Clinicopathological characteristics of multiple intracranial Rosai-Dorfman disease with increased IgG4-positive plasma cells: a report of two cases

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Abstract
Rosai-Dorfman disease (RDD) is an uncommon condition characterized by the proliferation of histiocytes and multiple intracranial involvements and it is extremely rare. Here, we present two cases of multiple intracranial RDD mimicking meningioma. These patients underwent surgery for tumour resection and pathological findings revealed an increased number of IgG4-positive plasma cells in RDD. The radiographic appearance and histology may contribute to a diagnostic dilemma, and immunohistochemical and serological examinations are a necessary complement for definitive diagnosis. Treatment protocols pertaining to such types of RDD cases are reviewed. Currently, surgical resection is the most effective therapy, and steroid therapy, radiotherapy, or chemotherapy may be provided as adjuvant treatments in some selected patients.

Key words: Rosai-Dorfman disease, multiple intracranial lesions, IgG4, diagnosis, treatment.

Introduction
Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare, idiopathic, and non-neoplastic histioproliferative disorder of unknown aetiology. Although it mainly affects lymph nodes and manifests with massive painless cervical lymphadenopathy in the presence of fever in young individuals, RDD has also been found to occasionally involve extranodal sites (accounting for about 40% of RDD cases), including skin, nasal cavity, bone, lung, soft tissue, and salivary glands [6]. However, intracranial presentation of this disease is extremely uncommon (approx. 5%), and as revealed by previous studies, central nervous system (CNS) involvement usually presents as an isolated lesion mimicking meningioma without systemic manifestations [2].

Recently, a few case studies have found that a subset of RDD patients exhibited increased immunoglobulin G4 (IgG4)-positive plasma cells and an elevated IgG4/IgG ratio in the affected organs [56]. These findings suggested that RDD is possibly char-
characterized by some features of IgG4-related disease (IgG4-RD). To date, few intracranial RDD cases with variable infiltration of IgG4-positive cells been reported [34,50,56,58] (Table I). Herein, we provide clinicopathological features of additional two multiple meningeal RDD cases with a highly increased level of IgG4-positive plasma cells in our institute to facilitate a comprehensive understanding of these diseases.

**Case 1**

A male patient aged 47 was admitted to our hospital with a 4-month history of intermittent right upper extremity numbness and an acute focal seizure of the right upper extremity and right face. Neurological examination was normal and no superficial lymph node enlargement or fever was found. A magnetic resonance imaging (MRI) revealed multiple carpet-like masses in the left frontal, parietal, and temporal regions and bilateral occipital meningeal. All lesions were slightly hypointense on T1-weighted images and hyperintense on T2-weighted images, with homogeneous enhancement and a dural tail sign on contrast (Fig. 1A-C). Obvious cerebral oedema was seen around the left temporal lesion, the maximum diameter of which was approximately 6 cm. Computed tomography (CT) scan demonstrated neither bone destruction nor hyperplasia. Preoperative diagnosis suggested the possibility of multiple intracranial meningiomas or metastases. Then, the patient underwent a left frontotemporal craniotomy to remove the giant mass, which was relatively avascular, yellowish, and extremely hard, with an unclear boundary between parts of the lesion and the cortex. Histopathological examination revealed cellular infiltrates of abundant lymphocytes, plasma cells, and histiocytes. On immunohistochemical staining, the histiocytes were strongly positive for S-100 protein, the monocyte-associated antigen CD68, and CD163 (Fig. 2A-C), but negative for BRAFV600E, Vimentin, EMA, CD34, GFAP, Olig-2, PR, BCL-2, and P53, and Ki67 was less than 4%. Emperipolesis was confirmed by the presence of intact intracytoplasmic CD3 and CD20 positive lymphocytes of the histiocytes. The average number of IgG4-positive plasma cells was higher than 35 by high-power field (HPF) (Fig. 2D). The ratio of IgG4+/IgG+ cells was about 50%, and IgG+ cells were positively stained by both Kappa and Lambda. The serum IgG4 level was detected normally at 21.4 mg/dl (range: 1-291 mg/dl). Based on these findings, thus the final diagnosis was RDD. Unresected small lesions located in the bilateral occipital region had not progressed during the two-year follow-up.

**Case 2**

A 19-year-old man presented with a history of headache and dysphagia for three months. He had no previous medical history. Physical examination

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**Table I. Summary of previously reported cases with intracranial Rosai-Dorfman disease mimicking IgG4-related disease**

| Authors and year | No. | Age (year) | Sex | Location(s) | IgG4 (cells/HPF) | IgG4/ IgG | Treatment(s) | Outcome and follow-up |
|------------------|-----|------------|-----|-------------|------------------|-----------|--------------|-----------------------|
| Menon et al., 2012 [34] | 1 | 55 | F | Dural involvement | 86 | 47% | NA | NA, NA |
| | 2 | 65 | F | Dural involvement | 47 | 24% | NA | NA, NA |
| | 3 | 11 | F | Frontal lobe | 10 | 5% | NA | NA, NA |
| Tauziede-Espariat et al., 2015 [50] | 4 | 35 | F | Bilateral parieto-occipital leptomeningeal involvement | > 30 | 50% | Biopsy + steroid therapy | Completely recovered, 6 months |
| Yang et al., 2017 [58] | 5 | 14 | F | Left petroclival region | 15 | NA | STR + gamma-knife surgery | Residual lesion significantly retrogressed, 12 months |
| | 6 | 50 | M | Dural involvement | 30 | 20 | Surgery + steroid therapy + rituximab | Condition improved, NA |
| Wang et al., 2020 [56] | 7 | 29 | M | Dural involvement and subcutaneous mass | > 50 | 40 | Surgery + steroid therapy + cyclophosphamide | Condition improved, NA |
| | 8 | 37 | M | Sellar region and orbital cavity | > 100 | > 40 | Surgery | Remain the same, NA |

F – female, M – male, NA – not available, STR – stereotactic radiotherapy
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found mild difficulty swallowing revealing a posterior cranial nerves deficit. MRI showed two homogenous contrast enhancement masses occupying bilateral cerebellopontine angle (CPA) regions and extending into the left foramen magnum (Fig. 1D-F). All lesions appeared isointense to the brain in T1- and T2-weighted sequences, without bone erosion or destruction in CT scan. The diagnosis of meningioma was made before surgery. As the mass effect of the left lesion on the brain stem was evident, the excision of the left lesion was performed first with intraoperative neurophysiological monitoring.

Fig. 1. Neuroimaging findings. A-C) T1-weighted contrast-enhanced MRI scan showing multiple masses in the left frontal, parietal, and temporal regions and bilateral occipital meningeal. D-F) Enhancement lesions in bilateral cerebellopontine angle regions and extending into the left foramen magnum. G-I) Progression of the lesion in the right cerebellopontine angle region.
through a far lateral approach. The lesion was red and extremely hard and grew in a creeping type with a rich blood supply. Histopathological findings included the proliferation of histiocytic cells and infiltration of lymphocytes and plasma cells (Fig. 2E). Immunohistochemical staining revealed strongly positive for S-100 and CD68 in the large histiocytes (Fig. 2F, G), and a significant portion of plasma cells was immunoreactive to IgG4 (15/HPF) and the ratio of IgG4+ to IgG+ cells was about 35% (Fig. 2H). The postoperative IgG4 serum level was normal at 36.6 mg/dl. After surgery, the patient had left facial paralysis (House-Brackmann [H-B] II degree) and a relatively mild hearing loss in his left ear, and the symptom of dysphagia also remained mild. Ionizing radiation was given to prevent further neurological impairment. According to the suggestion of a multidisciplinary team in our hospital, the patient received steroid treatment (prednisone of 30 mg qd) for one month. However, during the follow-up of the next nine months, the patient gradually developed a hearing loss in his right ear. MRI showed progression of the right lesion (Fig. 1G-I). Subsequently, he underwent excision of the lesion via the right suboccipital retrosigmoid approach. Pathological examination demonstrated typical features of RDD with increased IgG4-positive plasma cells (20/HPF) and a high ratio of IgG4/IgG (40%). Postoperative MRI
confirmed total removal of the lesion, and no recurrence occurred at the one-year follow-up. However, the patient had not completely recovered from his bilateral hearing loss, right facial paralysis (H-B III degree), and dysphagia.

Discussion

Rosai-Dorfman disease is an uncommon disorder that rarely involves CNS, moreover, multiple intracranial lesions have also been seldom reported. As indicated in previous studies, this specific type of RDD has a propensity to occur in males (male : female 3.3 : 1) and the age of patients at diagnosis ranges from 2 to 72 years [1,4,7,8,10,13,16,20,25-27,31-33,35,38-41,44,48,51,53,55] (Table II). Presenting symptoms associated with neurological deficits and mass effects depend on the location, size, and growth pattern of lesions. The common sites of multiple RDD are reported in cerebral convexity, parasellar and suprasellar regions, cranial base, and supratentorial ventricular system. In our study, we presented a case with lesions involving cerebral convexity and a patient with masses in bilateral CPA regions.

Radiologically, the typical appearance of multiple RDD is dural-based, extra-axial, and well-circumscribed enhancing masses mimicking meningioma in the cranium. On CT scans, the lesions show homogeneous hyperdensity or isodensity with possible hypodense perilesional cerebral oedema. Hyperostosis, bone erosion, and calcification are usually absent but can be common in meningioma. Moreover, digital subtraction angiography shows hyper-vascularity in a majority of meningiomas which are largely fed by branches of the external carotid artery or the internal carotid artery system [52]. In contrast, RDD varies in blood supply from avascularity to abundant vascularity [29]. MRI plays an important role in RDD diagnosis. When viewed on T1-weighted images, they usually appear as isointense or hyperintense masses with clear borders relative to the brain parenchyma, while T2-weighted and FLAIR sequences show an isointense signal with possible intralesional foci of low signal intensity, which may be helpful to distinguish RDD from meningioma. It is presumed that the specific hypointense foci are formed as a result of free radicals produced by

Table II. Published cases with multiple intracranial Rosai-Dorfman disease in the last 30 years

| Authors and year | No. | Age (year) | Sex | Locations | Treatment(s) | Outcome and follow-up |
|------------------|-----|------------|-----|-----------|--------------|-----------------------|
| Kim et al., 1995 [25] | 1 | 50 | M | Lesions involving the right parietal convexity | Complete removal | No residual or recurrent lesion, 6 months |
| Resnick et al., 1996 [41] | 2 | 38 | M | Lesions involving the frontal convexity in both sides and the left CPA region | Partial resection of the posterior fossa mass | Stable, 18 months |
| Udono et al., 1999 [53] | 3 | 67 | F | A large lesion in the right frontal parasagittal area and several satellite lesions involving the right parietal convexity, frontal base, right sphenoid ridge, left frontal convexity, tentorium, and pharynx | Removal of the frontal convexity mass | No regrowth of the satellite lesions, 20 months |
| Chen, 2003 [10] | 4 | 70 | M | Lesions involving the right parietal and suprasellar regions | Resection of the right parietal mass and biopsy for the suprasellar lesion | Died of respiratory failure, 3 years after the second craniotomy |
| Purav et al., 2005 [38] | 5 | 50 | M | Multiple intraparenchymal lesions | Biopsy for the right parietal lesion | Died, 10 days post-operation |
| Gupta et al., 2006 [16] | 6 | 15 | M | Bilateral petroclival lesions extending to the cavernous sinus region bilaterally and a lesion in the bilateral anterior frontal parasagittal region | Steroid therapy + near-total resection of the lesion in the left side and gross total resection of the lesion in the right side | A small residual tumor (stable), 12 months |
| McPherson et al., 2006 [33] | 7 | 53 | M | Multiple skull base lesions including a lesion at the planum sphenoidale and tuberculum sellae and bilateral lesions from the CPA to the foramen magnum | Resection of the planum sphenoidale/tuberculum sellae lesion debulked maximally + steroid therapy | Marked resolution of the remaining lesions, 11 months |
| Authors and year                        | No. | Age (year) | Sex | Locations                                                                 | Treatment(s)                                                                                           | Outcome and follow-up                              |
|----------------------------------------|-----|------------|-----|---------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------|-----------------------------------------------------|
| Russo et al., 2009 [43]                | 8   | 72         | M   | Bilateral frontobasal lesions                                             | Resection of the two masses                                                                             | Free of neurological symptoms, 14 months            |
| La Barge 3rd et al., 2009 [27]         | 9   | 51         | M   | Lesions in bilateral CPA regions extending to the cervicomедullary junction | NA                                                                                                      | NA, NA                                              |
| Symms et al., 2010 [48]                | 10  | 21         | M   | Lesions involving the tentorium inferotemporally on both sides and the cavernous sinuses | Subtotal resection of the right lesion                                                                | Progression of the lesion bilaterally, 6 months     |
| Raslan et al., 2011 [40]               | 11  | 50         | M   | Lesions involving bilateral paracavernous regions, right temporal lobe, left frontal convexity, and spinal canal at L1 | Resection of brain and spinal lesions                                                                  | NA, NA                                              |
|                                        | 12  | 54         | M   | Sellar and suprasellar lesions, bilateral CPA lesions extending into the upper spinal canal | Surgical resection of the sellar lesion + steroids therapy                                            | NA, NA                                              |
| Nalini et al., 2012 [35]               | 13  | 35         | M   | Lesions involving parasellar, tuberculum sellae, planum sphenoidale, and tentorium, and inferiorly along the CPA region and clivus into the spinal canal | Steroid therapy + biopsy + radiotherapy                                                                | Disease progression, NA                              |
| Catalucci et al., 2012 [8]             | 14  | 57         | M   | Lesions involving the cerebral falx within the interhemispheric fissure, the right periventricular region, the right temporo-polar/sphenoidal and insular region, the left tentorium, the right petroclival region, and the lateral ventricle | Surgical resection of the infratentorial petroclival right mass                                         | NA, NA                                              |
| Camp et al., 2012 [7]                  | 15  | 31         | F   | Lesions involving the left frontal region, the right frontal white matter, and the right parietal region | Resection of the left frontal lesion + steroid therapy                                                | No evidence of disease recurrence, 12 months        |
| Antuna Ramos et al., 2012 [4]          | 16  | 10         | F   | Intracranial lesions involving the right parasagittal basal frontal region, the left middle cerebellar peduncle, and the right ventral pons Intraspinal lesions at T9-T10, T6-T7 and T10-T11 levels | Complete removal of three frontal brain lesions, partial resection of the intraspinal lesion and the cerebellopontine lesion + steroid therapy + radiotherapy | Died, 5 months after the initial diagnosis          |
| Abdel-Razek et al., 2013 [1]           | 17  | 43         | M   | Lesions involving the right frontal convexity, the left frontal region, cerebral falx, the anterior clinoind process, and the right petrous bone | Total resection of the large frontal lesion                                                            | NA, NA                                              |
| Forest et al., 2013 [13]               | 18  | 38         | M   | Ethmoidal, frontal, and cerebral falx lesions                             | Resection                                                                                              | NA, 8 years                                         |
| Sandoval-Sus et al., 2014 [44]         | 19  | 51         | M   | Lesions involving bilateral internal auditory canals and CPA regions and the foramen magnum | Partial surgical resection + radiotherapy                                                               | Stable, 7 months                                    |
|                                        | 20  | 18         | M   | Multiple brain, base of skull, and cervical spine masses                  | Partial surgical resection followed + chemotherapy (V, MTX, 6-MP and prednisone) + radiotherapy          | Stable, 7 years                                     |
| Lüdemann et al., 2015 [31]             | 21  | 2          | M   | Lesions involving bilateral frontal lobes and the occipital horn of the left ventricle | Steroid therapy + partial resection of the occipital intraventricular mass                             | Stable, 16 months                                   |
| Tian et al., 2015 [51]                 | 22  | 6          | F   | Lesions in the lateral ventricle, tentorium of cerebellum on both sides and in frontal falx | Total resection of the largest mass in the left ventricle                                              | Stable, NA                                          |
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Macrophages during active phagocytosis [53]. Contrast enhancement is usually marked and presents as inhomogeneous or homogeneous after contrast administration, but can also be little or absent in some cases [18]. The dural tail sign is demonstrated in most cases, however, few purely intraparenchymal RDD cases without dural attachment do not have the classical characteristic, which further increases the difficulty of differential diagnosis [5,14,21]. Newer types of MRI sequences can be used for a better preoperative evaluation as recommended by some authors. On apparent diffusion coefficient maps, RDD displays a higher signal value than lymphoma but lower than meningioma [9]. Magnetic resonance spectroscopy combined with the perfusion profile has particular specificity for some patients, which suggests RDD as an inflammatory rather than a neoplastic process [7,48]. Differentials from imaging diagnosis should consider meningioma, lymphoma, metastases, Wegener's granulomatosis, sarcoidosis, Langerhans cell histiocytosis, and pseudotumour.

Definite diagnosis of RDD is totally dependent on histopathological and immunohistochemical examinations. Microscopically, lymphoid follicles with germinal centres, fibrosis, sclerosis, and fewer histiocytes is a prominent feature that is consistent in extra-nodal RDD [21]. Another typical finding is seen as phagocytic vacuoles of intact and viable lymphocytes within the cytoplasm of histiocytes, but this phenomenon of emperipolesis is present in only 70% of cases and is less often found in CNS RDD than nodal RDD [38]. The staining of histiocytes is positive for CD68 and S-100, but negative for CD1a and EMA, which has important diagnostic values and help rule out other histiocytic diseases and meningioma. Immunoreactivities of CD3 and CD20 indicate mixed populations of B and T lymphocytes in the background. Positive staining of Kappa and Lambda

| Authors and year | No. | Age (year) | Sex | Locations | Treatment(s) | Outcome and follow-up |
|-----------------|-----|------------|-----|-----------|--------------|-----------------------|
| Luo et al., 2017 [32] | 23 | 41 | M | Lesions involving ventricles, left parasellar and CPA region | Complete resection of the fourth ventricle lesion + steroid therapy + radiotherapy | Residual lesions markedly enlarged, 42 months |
| | 24 | 31 | M | Lesions involving bilateral sphenoidal crest, right parasellar, cavernous sinus, bilateral frontoparietal meningeal and foramen magnum | Resection of the right parasellar mass + steroid therapy + radiotherapy | Residual lesions slightly enlarged, 24 months |
| Jiang and Jiang, 2018 [20] | 25 | 39 | M | Lesions involving the left frontal lobe and right frontoparietal meningeal regions | Radiotherapy + total resection of lesions in two-stage surgery | No recurrence, 78 months |
| | 26 | 53 | M | Lesions involving the left parietal, temporal, occipital meningeal regions | Resection of the largest left occipital mass | No size increases of residual lesions, 8 months |
| | 27 | 9 | F | Lesions involving the right parietal meningeal region | Total resection | No recurrence, 42 months |
| Krueger et al., 2019 [26] | 28 | 52 | F | Lesions involving the bilateral paramidline frontal and right parietal lobes at the vertex, left frontal lobe near the vertex, bilateral posterior parietal lobes, and bilateral frontal-temporal lobes | Gross total resection of the largest lesion in the right frontal-temporal region | Stable, 3 months |
| Qin et al., 2019 [39] | 29 | 43 | M | Lesions involving frontal falx, parietal falx and tentorium cerebelli, and intraspinal dural T3 level | Resection of the intraspinal lesion + steroid therapy | Stable, 2 years |
| Wang et al., 2019 [55] | 30 | 57 | F | Lesions involving bilateral petroclival, parasellar regions | Subtotal resection of the lesion in the left base of anterior cranial fossa | Alive with disease, 18 months |

CPA – cerebellopontine angle, F – female, L – lumbar, M – male, NA – not available, T – thoracic
light chain shows polytypic staining in the plasma cell infiltrates, which excludes the diagnosis of plasmacytoma with monoclonal plasma cell infiltrates.

However, the presence of a high proportion of IgG4+ plasma cells in RDD requires the exclusion of IgG4-RD in the diagnostic process. IgG4-RD is a multi-organ immune-mediated fibroinflammatory condition characterized by specific clinical, serological, and pathological features involving tumefactive tissue, elevated serum IgG4 level, lymphoplasmacytic infiltration enriched in IgG4+ plasma cells, storiform fibrosis, and obliterator phlebitis [22,23]. Though the enrichment of IgG4+ plasma cells and increased IgG4/IgG ratio in RDD mimics some characteristics of IgG4-RD, the former lacks storiform fibrosis and obliterator phlebitis. Elevated serum IgG4 concentration has also been found in some RDD cases, but in general, the level is still lower than that in IgG4-RD patients [30,56]. Moreover, negatively stained S-100 and CD68 and higher number of Foxp3+ regulatory T cell infiltration in the IgG4-RD group, while neoplastic point mutations affecting KRAS, NRAS, SMAD4, ARAF, or MAP2K1 occurring in 33-50% of RDD cases but not IgG4-RD, provide evidence that the two entities are apparently different from each other [12,15,30,46].

Sporadic RDD frequently presents a self-limited clinical course, with up to 50% of cases having spontaneous remission without any therapy [6]. However, spontaneous resolution is not observed in RDD patients with CNS involvement [44]. Consensus on the management of these RDDs should be reached to confer benefit in patients. Currently, long-term satisfactory results can be obtained from surgical intervention, which is aimed at gross total excision (GTR), in a majority of isolated intracranial RDD cases [13], but recurrence still occurs in about 14% of patients receiving initial GTR alone over a mean of 10.1 years [2]. In cases of disease located in relatively less accessible regions such as the petroclival or clival area or with multiple lesions, subtotal excision or biopsy or single resection of single foci will be performed sometimes, which significantly increases risks of RDD growth and progression, then close follow-up with clinical and radiological observation is strongly recommended and adjuvant treatments can also be considered.

Systemic steroids are the preferred treatment advocated by few investigators for residual or refractory intracranial or even non-surgically RDD lesions. However, only a portion of cases may benefit from this therapeutic regimen, moreover, in the subgroup of RDD mimicking IgG4-RD, response to steroids is not as good as that in IgG4-RD [11,33,47,56,59]. Once the steroid therapy fails, most patients need treatment of high intensity, such as combining with radiotherapy or chemotherapy or using immunosuppressive drugs [45,57]. Radiation offered as fractionated therapy or stereotactic radiotherapy has been used in a few cases. However, no standard doses have yet been established, and the response to radiotherapy varies in patients from poor control to complete disappearance of the lesion [11,17,24,37,44]. Many chemotherapy agents including cytarabine, methotrexate, 6-mercaptopurine, anthracyclines, vinca alkaloids, 2-chlorodeoxyadenosine, and etoposide have been tried for RDD with mixed results [11,19,42,49,57]. In general, chemotherapy has not provided significant efficacy in the long-term remission of RDD lesions. Additionally, immuno-modulators like interferon- targetted therapies, such as Imatinib and Rituximab, have also been introduced in the management of RDD, but sufficient evidence is currently lacking to support intracranial disease control by these treatments [3,28,36,54].

Conclusions

Multiple intracranial RDD mimicking IgG4-RD is an extremely rare disease. Our study underlines the pivotal roles of histopathological and immunohistochemical examinations in its definitive diagnosis and provides comprehensive understanding through clinical manifestations, laboratory parameters, imaging findings, and other pathological characteristics. Maximum safe surgical removal is the preferred treatment approach for almost all patients, as spontaneous regression has not been observed in cases of CNS involvement. Steroids, radiation therapy, and chemotherapy reserve treatment options for residual or refractory RDD lesions, but their variable efficacies should be optimized in future investigations.

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Disclosure

The authors report no conflict of interest.
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