Case Report

Rosai-Dorfman disease mimicking images of meningiomas: Two case reports and literature review

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INTRODUCTION

Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytic proliferative disorder classically as a massive cervical lymphadenopathy. However, over the years, extranodal locations were confirmed with the central nervous system involvement in less than 5% of cases, which is marked as a significant differential diagnosis of meningiomas, with which they are widely confused due to the similarity of their radiological images.

ABSTRACT

Background: Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytic proliferative disorder classically as a massive cervical lymphadenopathy. However, over the years, extranodal locations were confirmed with the central nervous system involvement in less than 5% of cases, which is marked as a significant differential diagnosis of meningiomas, with which they are widely confused due to the similarity of their radiological images.

Case Description: We report a 37-year-old man and 45-year-old man who were diagnosed with intracranial RDD but whose radiological images mimic meningiomas, requiring anatomopathological and tumor’s immunohistochemistry for definitive diagnosis. Moreover, a review of 184 publications with 285 cases of intracranial involvement of this disease was also performed, comparing these findings with those brought in the previous studies.

Conclusion: Intracranial Rosai-Dorfman tumors should always be remembered as differential diagnosis of meningiomas since they are similar radiologically and macroscopically. Once remembered and diagnosed, the lesion must be treated following the same pattern of resection done in meningiomas and, treatment’s differences will not occur in the surgical excision technique, but in complementary chemotherapy implementation, radiotherapy, and even with radiosurgery aid, depending on the case. Thus, it is possible to obtain better results than with just the isolated surgical procedure.

Keywords: Central nervous system, Histiocytosis, Magnetic resonance imaging, Meningioma, Rosai-Dorfman disease

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His first description dates back to 1965 by a French pathologist - Pierre Paul Louis Lucien Destombes, and 4 years later was exhaustively studied by Rosai and Dorfman, who analyzed 34 cases of the same disease under the name of sinus histiocytosis with massive lymphadenopathy. Classically, RDD presents as a bilateral cervical lymphadenopathy, but in 43% of patients the disease presents as an extranodal location. It is important to point out that RDD is a rare disease, with prevalence of approximately 1:200,000 people, with involvement of the central nervous system (CNS) occurring in less than 5% of cases of RDD and, within this portion, approximately 75% present themselves as intracranial lesions and 25% as spinal lesions. As for the age group, nodal RDD is more frequently observed in children and young adults (mean age 20.6 years), being more present in men (male/female ratio 1.4:1). However, when intracranial RDD is involved, the age group most affected changes, generally affecting adult men in the fourth and fifth decades of life (mean age 39.5 years).

In intracranial RDD, the most involved structures are the suprasellar region, cerebral convexity, parasagittal region, cavernous sinus, and petroclival region with infratentorial parenchymal lesions being the most frequent while supratentorial, intraventricular, and multilocal lesions are observed with significantly lower frequency. Radiologically, intracranial RDD is commonly confused with meningioma and requires tumor histopathology and immunohistochemistry for its definitive diagnosis. In anatomopathological examination of the lesions, histiocytes with large and discolored cytoplasm with large hypochromatic nucleus and prominent nucleolus are present. Emperipolysis is a useful finding, although not necessary for diagnosis. In immunohistochemical examination, histiocytes are positive for S-100 and CD-68 protein and negative for CD1a and increased doses in exacerbations, and he presented with pulsatile headache which was often disabling. A skull MRI was performed which revealed an expansive lesion in the left frontal region, requiring hospitalization, and use of prophylactic anticonvulsant. A microsurgery was performed for total resection of the brain tumor, which in the anatomopathological examination showed proliferation of histiocytes of ample cytoplasm and vesicular nuclei with prominent nucleoli, forming aggregates surrounded by lymphoplasmocytic infiltrate and with emperipolysis. The immunohistochemical examination demonstrated histiocytes positive for S-100 and CD-68 protein and negative for CD1a, thus confirming the diagnosis of RDD. A panel of mutations for solid tumors was also performed by Next-Generation Sequencing, with no relevant changes in the areas of interest of the analyzed genes. The patient was discharged 3 days later, remaining in follow-up until now well and without recurrence.

Second case report

Male patient, 45 years old, receives specialized neurological care with convergent strabismus and complaint of diplopia, headache, ringing in the left ear and hypoaacusis for 6 months. A gadolinium-contrast MRI examination was requested, which demonstrated a lesion in the petroclival region invading the cavernous sinus with extension into the posterior fossa, with contrast uptake compatible with

CASES REPORTS

First case report

Male patient, 37 years old, presented 4 years before with painless left supraclavicular adenomegaly, with progressive increase followed by intense pain in the left clavicle after physical activity. Imaging examinations demonstrated the presence of bone infiltration, supraclavicular and infracranial adenomegaly, as well as lesions in the orbit and cranial cap. Biopsy of supraclavicular lymph node confirmed lymphadenitis with massive sinus histiocytosis compatible with RDD, with immunohistochemical examination demonstrating CD68 and S100 positive and CD30 and CD1a negative. The patient initially presented an excellent response with corticoids using, noting significant regression of adenomegaly, and general improvement of symptoms. In the last year, however, he began to refer to migratory arthralgia with an increase in cervical adenomegalies, requiring the continuous use of corticoids and increased doses in exacerbations, and he presented with pulsatile headache which was often disabling. A skull MRI was performed which revealed an expansive lesion in the left frontal region, requiring hospitalization, and use of prophylactic anticonvulsant. A microsurgery was performed for total resection of the brain tumor, which in the anatomopathological examination showed proliferation of histiocytes of ample cytoplasm and vesicular nuclei with prominent nucleoli, forming aggregates surrounded by lymphoplasmocytic infiltrate and with emperipolysis. The immunohistochemical examination demonstrated histiocytes positive for S-100 and CD-68 protein and negative for CD1a, thus confirming the diagnosis of RDD. A panel of mutations for solid tumors was also performed by Next-Generation Sequencing, with no relevant changes in the areas of interest of the analyzed genes. The patient was discharged 3 days later, remaining in follow-up until now well and without recurrence.

Second case report

Male patient, 45 years old, receives specialized neurological care with convergent strabismus and complaint of diplopia, headache, ringing in the left ear and hypoaacusis for 6 months. A gadolinium-contrast MRI examination was requested, which demonstrated a lesion in the petroclival region invading the cavernous sinus with extension into the posterior fossa, with contrast uptake compatible with
meningioma. MRI also showed that the lesion reached the cervical region, descending through the petroclival portion, and bordering the clivus [Figure 3]. The patient was then submitted to a combined subtemporal and presigmoid route for partial resection – leaving only part of the lesion in the middle fossa [Figure 4] – of the possible meningioma, which after anatopathological analysis was concluded it was not a meningioma but a case of RDD. Then, continuous chemotherapy treatment with CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) and radiotherapy in specialized oncology services was started for 2 years and submitted to several sections throughout this period. The patient was operated on with tumor partial resection in 2005, remaining in follow-up until now without recurrence.

DISCUSSION

According to 285 RDD literature reviewed cases [1–5,7,8,10,12–24,26–39,41,44–48,50–56,68,70,72–96,98,99,102–111,113–115,117–123,126–129,130,132–135,137–142,144,146–148,150–154,157–161,163–175,177,181–185,187–191,193–196,198,200,202–210,212–215,217–221], added two newly cases presented in this study [Tables 1 and 2], we obtained a male/female ratio of 1.9:1 and a mean of 38.51 years and a median of 38 years of age, with 50% of patients aged between 26 and 53 years. The exclusively intracranial involvement was present in 77% (n = 221) of RDD cases, with a mean age of 39.5 years, while the exclusively spinal involvement was present in only 14% (n = 40) of them and with a mean age of 36.4 years. The mean age values obtained are quite close to the mean of 39.5 years described in the previous studies, mainly in cases with exclusive intracranial involvement. However, the 1.9:1 male/female ratio obtained in our review was much more remarkable in the prevalence of men than the 1:1.4 in the previous studies.

On the other hand, comparing the frequencies of intracranial or spinal involvement according to the systemic or non-systemic involvement of the disease, RDD with isolated CNS involvement, reported in 77% (n = 221) of all cases of CNS-RDD, showed that 84% (n = 186) of isolated cases of the CNS had exclusively intracranial involvement and only 10% (n = 23) had exclusively spinal involvement. As for systemics CNS-RDD cases, the exclusively intracranial involvement occurred in 56% (n = 37) of the cases, while the exclusively spinal involvement was present in 26% (n = 17) of them. Therefore, it should be noted that comparing RDD with isolated CNS involvement, systemic CNS-RDD has a lower prevalence of exclusively intracranial involvement and a greater involvement of the spinal cord; in 26% (n = 17) of the cases with systemic presentation there was exclusive involvement of the spinal and in approximately 18% (n = 12) of the cases there was intracranial and spinal involvement. Thus, it is interesting to note that when RDD has systemic involvement, spinal cord involvement is more frequent than in relation to RDD with exclusive CNS involvement, which may have different explanations, such as perhaps because of systemic disease focus origin, usually sinus and with massive lymphadenopathy in the region, be closer to the spinal cord, this will somehow facilitate the disease spread to this nearest neural structure. This would mean that, once systemic RDD
was present, it could spread more easily to any location in the CNS, without maintaining the preferential intracranial involvement of CNS exclusive cases.

The typical radiological findings of intracranial RDD show dural-based, extra-axial, well-circumscribed masses mimicking meningioma with MRI usually reveals multiple well-defined, dural-based or intraventricular, extra-axial masses with possible perilesional cerebral edema.\[80\] Intracranial RDD CT typically presents homogeneous hyperdense or isodense masses, but MRI is currently the optimal diagnostic modality for evaluating lesions. On T1-weighted images, the lesions usually appear as isointense or hyperintense masses with clear borders relative to the peripheral brain parenchyma\[56,150,187,196,201\] and possible perilesional cerebral edema hypo or isointense.\[80\] While on T2-weighted images, the lesions usually appear as isointense masses with possible intrallesional hypointense foci\[56,150,187,196,201\] although studies have described rather low signal intensity on in this type of image.\[7,87,196\] On the other hand, meningiomas on T2-weighted MR images show low to high signal

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**Table 1:** Characteristics of CNS-RDD cases according to present and previous reports from references \[1-5,7-9,10,12-24,26-39,41,44-48,50-54,68,70,72-96,99,102-111,113-115,117-123,126-129,130,132-135,137-142,144-146,150-154,157-161,163-1-75,177,178,181-185,187-191,193-196,198,200,202-210,212-215,217-221\]

| Characteristic     | RDD with isolated CNS involvement (n=221) | Systemic RDD with CNS involvement (n=66) | Total RDD with CNS involvement (n=287) |
|--------------------|------------------------------------------|------------------------------------------|---------------------------------------|
| Age – yr           | Mean 38.93                                | 37.11                                    | 38.51                                 |
|                    | Range 1–79                                | 3–78                                     | 1–79                                  |
| Sex                | Female 74                                 | 25                                       | 99                                    |
|                    | Male 147                                  | 41                                       | 188                                   |
| Location           | Intracranial 186                         | 37                                       | 223                                   |
|                    | Spine 23                                  | 17                                       | 40                                    |
|                    | Intracranial and spine 12                | 12                                       | 24                                    |

RDD: Rosai-Dorfman disease, CNS: Central nervous system

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**Figure 3:** Magnetic resonance imaging preoperated showing expansive lesion in the left petroclival region with left to right mass effect, invading the cavernous sinus with extension into the posterior fossa and reach the cervical region, descending through the petroclival portion, bordering the clivus. Lesion with contrast enhances compatible with meningioma. (a) Coronal gadolinium-enhanced image (GEI). (b and c) Axial GEI.

**Figure 4:** (a-c) Magnetic resonance imaging at the post operated showed partial resection of the left petroclival tumor with remaining lesion in the middle fossa. Axial gadolinium-enhanced image.
Table 2: Central nervous system involvement in 287 cases of Rosai-Dorfman disease.

| Case. No. | Authors et al., 2015 | Sex | Age (years) | Location | Exact location |
|-----------|----------------------|-----|-------------|----------|---------------|
| 1         | Tatit et al.         | Male | 37          | Intracranial | L] frontal   |
| 2         | Our study - case 2   | Male | 41          | Intracranial | Petrodival, cavernous sinus, posterior fossa and cervical region and spine |
| 3         | Abdel-Razek et al., 2013 | Male | 43          | Intracranial | [R] and [L] frontal, falx cerebri, anterior clinoid process and [R] petrous bone |
| 4         | Abdel-Razek et al., 2013 | Female | 38       | Intracranial | [R] parietal parasagittal |
| 5         | Adeleye et al., 2010 | Male | 61          | Intracranial | [L] tentorial dural and supra/infratentorial compartments |
| 6         | Adeleye et al., 2010 | Male | 38          | Intracranial | Tuberculum sella dural, [R] parietal and [L] foramen magnum |
| 7         | Agnoletto et al., 2019 | Male | 46          | Intracranial | [R] cerebellar hemisphere |
| 8         | Alimli et al., 2016  | Male | 1           | Intracranial | [L] parieto-occipital |
| 9         | Amagasa et al., 2001 | Male | 42          | Intracranial | Pituitary fossa |
| 10        | Andriko et al., 2001 | Male | 51          | Spine      | Spinal canal, thoracic epidural |
| 11        | Andriko et al., 2001 | Male | 50          | Intracranial | Base of skull, [L] petroclinoid ligament |
| 12        | Andriko et al., 2001 | Male | 42          | Spine      | Spinal canal, epidural space, T6–T8 |
| 13        | Andriko et al., 2001 | Male | 22          | Intracranial | [L] fronto-temporal and [R] parietal |
| 14        | Andriko et al., 2001 | Female | 63         | Intracranial | [R] frontal |
| 15        | Andriko et al., 2001 | Female | 25         | Intracranial | Falx cerebri and sagittal sinus |
| 16        | Andriko et al., 2001 | Female | 31         | Intracranial | Falx cerebri |
| 17        | Andriko et al., 2001 | Male | 35          | Spine      | Spinal cord, intramedullary, T4–T5 |
| 18        | Andriko et al., 2001 | Male | 62          | Intracranial | [R] parietal |
| 19        | Andriko et al., 2001 | Female | 43         | Intracranial | [R] parietal |
| 20        | Andriko et al., 2001 | Male | 24          | Intracranial | [L] occipital dura |
| 21        | Antuña Ramos et al., 2012 | Female | 10     | Intracranial | [R] frontal, [L] middle cerebellar peduncle, [R] ventral pons and and spine spinal cord, intramedullary, T9-T10 |
| 22        | Arnao et al., 2016   | Male | 70          | Intracranial | [L] fronto-parietal-temporal |
| 23        | Kumar et al., 2014   | Male | 43          | Intracranial | [R] fronto-temporal |
| 24        | Asai et al., 1988    | Male | 39          | Intracranial | [L] occipital subarachnoid space |
| 25        | Baassiri et al., 2020 | Male | 54          | Intracranial | Foramen magnum, the base of the odontoid process, [L] and spine anterolateral to the spinal cord and medullary spinal junction surrounding the [L] vertebral artery |
| 26        | Battal et al., 2017  | Male | 18          | Intracranial | [R] and [L] choroid plexus of ventricular atria, tentorial dural, falx cerebri and planum sphenoidale dural |
| 27        | Bernard et al., 1999 | Female | 10       | Spine | Spinal, epidural and subdural, L3-L5 |
| 28        | Beros et al., 2011   | Male | 41          | Intracranial | [R] cerebellar hemisphere |
| 29        | Bertero et al., 2018 | Female | 68         | Intracranial | [L] temporal |
| 30        | Lima et al., 2019    | Female | 60         | Intracranial | [L] parietal |
| 31        | Bhandari et al., 2006 | Female | 23       | Spine | Intradural extramedullary space, C3-C4 to C5-C6, T1-T2 to T4 and T5 |
| 32        | Bhat et al., 2015    | Male | 38          | Intracranial | [L] parietal convexity |
| 33        | Bhattacharyee et al., 1992 | Male | 78         | Intracranial | Suprasellar and planum sphenoidale |
| 34        | Bing et al., 2009    | Female | 32        | Intracranial | [L] parieto-occipital |
| 35        | Boissaud-Cooke et al., 2020 | Male | 52         | Intracranial | [R] frontal |
| 36        | Brandsma et al., 2003 | Female | 36        | Intracranial | [L] cerebellopontine angle, [R] and [L] supratentorial parietal extension |
| 37        | Brandsma et al., 2003 | Male | 41          | Intracranial | [R] occipital |
| 38        | Brandsma et al., 2003 | Female | 33        | Intracranial | Infratentorial, [R] and [L] internal auditory meatus and ganglion Gasseri, and cervical spinal cord |
| 39        | Breiner et al., 2013 | Female | 63        | Intracranial | [R] and [L] periventricular white matter and corpus callosum |
| 40        | Buchino et al., 1982 | Male | 13          | Intracranial | Base of skull and spinal cord, C6-T12 and spine |
| 41        | Camp et al., 2012    | Female | 31        | Intracranial | [L] frontal, [R] parietal and [R] frontal white matter |
| 42        | Cao et al., 2011     | Male | 35          | Intracranial | Tentorial dural in the [R] trigone and [R] lateral ventricle |

(Contd...)
Table 2: (Continued)

| Case. No. | Authors | Sex   | Age (years) | Location       | Exact location                                                                 |
|-----------|---------|-------|-------------|----------------|-------------------------------------------------------------------------------|
| 43        | Carey and Case, 1987 | Male   | 35          | Intracranial   | Parasagittal crossing the midline                                             |
| 44        | Castellano-Sanchez and Brat, 2003 | Female | 37          | Intracranial   | [L] parietal                                                                  |
| 45        | Catalucci et al., 2012 | Male   | 57          | Intracranial   | Anterior, middle and posterior cranial fossa                                  |
| 46        | Cavuoto et al., 2011 | Male   | 25          | Intracranial   | Pons, cerebral and cerebellar peduncles, [L] temporal, [L] frontal, [L] globus pallidus, [L] optic nerve, optic chiasm and proximal optic tracts |
| 47        | Chan et al., 1985 | Female | 7           | Spine          | Spinal cord, C5-C6                                                            |
| 48        | Chen, 2003          | Male   | 70          | Intracranial   | [R] parietal and suprasellar                                                  |
| 49        | Chen, 2003          | Male   | 62          | Spine          | Sacral canal                                                                  |
| 50        | Chivukula et al., 2014 | Female | 66          | Intracranial   | Hypothalamus                                                                  |
| 51        | Clark and Berry, 1996 | Female | 38          | Intracranial   | [L] parietal                                                                  |
| 52        | Cooper and Jenrette, 2012 | Male   | 6           | Intracranial   | [L] temporal, diffuse meningeal infiltration and spine                         |
|           |          |       |             |                | and spine                                                                     |
| 53        | Cunliffe et al., 2009 | Male   | 33          | Intracranial   | Skull base, with involvement of the sella and suprasella cistern             |
| 54        | Deodhare et al., 1998 | Male   | 41          | Intracranial   | [L] parieto-occipital                                                         |
| 55        | Deshayes et al., 2013 | Male   | 38          | Intracranial   | [L] parieto-occipital                                                         |
| 56        | Deshayes et al., 2013 | Female | 51          | Intracranial   | Floor of the third ventricle                                                  |
| 57        | Di Rocco et al., 2007 | Female | 13          | Intracranial   | [L] frontal                                                                   |
| 58        | El Majdoub et al., 2009 | Female | 17          | Spine          | Spinal cord, intradural extramedullary space, T1-T4                          |
| 59        | El Moghaddam et al., 2009 | Male   | 10          | Intracranial   | [L] white matter and basal ganglia                                           |
| 60        | El Molla et al., 2014 | Male   | 76          | Spine          | Spinal cord, intramedullary, C2-C3                                           |
| 61        | Forest et al., 2014 | Male   | 41          | Intracranial   | Frontal and temporal dural-based                                              |
| 62        | Forest et al., 2014 | Female | 35          | Intracranial   | Orbitary and optic pathways                                                  |
| 63        | Cooper and Jenrette, 2014 | Male   | 38          | Intracranial   | Multiple dural-based lesions                                                  |
| 64        | Fortea et al., 2008 | Female | 53          | Intracranial   | [L] occipital cortex and cerebellar hemispheres                               |
| 65        | Foucar et al., 1982 | Male   | 21          | Intracranial   | [L] cerebellopontine angle, epidural                                         |
| 66        | Foucar et al., 1982 | Female | 59          | Spine          | Spinal, T8                                                                    |
| 67        | Foucar et al., 1982 | Male   | 53          | Spine          | Spinal, epidural, C7-T3                                                       |
| 68        | Foucar et al., 1982 | Female | 12          | Spine          | Spinal, epidural, C2, C5-T2                                                   |
| 69        | Foucar et al., 1982 | Male   | 4           | Spine          | Spinal, subdural                                                             |
| 70        | Foucar et al., 1982 | Female | 55          | Intracranial   | Frontal, epidural                                                            |
| 71        | Foucar et al., 1982 | Female | 28          | Intracranial   | [L] parietal, epidural                                                        |
| 72        | Foucar et al., 1982 | Female | 11          | Spine          | Spinal, epidural: T3-T9, L5-S1                                                |
| 73        | Franco-Paredes and Martin, 2002 | Female | 57          | Intracranial   | Diffuse leptomeningeal                                                        |
| 74        | Friedman et al., 1984 | Male   | 32          | Intracranial   | Suprascellar                                                                  |
| 75        | Fukushima et al., 2011 | Female | 33          | Intracranial   | Cavernous sinus                                                               |
| 76        | Gaetani et al., 2000 | Female | 67          | Intracranial   | Cerebellar, Intracerebellar                                                  |
| 77        | Geara et al., 2004 | Male   | 3           | Intracranial   | [L] cerebellar hemisphere, tentorium, fourth ventricle, trigeminal nerves at their origins from the pons, choroid plexus in the [R] and [L] trigone |
| 78        | Ghosal et al., 2007 | Male   | 26          | Intracranial   | [R] parietal convexity                                                        |
| 79        | Gies et al., 2005  | Female | 40          | Intracranial   | [L] parietal                                                                  |
| 80        | Griffiths et al., 2004 | Male   | 9           | Intracranial   | [R] frontal                                                                   |
| 81        | Gui et al., 2014 | Male   | 60          | Intracranial   | Corpus callosum                                                               |
| 82        | Gui et al., 2014 | Female | 54          | Intracranial   | [L] frontal and parietal                                                      |
| 83        | Gupta et al., 2015 | Female | 20          | Intracranial   | [L] and [R] bulky intraconal                                                  |
| 84        | Gupta et al., 2006 | Male   | 15          | Intracranial   | Parasellar and petroclival                                                   |
| 85        | Gupta et al., 2011 | Male   | 14          | Intracranial   | [L] skull base                                                                |
| 86        | Haas et al., 1978   | Female | 12          | Spine          | Epidural, C2 and C5-T2                                                        |
| 87        | Hadjipanayis et al., 2003 | Male   | 52          | Intracranial   | [L] cavernous sinus and petroclival                                           |
| 88        | Halelidad et al., 2007 | Female | 65          | Intracranial   | [L] temporal convexit                                                         |

(Contd...)
Table 2: (Continued)

| Case. No. | Authors                     | Sex  | Age (years) | Location                     | Exact location                                                                 |
|----------|-----------------------------|------|-------------|------------------------------|--------------------------------------------------------------------------------|
| 89       | Hargett and Bassett, 2005   | Female | 29          | Spine                        | Spinal cord, T5-T9                                                             |
| 90       | Hashimoto et al., 2014      | Male  | 53          | Intracranial                 | [R] frontotemporal and sphenoid wing                                           |
| 91       | Hinduja et al., 2009        | Male  | 42          | Intracranial                 | [L] orbital apex, middle cranial fossa and cavernous sinus                     |
| 92       | Hollowell et al., 2000      | Male  | 78          | Spine                        | Spinal cord, C4-C8                                                             |
| 93       | Hong Cheng et al., 2017     | Male  | 64          | Intracranial                 | [R] frontal                                                                   |
| 94       | Hong et al., 2016           | Female | 59          | Intracranial                 | Intraparenchymal cerebellar                                                   |
| 95       | Huang et al., 1998          | Male  | 38          | Intracranial                 | Parietal, dural-based                                                         |
| 96       | Huang et al., 2016          | Male  | 55          | Spine                        | Spine, epidural, T1-T9                                                        |
| 97       | Huang et al., 2016          | Male  | 40          | Spine                        | Spine, epidural, C3-C6                                                        |
| 98       | Huang et al., 2016          | Female | 14          | Spine                        | Spine, intervertebral foramen, S1-S2                                           |
| 99       | Huang et al., 2016          | Male  | 43          | Spine                        | Spine, epidural, C5-C6                                                        |
| 100      | Idir et al., 2011           | Female | 28          | Intracranial                 | [L] frontal                                                                   |
| 101      | Imada et al., 2015          | Female | 68          | Intracranial                 | Brainstem                                                                    |
| 102      | Jayaram et al., 2020        | Male  | 24          | Spine                        | Intraspinal, epidural, C7-T4                                                  |
| 103      | Jiang and Jiang, 2018       | Male  | 39          | Intracranial                 | [R] fronto-parietal and [L] frontal meningeal                                  |
| 104      | Jiang and Jiang, 2018       | Male  | 53          | Intracranial                 | [L] parietal, temporal and occipital meningeal                                 |
| 105      | Jiang and Jiang, 2018       | Female | 9           | Intracranial                 | [R] parietal meningeal                                                        |
| 106      | Johnston et al., 2009       | Male  | 14          | Intracranial                 | [R] cerebellum                                                               |
| 107      | Jones and Rueda-Pedraza, 1997| Male  | 34          | Spine                        | Intramedullary spinal cord                                                   |
| 108      | Joshi et al., 2019          | Male  | 58          | Intracranial                 | [L] medial occipito-parietal                                                 |
| 109      | Joshi et al., 2019          | Female | 42          | Intracranial                 | [R] parietal dural-based                                                       |
| 110      | Joshi et al., 2019          | Male  | 40          | Intracranial                 | [R] parietal dural based                                                      |
| 111      | Joshi et al., 2019          | Male  | 46          | Intracranial                 | [R] parietal dural based                                                      |
| 112      | Joshi et al., 2019          | Male  | 36          | Spine                        | Spinal column                                                               |
| 113      | Joubert et al., 2013        | Male  | 38          | Intracranial                 | Frontal falk cerebri, lateral ventricles and [L] and [R] tentorium cerebelli |
| 114      | Jurić et al., 2003          | Male  | 39          | Intracranial                 | [R] temporal                                                                  |
| 115      | Kaminsky et al., 2005       | Male  | 32          | Intracranial                 | Petroclival, cavernous sinuses, suprasellar and anterior cranial fossa       |
| 116      | Kattner et al., 2000        | Male  | 33          | Intracranial                 | [R] parasagittal                                                              |
| 117      | Katz et al., 1993           | Male  | 20          | Intracranial                 | Foramen magnum, posterior fossa and cervical Region                             |
| 118      | Kayali et al., 2004         | Male  | 31          | Intracranial                 | [L] temporal                                                                  |
| 119      | Kelly et al., 1999          | Female | 45          | Intracranial                 | [L] cavernous sinus, posterior pituitary gland, optic chiasma, third ventricle and spinal cord, T2 |
| 120      | Kessler et al., 1976        | Male  | 53          | Spine                        | Spinal Cord, C7-T3                                                            |
| 121      | Kidd et al., 2006           | Female | 37          | Intracranial                 | Anterior cranial fossa, [R] cerebellopontine angle, clivus and spinal cord, C5 |
| 122      | Kidd et al., 2006           | Male  | 68          | Intracranial                 | Suprasellar region and [R] parasellar region                                  |
| 123      | Kim et al., 2011            | Male  | 39          | Intracranial                 | [R] frontal, temporal, cerebellopontine angle, [L] clinoidal, pteroclival, [R] [L] Meckel's Cave |
| 124      | Kim et al., 1995            | Male  | 50          | Intracranial                 | [R] parietal convexity                                                        |
| 125      | Kitai et al., 2001          | Male  | 36          | Intracranial                 | [L] occipital convexity                                                       |
| 126      | Kitai et al., 2001          | Female | 42          | Intracranial                 | [R] frontal base                                                              |
| 127      | Kitai et al., 1996          | Male  | 25          | Intracranial                 | Tentorium                                                                    |
| 128      | Kong et al., 2019           | Male  | 10          | Intracranial                 | Posterior pituitary                                                           |
| 129      | Konishi et al., 2003        | Female | 68          | Intracranial                 | [L] frontal region                                                            |
| 130      | Krishnamoorthy et al., 2011 | Male  | 51          | Intracranial                 | [R] frontal region                                                            |
| 131      | Kumar et al., 2008          | Male  | 45          | Intracranial                 | [L] parietal and temporal convexity                                           |
| 132      | Lauwers et al., 2000        | Female | 28          | Intracranial                 | Central nervous system, meninges                                              |
| 133      | Le Guenno et al., 2012      | Male  | 57          | Intracranial                 | [L] frontotemporal                                                            |
| 134      | Leung et al., 2003          | Male  | 35          | Intracranial                 | [L] parietal convexity and [R] tentorium                                      |

(Contd...)
### Table 2: (Continued)

| Case. No. | Authors               | Sex  | Age (years) | Location | Exact location                                                                 |
|----------|-----------------------|------|-------------|----------|--------------------------------------------------------------------------------|
| 135      | Li et al., 2012       | Male | 40          | Intracranial | [R] parietal                                                                       |
| 136      | Löhr et al., 1995     | Female | 40    | Spine | Cervical region                                                                 |
| 137      | Lopez and Estes, 1989 | Male | 35          | Intracranial | [L] cavernous sinus and Meckel's cave, [L] cerebellopontine angle cistern. Internal auditory canal, middle cranial fossa. |
| 138      | Lou et al., 2012      | Male | 27          | Intracranial | Suprasellar and intrasellar region                                               |
| 139      | Lou et al., 2012      | Male | 29          | Intracranial | Suprasellar and intrasellar region                                               |
| 140      | Lou et al., 2012      | Female | 26     | Intracranial | Suprasellar region                                                               |
| 141      | Lou et al., 2012      | Male | 14          | Intracranial | Suprasellar region                                                               |
| 142      | Lou et al., 2012      | Female | 22     | Intracranial | Suprasellar region, midbrain and cerebellum                                     |
| 143      | Lu et al., 2012       | Female | 48     | Intracranial | [L] frontal area                                                                  |
| 144      | Lu and Guo, 2010      | Male | 34          | Intracranial | [L] frontal lobe and corona radiata                                               |
| 145      | Lüdemann et al., 2015 | Male | 2          | Intracranial | [R] and [L] frontal lobe, [L] ventricle                                           |
| 146      | Lungenre et al., 2009 | Female | 2      | Intracranial | Frontal convexity and parafalcine region                                           |
| 147      | Luo et al., 2017      | Male | 41          | Intracranial | Fourth ventricle, [R] and [L] posterior horn of lateral ventricles, [L] parasellar and cerebellopontine angle |
| 148      | Luo et al., 2017      | Female | 31     | Intracranial | Cavernous sinus, foramen magnum, [R] parasellar, [R] and [L] sphenoidal crest and fronto-parietal meningeal |
| 149      | Luo et al., 2017      | Female | 73     | Intracranial | [L] posterior cranial fossa                                                        |
| 150      | Lutterbach et al., 2003 | Female | 60    | Intracranial | [L] hemisphere                                                                     |
| 151      | Maiti et al., 2011    | Female | 19     | Spine | Extradural lesion, C3-C6                                                           |
| 152      | McPherson et al., 2006 | Male | 53        | Intracranial | Skull base, planum sphenoidale and tuberculum sella                               |
| 153      | Kim et al., 2011      | Male | 39          | Intracranial | [R] frontal, temporal, and cerebellopontine angle. [L] clinoidal and spine         |
| 154      | Miletic et al., 2008  | Female | 8       | Intracranial | [L] occipital horn of lateral ventricle. [R] and [L] frontal periventricular      |
| 155      | Mir et al., 1985      | Male | 33          | Intracranial | Suprasellar                                                                       |
| 156      | Mirra et al., 1983    | Female | 11     | Intracranial | [R] frontal area                                                                  |
| 157      | Mirra et al., 1983    | Female | 38     | Spine | Spinal cord, C7                                                                   |
| 158      | Morandi et al., 2000  | Female | 22     | Intracranial | Fourth ventricle                                                                  |
| 159      | Nalini et al., 2012   | Male | 18          | Intracranial | [R] and [L] Parasellar region. Tuberculum sella, planum sphenoidale, tentorium, clivus and cervical spinal canal |
| 160      | Nassif and Boulos, 2015 | Male | 42        | Intracranial | N/A                                                                               |
| 161      | Natarajan et al., 2000 | Female | 45     | Intracranial | [R] frontal lobe                                                                  |
| 162      | Ng and Poon, 1995     | Male | 22          | Intracranial | Posterior pituitary                                                                |
| 163      | Olsen et al., 1988    | Male | 69          | Intracranial | [L] temporal lobe                                                                  |
| 164      | Osenbach, 1996        | Male | 35          | Spine | Spinal cord, T4-T5                                                                |
| 165      | Mahzoni et al., 2012  | Male | 33          | Intracranial | [L] parietal region                                                               |
| 166      | Panicker et al., 1996 | Female | 58     | Intracranial | Middle cranial fossa, dural-based                                                 |
| 167      | Parmar et al., 2013   | Female | 64     | Intracranial | Planum sphenoidale, clivus, [L] temporal lobe and spine, C5-C6 and spine          |
| 168      | Patwardhan and Goel, 2018 | Female | 40     | Intracranial | Occipital horn of the lateral ventricle                                            |
| 169      | Petzold et al., 2001  | Male | 47          | Intracranial | Cerebellopontine angle, foramen magnum, chiasmatic cistern, planum sphenoidale and [R] parafalcine region |
| 170      | Prayson and Rowe, 2014 | Male | 29        | Intracranial | [R] posterior parietal convexity                                                  |
| 171      | Purav et al., 2005    | Male | 18          | Intracranial | [L] Meckel's cave and spine, C2-C3 level and spine                                |
| 172      | Purav et al., 2005    | Male | 23          | Intracranial | [L] parietal                                                                       |
| 173      | Purav et al., 2005    | Male | 31          | Intracranial | [L] parietal                                                                       |
| 174      | Purav et al., 2005    | Male | 37          | Intracranial | [R] parieto-ccipital                                                              |
| 175      | Purav et al., 2005    | Male | 37          | Intracranial | [L] parietal                                                                       |
| 176      | Purav et al., 2005    | Male | 39          | Intracranial | [L] frontal                                                                       |
| 177      | Purav et al., 2005    | Male | 50          | Intracranial | Multiple intraparenchymal                                                        |
| 178      | Purav et al., 2005    | Female | 51    | Intracranial | [L] frontal convexity                                                             |

(Contd...)
Table 2: (Continued)

| Case. No. | Authors | Sex  | Age (years) | Location | Exact location |
|-----------|---------|------|-------------|----------|---------------|
| 179       | Purav et al., 2005 | Male | 56          | Intracranial | [R] parietal parasagittal |
| 180       | Purav et al., 2005 | Female | 60         | Intracranial | [R] parietal convexity |
| 181       | Qin et al., 2019 | Male | 43          | Intracranial and spine | Frontal falx, parietal falx, tentorium cerebelli and spinal, T3 |
| 182       | Raslan et al., 2011 | Male | 50          | Intracranial and spine | Skull base, convex and spinal, meningeal |
| 183       | Raslan et al., 2011 | Male | 54          | Intracranial and spine | Sellar and suprasellar, [R] and [L] cerebellopontine angle, and cervical canal, epidural |
| 184       | Raslan et al., 2011 | Female | 50         | Intracranial | Pituitary |
| 185       | Raslan et al., 2011 | Female | 57         | Spine | Spinal, meningeal, T9 |
| 186       | Resnick et al., 1996 | Male | 38          | Intracranial | [L] cerebellopontine angle |
| 187       | Richardson et al., 2018 | Female | 64         | Intracranial | [R] and [L] cerebellar hemispheres, basal ganglia, and corpus callosum |
| 188       | Rocha-Maguey et al., 2016 | Female | 27         | Spine | Spinal cord, intramedullary, C7-T1 |
| 189       | Rotondo et al., 2010 | Female | 63         | Intracranial | spread to the pars tuberalis, the lower portion of the pituitary stalk and to the adjacent dura |
| 190       | Russo et al., 2009 | Male | 72          | Intracranial | Pituitary |
| 191       | Russo et al., 2009 | Male | 57          | Intracranial | [R] and [L] frontal |
| 192       | Said et al., 2011 | Male | 74          | Intracranial | [R] temporal |
| 193       | Sakai et al., 1998 | Male | 60          | Intracranial | [L] cerebellopontine angle, [R] temporal fossa and convexity, [R] and [L] frontal convexity |
| 194       | Sandoval-Sus et al., 2014 | Female | 32         | Intracranial | Extraaxial brainstem |
| 195       | Sandoval-Sus et al., 2014 | Male | 51          | Intracranial | Extraaxial brainstem |
| 196       | Sandoval-Sus et al., 2014 | Male | 53          | Intracranial | Extraaxial brainstem and spinal, extramedullary and intramedullary |
| 197       | Sandoval-Sus et al., 2014 | Male | 18          | Intracranial and spine | Extraaxial brainstem and spinal, extramedullary and intramedullary |
| 198       | Sandoval-Sus et al., 2014 | Male | 38          | Intracranial | Extraaxial brainstem and cerebral |
| 199       | Sandoval-Sus et al., 2014 | Male | 60          | Intracranial | Extraaxial cerebral |
| 200       | Sato et al., 2003 | Female | 59         | Intracranial | Suprasellar region, [R] temporal convexity, [L] frontal convexity and cerebello-pontine angle, and spine, C5 level |
| 201       | Schmidt et al., 2004 | Male | 4           | Intracranial and spine | [R] and [L] retro-orbita and spine, epidural, L1-S2 |
| 202       | Scumpia et al., 2009 | Male | 22          | Intracranial | [R] middle cranial fossa |
| 203       | Seydinejad et al., 2007 | Female | 43         | Intracranial | Anterior and posterior cranial fossa, and spine, C5–C6 level |
| 204       | Shah et al., 2020 | Female | 63         | Intracranial | Cavernous sinus and superior orbital fissure |
| 205       | Shaver et al., 1993 | Male | 5           | Intracranial | [L] cavernous sinus, with extension over the tentorial margin into the posterior fossa |
| 206       | Shuangshoti et al., 1999 | Female | 55         | Intracranial | [R] fronto-parietal |
| 207       | Siadati et al., 2001 | Female | 48         | Intracranial | [L] parieto-occipital |
| 208       | Simos et al., 1998 | Male | 62          | Intracranial | [L] parietal, with base of attachment along the falx |
| 209       | Siu et al., 2015 | Male | 60          | Intracranial | [L] anterior cranial fossa |
| 210       | Song et al., 1989 | Male | 30          | Intracranial | Frontal, middle and posterior fossa |
| 211       | Sundaram et al., 2005 | Male | 35          | Intracranial | Dural-based lesions |
| 212       | Sundaram et al., 2005 | Male | 35          | Intracranial | Dural-based lesions |
| 213       | Sundaram et al., 2005 | Female | 35 | Intracranial | Dural-based lesions |
| 214       | Symss et al., 2010 | Male | 21          | Intracranial | Infratentorial, extending on both sides along the tentorium, up to the cavernous sinuses |
| 215       | Symss et al., 2010 | Male | 35          | Intracranial | [R] and [L] frontal dural based lesion with involvement of the falx |
| 216       | Symss et al., 2010 | Female | 17         | Intracranial | Suprasellar region |
| 217       | Tan et al., 2018 | Male | 66          | Intracranial | [L] temporal |

(Contd...)
Table 2: (Continued)

| Case No. | Authors                            | Sex   | Age (years) | Location                       | Exact location                                                                 |
|----------|------------------------------------|-------|-------------|--------------------------------|--------------------------------------------------------------------------------|
| 218      | Tanboon et al., 2003               | Male  | 22          | Intracranial                   | Sellar region                                                                 |
| 219      | Tavangar et al., 2006              | Male  | 79          | Intracranial                   | [L] parasagittal region                                                       |
| 220      | Thee et al., 2008                  | Female| 56          | Intracranial                   | [R] fronto-parietal convexity                                                |
| 221      | Tian et al., 2015                  | Male  | 6           | Intracranial                   | Frontal falx cerebri, lateral ventricles and both sides of the tentorium     |
| 222      | Tian et al., 2015                  | Male  | 17          | Intracranial                   | Falx cerebri and midline in both parietal lobes                              |
| 223      | Tian et al., 2015                  | Male  | 26          | Intracranial                   | [R] middle and posterior fossa                                               |
| 224      | Tian et al., 2015                  | Female| 49          | Intracranial                   | Falx cerebri and midline in both parietal lobes                              |
| 225      | Tian et al., 2015                  | Male  | 68          | Intracranial                   | [R] frontal                                                                  |
| 226      | Tian et al., 2015                  | Male  | 7           | Intracranial                   | Multiple intracranial lesions                                                |
| 227      | Tian et al., 2015                  | Male  | 40          | Spine                          | Spine, C3-C5, C6 level                                                       |
| 228      | Tian et al., 2015                  | Male  | 43          | Spine                          | Spine, C5-C6 level                                                           |
| 229      | Wu and Xu, 2014                    | Female| 60          | Intracranial                   | [R] occipital lobe and the [R] cerebellar hemisphere                         |
| 230      | Tomio et al., 2012                 | Male  | 53          | Intracranial                   | [R] and [L] frontal lobe with a base of attachment along the falx             |
| 231      | Triana-Pérez et al., 2011          | Male  | 40          | Intracranial                   | [R] parieto-occipital extending into the posterior fossa                      |
| 232      | Tripathi et al., 2017              | Female| 7           | Intracranial                   | [R] and [L] third, fifth, sixth, seventh and eighth cranial nerves, and spine |
| 233      | Trudel, 1984                       | Male  | 28          | Intracranial                   | [L] middle cranial fossa                                                     |
| 234      | Tu et al., 2017                    | Male  | 41          | Spine                          | Spine, T2-T3 level with dura tail sign                                       |
| 235      | Tubbs et al., 2005                 | Male  | 13          | Spine                          | Cranio cervical junction                                                     |
| 236      | Türe et al., 2004                  | Male  | 29          | Intracranial                   | [R] cavernous sinus, reaching the rostrum along the interhemispheric fissure  |
| 237      | Varan et al., 2015                 | Male  | 5           | Intracranial                   | Pontomesencephalic junction                                                  |
| 238      | Walker et al., 2011                | Female| 54          | Intracranial                   | Multiple dural-based lesions                                                 |
| 239      | Wang et al., 2001                  | Female| 19          | Intracranial                   | Infratemporal fossae                                                         |
| 240      | Wang et al., 2001                  | Male  | 38          | Intracranial                   | [R] frontal and [R] temporal and spine                                        |
| 241      | Wang et al., 2010                  | Male  | 22          | Intracranial                   | [L] parietal                                                                 |
| 242      | Wang et al., 2010                  | Female| 40          | Intracranial                   | [L] middle fossa                                                             |
| 243      | Wang et al., 2010                  | Male  | 38          | Intracranial                   | [L] petrous orbit                                                            |
| 244      | Wang et al., 2010                  | Male  | 47          | Intracranial                   | [L] petrous region                                                            |
| 245      | Wang et al., 2010                  | Male  | 58          | Spine                          | Spine, T8-T10 level                                                          |
| 246      | Wang et al., 2010                  | Male  | 26          | Intracranial                   | [R] occipital                                                                |
| 247      | Warnier et al., 2012               | Male  | 6           | Intracranial                   | Meningeal infiltrate to the cortex and spine and spine                       |
| 248      | Wen et al., 2019                   | Female| 54          | Intracranial                   | [R] frontal                                                                  |
| 249      | Wen et al., 2019                   | Male  | 40          | Intracranial                   | [L] occipital                                                                |
| 250      | Wen et al., 2019                   | Male  | 54          | Intracranial                   | Falx cerebri                                                                 |
| 251      | Woodcock et al., 1999              | Female| 15          | Intracranial                   | Pituitary infundibulum and around optic chiasm and lamina terminalis          |
| 252      | Wrzolek and Zagzag, 2002           | Male  | 38          | Intracranial                   | [L] frontal                                                                  |
| 253      | Wrzolek and Zagzag, 2002           | Female| 69          | Intracranial                   | [L] tentorial                                                                |
| 254      | Wu et al., 2014                    | Male  | 43          | Spine                          | Spine, C3-C6 level                                                           |
| 255      | Wu et al., 2001                    | Male  | 35          | Intracranial                   | [L] temporal and [L] occipital                                               |
| 256      | XiaoWen et al., 2010               | Male  | 38          | Intracranial                   | Skull base, supra-sellar region, basal cistern, tentorium, bilateral cavernos |
| 257      | XiaoWen et al., 2010               | Male  | 14          | Intracranial                   | Petroclival, [L] cavernous sinus                                            |
MRI spectroscopy meningiomas have been shown to show elevated lipid and N-acetyl aspartate peaks, suggestive of granulomatous inflammatory pathology, and a raised choline peak.\[199\] Furthermore, perfusion MRI imaging can provide useful information on menigioma vascularity which is not available from conventional MRI. Measurement of maximal rCBV and corresponding rMTE values in the peritumoral edema is useful in the preoperative differentiation between benign and malignant menigiomas\[216\] and the relatively low rCBV perfusion values in CNS-RDD.\[199\] Regarding to the neuroimaging, the best diagnostic clues for diagnosing CNS-RDD appear to be represented by hypo-isointensity in the T2-weighted sequences and the relatively low rCBV perfusion values, likely due to abundant fibrous tissue; however, these findings are not specific and not always present, and the final diagnosis is often still histological.\[199\] Therefore, preoperative radiological findings using current MRI sequences are difficult to distinguish between menigiomas and RDD; however, it has already been described that the absence of hyperostosis, bony erosion, or calcification – characteristically absent in the RDD\[177\] – should suggest RDD as a differential diagnosis of menigiomas.\[150\]

As for the two reported cases of RDD, they were very similar to the expected age group and sex grouping, according

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**Table 2: (Continued)**

| Case No. | Authors | Sex | Location | Exact location |
|----------|---------|-----|----------|---------------|
| 261      | Yao et al., 2013 | Female | 12 | Spine | Spine, C4-C5 level |
| 262      | Yetiser et al., 2004 | Male | 7 | Intracranial | [R] parietal convexity near to interhemispheric fissure |
| 263      | Yetiser et al., 2004 | Male | 6 | Intracranial | Occipital region |
| 264      | Z’Graggen et al., 2006 | Male | 35 | Intracranial | [L] cerebral convexity, including the falx cerebri and superior sagittal sinus |
| 265      | Zhang et al., 2010 | Male | 27 | Intracranial | Sellar region |
| 266      | Zhang et al., 2010 | Female | 38 | Intracranial | Pituitary fossa with a suprasellar extension on the sagittal |
| 267      | Zhang et al., 2010 | Female | 26 | Intracranial | Pituitary fossa with a suprasellar extension on the sagittal |
| 268      | Zhang et al., 2010 | Male | 30 | Intracranial | Anterior cranial fossa |
| 269      | Zhang et al., 2018 | Female | 43 | Intracranial | [R] and [L] fronto-parietal and tentorium |
| 270      | Zhang et al., 2018 | Male | 16 | Intracranial | [L] temporal fossa |
| 271      | Zhu et al., 2013 | Male | 54 | Intracranial | Cerebral subdura |
| 272      | Zhu et al., 2013 | Male | 60 | Intracranial | Cerebral subdura |
| 273      | Zhu et al., 2013 | Male | 4 | Intracranial | Cerebral parenchyma |
| 274      | Zhu et al., 2013 | Female | 26 | Intracranial | Saddle area |
| 275      | Zhu et al., 2013 | Male | 38 | Intracranial | Saddle area |
| 276      | Zhu et al., 2013 | Male | 27 | Intracranial | Saddle area |
| 277      | Zhu et al., 2013 | Male | 53 | Spine | Spinal subdura |
| 278      | Zhu et al., 2012 | Male | 25 | Intracranial | [L] frontal, attaching to falx cerebri |
| 279      | Zhu et al., 2012 | Female | 38 | Intracranial | [L] temporal, attaching to dura |
| 280      | Zhu et al., 2012 | Male | 46 | Intracranial | [L] temporal, attaching to dura |
| 281      | Zhu et al., 2012 | Male | 26 | Intracranial | [L] occipital, attaching to dura |
| 282      | Zhu et al., 2012 | Male | 41 | Intracranial | [R] temporal, attaching to dura |
| 283      | Zhu et al., 2012 | Male | 40 | Intracranial | [R] paraseptal, attaching to dura |
| 284      | Zhu et al., 2012 | Male | 68 | Intracranial | [R] frontal, attaching to sagittal sinus and falx cerebri |
| 285      | Zhu et al., 2012 | Male | 35 | Intracranial | [R] occipital, attaching to [R] tentorium cerebelli |
| 286      | Zhu et al., 2012 | Male | 58 | Spine | Spinal canal, attaching to spinal meninges, T8-T10 |
| 287      | Zhu et al., 2012 | Male | 47 | Intracranial | [L] parietal, attaching to dura and falx cerebri |

R: Right, L: Left, N/A: Not available

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intensity, varying this according to the histological subtype, and on angiograms are commonly seen as hypervascular lesions,\[25\] whereas in RDD this results are variable.\[176,178\]

On RDD, in addition, on contrast-enhanced T1-weighted images with gadolinium, the lesions are markedly enhanced, homogeneously or inhomogeneously, and the dural tail sign can commonly be found.\[177,25,184,187,196,201\]

Recently, new MRI sequences have been recommended for the diagnosis of RDD, such as diffusion tensor imaging (DTI), susceptibility-weighted imaging, and perfusion-weighted imaging.\[172,177\] In addition, the use of 18F-FDG PET/CT has been described to diagnose relapsed intracranial RDD of the hypothalamus in a patient.\[199\] MRI spectroscopy menigiomas have been shown to have elevated Cho and decreased NAA, which is also seen in many other neoplastic processes, decrease in Cr and prominent Ala, much more so than in other neoplastic processes and is considered a spectroscopic signature for menigiomas.\[176\]

On the other hand, in RDD lesions, spectroscopy generally shows elevated lipid and N-acetyl aspartate peaks, suggestive of granulomatous inflammatory pathology, and a raised choline peak.\[199\] Furthermore, perfusion MRI imaging can provide
to the literature and our review. As for the location of the lesion, which can happen in many regions, including the supratentorial region, where meningiomas occur and in which one of them mimics, the two cases presented in this study are very representative, especially the second one, since at first moment it was thought that it was one of those. Furthermore, the involvement reaching the cervical portion of the second case is compatible with a higher prevalence location of spinal cord injuries according to previous studies.[101] Regarding its severity, although the involvement of CNS is often progressive and fatal, patients undergoing surgical resection have favorable prognosis in many cases.[106] However, surgical resection without complementary or additional therapy is frequently associated with recurrences of the disease[47] and should be associated with complementary or adjuvant treatments. As examples of these, chemotherapy and radiotherapy, which were very successful in the present case, may be indicated and instituted as several authors suggest,[8,9,43,55,97,124,145,153,162,179,186] especially when radical surgical resection of the tumor is not possible – which would be the best approach as several authors defend.[70,151,165] Treatment of resectable intracranial RDD and high risk by means of radiosurgery may also be a therapeutic option to be instituted,[45] obtaining very favorable results when combined with neurosurgical excision.[65,163,201] Furthermore, possibly promising, the use of Brachytherapy, a special way of applying radiation, may be a possible option to be analyzed, having already well documented therapeutic results in the treatment of other pathologies such as gliomas and some extracranial solid tumors.[11,106,143,190,223] Alternatively, the use of glucocorticoids has also shown quite beneficial effects on the regression and resolution of multiple and isolated intracranial lesions[47,126,217] and should be considered as an effective option in the treatment of RDD in certain cases where surgical resection is not applicable.

More importantly, surgical resection should follow the same pattern as meningiomas, since the texture of both is very similar, and it is extremely unlikely that with only radiological images the two pathologies can be differentiated before neurosurgical removal for anatomopathological analysis. At present, the best treatment for intracranial RDD involves surgical excision,[201] as employed in the two cases reported here.

CONCLUSION

Thus, we conclude that intracranial Rosai-Dorfman tumors should always be remembered as differential diagnosis of meningiomas, since they are similar radiologically and macroscopically. Once remembered and diagnosed, the lesion must be treated following the same pattern of section done in meningiomas and, treatment's differences will not occur in the surgical excision technique, but in complementary chemotherapy implementation, radiotherapy, and even with radiosurgery aid, depending on the case. Thus, it is possible to obtain better results than with just the isolated surgical procedure.

Declaration of patient consent

Patient’s consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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