Intracranial Rosai Dorfman Disease: report of three cases and literature review.

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Background: Rosai-Dorfman Disease (RDD) is a rare idiopathic non-neoplastic histioproliferative disease characterized clinically by massive painless cervical lymphadenopathy, fever and weight loss. Extranodal involvement has also been recognized. Central nervous system (CNS) manifestations are extremely rare and patients with intracranial involvement usually present with clinical and radiological findings suggestive of a meningioma.

Case description: We report our experience in the management of three patients with RDD. Two patients had dural based lesions, radiologically in favour of a meningioma, and one patient had a parenchymal lesion suggestive of a tuberculous granuloma. Treatment consisted of total excision in one case, and subtotal excision followed by conventional radiotherapy in two cases. The diagnosis was confirmed by histopathology and immunochemistry which is essential for a definite diagnosis of RDD.

Keywords: Rosai-Dorfman Disease, Meningioma, Histoproliferative disease, Lymphnode, Tuberculous granuloma.
were normal and there was no lymphadenopathy. His immune status was normal too. His MRI scan of the brain (plain and contrast study) showed extensive infratentorial lesions extending on both sides along the tentorium, upto the cavernous sinuses, with a larger component on the right side. On the right side, lesion was exerting pressure on the pons, with effacement of the cisterns, and deformity of the fourth ventricle. The lesion was isointense on T1 WI, hypointense on T2 WI and uniformly enhancing with contrast (fig 1a). Based on these imaging findings a provisional diagnosis of an en-plaque meningioma was made. The patient underwent a right retromastoid craniectomy and subtotal excision of the lesion. The lesion was tough, fibrous in consistency, avascular, was extending along the tentorium, and had breached the arachnoid. Only subtotal excision of the lesion could be achieved as it was too extensive. The portion causing pressure on the brainstem was excised. Post operative period was uneventful. The histopathology report was suggestive of a non-specific diffuse pachymeningitis, possibly of tuberculous origin. The patient was started on a course of ATT (Rifampicin, Isoniazid, Pyrazinamide). He was under regular follow-up and remained asymptomatic. He presented six months later with recurrence of symptoms of headache, vomiting and giddiness. CT scan of the brain plain and contrast study showed progression of the lesion bilaterally (fig 1b). The histopathological specimen was reviewed by the same pathologist at the same centre and immunohistochemical analysis demonstrated that most of the histiocytes stained diffusely positively for S100 protein and CD 68. Hence the lesion was diagnosed as intracranial RDD. Since the lesion was too extensive to surgically excise, he underwent low dose (20Gy) conventional radiotherapy to the brain. At eight years follow up the patient continues to be symptomatic and repeat imaging (fig 1c) shows the lesion to be progressing in size, extending into the cavernous sinuses upto the orbit.

**Case 2**

A 35 years male, presented with complaints of three episodes of generalized tonic clonic

![Figure 1a. MRI scan of the brain plain and contrast showing lesion which is isointense in T1 WI, hypointense in T2 WI and uniformly enhancing with contrast. Lesion seen infratentorially extending on both sides along the tentorium, upto the cavernous sinuses, with a larger component on the right side. There is pressure on the right side of the pons, with effacement of the cisterns, and deformity of the fourth ventricle.](image-url)
seizures over a period of one year. He had no complaints suggestive of raised intracranial pressure. There was no history of fever, chills or rigors, and no significant family history. He was radiologically investigated at another neurological centre, found to have a bifrontal dural lesion and was started on anti-tubercular treatment (ATT), which he took for 4 months. On examination his neurological functions were normal. His other systems were normal, there was no lymphadenopathy, and his immune status was normal also. MRI of the brain (fig 2a) with gadolinium (fig 2b) showed a bifrontal dural based lesion with involvement of the falx. It was intensely enhancing with contrast, with perilesional oedema and mass effect. He underwent a bifrontal craniotomy and total excision of the lesion. The involved dura, falx and anterior end of the superior sagittal sinus were also excised. The tumour was well-circumscribed, extra-axial, relatively avascular and tough in consistency. There was no clear arachnoid plane between the lesion and the underlying cortex. Histology: showed dural collagen densely infiltrated by sheets of mature plasma cells. These were admixed with a smaller population of mature lymphocytes and sheets of large histiocytes. These cells were positive for CD 68 and S-100 protein. Russell bodies were also prominent. His post operative CT scan of the brain showed total excision of the lesion (fig 2c). After three years of follow up, the patient remains asymptomatic, with no clinical evidence of recurrence.
A 17 years old female, presented with complaints of headache and episodes of vomiting for 16 months duration. She also had amenorrhea, excessive weight gain, and memory lapses for one year. She was initially evaluated at another hospital and her MRI scan of the brain was done in December 2006, which showed an ill-defined contrast enhancing lesion in the suprasellar region (fig 3a). Her cerebrospinal fluid analysis revealed elevated protein and cell counts, which was suggestive of tuberculous infection. Based on this she was started on antituberculous treatment (Isoniazide, Rifampicin, Pyrazinamide). Eight months later she presented to us with complaints of insidious onset, painless, progressive loss of vision in
both eyes for last 5 months, with persistent headache and vomiting. On examination her higher intellectual functions were normal, fundus showed bilateral primary optic atrophy, visual acuity in the right eye was 3/60, and left eye 6/60, and she had bitemporal hemianopia. She also had a right third and sixth cranial nerve paresis. Her other cranial nerves were normal, and she had no spinomotor sensory deficits. In August 2007, her repeat contrast MRI scan of the brain (fig 3b) showed multiple ring enhancing lesions in the sellar, suprasellar, retrosellar and interpeduncular regions, with an intra-axial lesion in the left medial temporal region, with surrounding vasogenic oedema and mass effect. She was continued on antituberculous treatment and was on regular follow up with no further deterioration in her vision.

She was re-admitted in March 2008 with complaints of rapid visual deterioration, and increased thirst for last one week. On examination her fundus showed bilateral optic atrophy. Visual acuity in the right eye was 6/60 and in the left eye, she could perceive only hand movements. The right third and sixth cranial nerve paresis persisted. The rest of her neurological examination was normal. Her other systems were normal, there was no lymphadenopathy or immune deficiency. Her repeat MRI scan of the brain plain and gadolinium, (fig 3c) showed an increase in the size of the lesions. Magnetic resonance spectroscopy showed elevated lipid and N-Acetyl Aspartate peaks suggestive of a granulomatous lesion (fig 3d). Her endocrine analysis by radio-immunoassay was as follows: T3-124 ng/dl; T4- 9.00 ug/dl; Thyroid stimulating hormone- 2.00 Ui/ml; Growth hormone 0.29
Follicular stimulating hormone-1.00 mIU/ml; Leutinizing hormone-1.00 mIU/ml; Prolactin 13.00 ng/ml; Cortisol- 1.00ug/dl. Alpha feto protein was 1.7 ng/ml, beta human chorionic gonadotropin was 2.2 mIU/ml.

As the lesion had increased in size following one year of antituberculous treatment and since the patient was rapidly losing vision bilaterally, she underwent a right frontal craniotomy and subtotal excision of the lesion. Per-operatively, the lesion had splayed both optic nerves, the chiasm was stretched, there were arachnoidal adhesions, and the lesion was densely adherent to the hypothalamus. Due to this, only subtotal excision of the lesion could be achieved. The lesion was tough in consistency, moderately vascular with central areas of necrosis. The histopathological examination showed chronic inflammatory granulation tissue, admixed with multinucleated giant cells, revealing emperipolesis. Immunostaining with antibodies against growth hormone and prolactin did not reveal remnants of adenohypophysis in the resected tissue. S-100 revealed cytoplasmic labelling of the histiocytes and only an occasional one showed nuclear labeling. These findings were consistent with RDD. Her post operative CT scan showed multiple ill defined hyperdense, contrast enhancing residual lesions in the sellar, suprasellar, retrosellar, interpeduncular , left basifrontal and left temporal lobe regions with perilesional oedema and mass effect on the lateral ventricles, with post operative changes (fig 3e). The patient underwent low dose conventional radiotherapy (20 Gy) for the residual lesion, which led to some resolution of the intracranial lesion, and partial recovery of her vision. On one year follow up, she developed hydrocephalus and required a ventriculoperitoneal shunt. On two years follow up, the patient is symptom free with further improvement in her vision, and the contrast CT shows further resolution of the lesion (fig 3f).
RDD is a well described entity, the etiology of which is still unknown. It often occurs in the setting of nonspecific immune dysfunction with many cases occurring after a viral illness (21). Levine et al recommended that the human herpes virus 6 and to a lesser extent Epstein Barr virus may be involved in the aetiology (13). The mean age of onset with nodal disease is 20.6 years with a male to female ratio of 1.4:1 (6,15). Patients who develop intracranial involvement, however, become symptomatic at a mean age of 34.9 years, with a strong male preponderance. Ninety percent of the patients present with massive painless bilateral cervical lymphadenopathy (21). Fever, anaemia, leukocytosis, elevated erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia may also be found. One third of the patients

Discussion

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have extranodal involvement, such as the eyes, skin, upper respiratory tract, bone, salivary gland, testis and in extremely rare circumstances, the central nervous system can be affected (6,22). Massive cervical lymphadenopathy is not always seen with intracranial RDD (21). To the best of our knowledge 49 previous case reports of isolated intracranial RDD have been published in major scientific journals and their details are available in table 1. In all our cases the lesion was confined to the central nervous system, and there was no evidence of lymphadenopathy. Occasionally, only extranodal disease has been found, and for these patients, the lack of lymphadenopathy makes the term sinus histiocytosis with massive lymphadenopathy inappropriate. Foucar et al have described in their patients with sinus histiocytosis with massive lymphadenopathy, a poor outcome when associated immunocompromised state exists. (6) But does this fact hold good for isolated intracranial RDD, is still unclear. None of our patients had diseases predisposing for immunocompromised states, and they were not on immunosuppressive medications. HIV-ELISA was done to rule out acquired immunodeficiency syndrome(AIDS). A normal peripheral blood smear with a complete blood count within normal limits ruled out common lymphoreticular malignancies. Also none of them had a clinical picture of primary immunodeficiency diseases like immunoglobulin deficiency. Hence, further tests to identify them were not attempted. In patients suspected to have primary immunodeficiency disease, quantitative assays of IgG, IgM, IgA, IgE and complement factors can be done.

Intracranial RDD usually presents as a solitary dural-based lesion though multiple intracranial lesions have also been reported. (3,11). Purely intraparenchymal (I,5,10), and intraventricular lesions (17) have also been reported. They can occur in the suprasellar region, convexity, parasagittal region, cavernous sinus, petroclival region and cerebellum. Cheng et al have reported two cases of RDD with locally aggressive features and dural sinus invasion (2). RDD can also present as pachymeningitis (12). Patients with intracranial involvement usually present with headache and seizures. They can also present with dysphasia, cranial nerve deficit, progressive loss of vision, hemiparesis, neglect, and endocrine dysfunction depending on the location of the lesion. Suprasellar involvement has been reported in only five other cases (table-1), and hypothalamic-pituitary axis dysfunction has been described in only one case, in which diabetes insipidus was present (18). CT scan findings typically shows a hyperattenuating, contrast enhancing meningeal-based mass with a variable amount of oedema surrounding the lesion, mimicking a meningioma. On MRI scans the lesion is usually isointense to gray matter on T1-weighted images and hyperintense on T2-weighted. Homogenous enhancement is typical.

### Table 1. Summary of reported literature on isolated intracranial Rosai-Dorfman Disease.

| Source          | Year | Cases | Age/Sex | Location            | Treatment          | Outcome         |
|-----------------|------|-------|---------|---------------------|--------------------|-----------------|
| Foucar et al    | 1982 | one   | 21/M    | CP angle            | Total excision     | N/A             |
| Tradel          | 1984 | one   | 28/M    | Petrous bone       | 90% excised        | 14 months       |
|                 |      |       |         |                     | Irradiated         | No regrowth     |
| Lopez et al     | 1989 | one   | 35/M    | Cavernous sinus/CPangle | Subtotal excision | 1 year, no progression |
| Bhattacharjee   | 1992 | one   | 78/M    | Suprasellar        | Subtotal excision  | 1 year, no progression |
| Shaver et al    | 1993 | one   | 5/M     | Cavernous sinus/postfossa | Subtotal excision | 1 month, no Progression |
| Kim et al       | 1995 | one   | 50/M    | Parietal convexity  | Total excision     | 6 months, no progression |
| Ng et al        | 1995 | one   | 22/M    | Suprasellar        | Aspiration         | N/A             |
| Clark et al     | 1996 | one   | 38/F    | Parietal mass      | Surgery            | 4 years, no Progression |
| Kitit et al     | 1996 | one   | 25/M    | Occipital          | N/A                | N/A             |
| Panicker et al  | 1996 | one   | N/A     | N/A                | N/A                | N/A             |
| Deodhar         | 1998 | two   | 41/M    | Parietooccipital   | Subtotal excision  | 4 years, no Progression |
|                 |      |       | 38/M    | Parietooccipital   | Total excision     | 6 months, no Progression |
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| Source            | Year | Cases | Age/Sex | Location             | Treatment                  | Outcome                  |
|-------------------|------|-------|---------|----------------------|----------------------------|--------------------------|
| Simos et al       | 1998 | one   | 63/M    | Parasagittal         | Total excision             | 2 years, no progression |
| Huang et al       | 1998 | one   | 58/M    | N/A                  | N/A                        | N/A                      |
| Kattner et al     | 1999 | one   | 33/M    | Parasagittal         | Total excision             | 1 year, no progression  |
| Woodcock et al    | 1999 | one   | 15/F    | Suprasellar          | Biopsy, steroids           | 9 months, Progressed.   |
| Udono et al       | 1999 | one   | 67/F    | Frontal              | Excision                   | 20 months, no progression|
| Natarajan et      | 2000 | one   | 45/F    | Frontal intraparenchymal | Excision.                  | N/A                      |
| Gaetani et al     | 2000 | one   | 67/F    | Cerebellar           | Total excision             | N/A                      |
| Morandi X et al   | 2000 | one   | 22/F    | Fourth ventricle     | Total excision             | 5 years, no progression |
| Petzold et al     | 2001 | one   | 78/M    | Suprasellar          | ST excision, irraditated    | 11 years, progressed    |
|                   |      |       |         |                      |                             | 47/M                     |
|                   |      |       |         |                      |                             | Multiplesions             |
|                   |      |       |         |                      | ST excision, irraditated    | 2 years, progressed      |
| Andriko et al     | 2001 | one   | 61/F    | Dural-based parenchymal | Biopsy/Excision           | Mean: 15 months, no progression |
| Hadjipanayis et al| 2003 | one   | 52/M    | Petrocival and cavernous sinus | Subtotal excision/SRS | Regression               |
| Juric et al       | 2003 | one   | 39/M    | Temporal             | Total excision             | N/A                      |
| Sato et al        | 2003 | one   | 59/F    | Multiple             | Subtotal/ SRS              | 2 years, no progression |
| Gao Jin et al     | 2004 | one   | 58/M    | Frontal              | Total excision             | N/A                      |
| Kinoshita et      | 2004 | one   | 69/M    | Frontal              | Total excision             | N/A                      |
| Grillo et al      | 2004 | one   | 9/M     | Frontal              | N/A                        | N/A                      |
| Kayali et         | 2004 | one   | 31/M    | Temporal             | Total excision             | N/A                      |
| Jin Kyu Park et   | 2004 | one   | N/A     | Foramen magnum       | Excision                   | N/A                      |
| Cheng Hong Toh    | 2005 | two   | 60/M    | Occipital lobe sinus | ST Excision                | 4 months, no progression|
|                   |      |       | 59/M    | Frontal lobe invasion | ST Excision                | 30 months, no progression|
| Werner at al      | 2005 | one   | 35/M    | Left convexity       | Biopsy/steroids            | N/A                      |
| Sundaram et       | 2005 | Three | Above 35 years | N/A                  | N/A                        | 10 to 21.5 years, no progression |
| Sharma et         | 2005 | one   | 40/M    | Sphenoid wing        | Near total excision        | 3 months, no progression |
| Pavav et al       | 2005 | nine  | N/A     | Intracranial         | Surgical excision to biopsy| 3 months to 8 years, no progression |
| Sang Ha Shin et   | 2006 | one   | 61/M    | Posterior fossa      | Total excision             | 5 months, no progression |
| McPherson et      | 2006 | one   | 53/M    | Multiple skull base lesions | Excision/ steroids | Regression all lesions |
| Gupta et al       | 2006 | one   | 15/M    | Petrocival bilateral | Near total excision        | N/A                      |
| Z Gragggen et     | 2006 | one   | 35/M    | Dural cerebral hemisphere | Biopsy/steroids | 2 months, remission of symptoms |
| Shang Chi et      | 2007 | two   | 57/M    | Left CP angle        | Excision                   | No recurrence            |
|                   |      |       | 60/M    | Left frontal parafalcine | Excision                   | No recurrence            |
| Xiaoping et       | 2007 | one   | 17/F    | N/A                  | N/A                        | N/A                      |
| Di Rosco et       | 2007 | one   | 13/F    | Frontal              | Biopsy/Excision            | 1 year, no progression  |
| Seyward K et      | 2007 | one   | 45/M    | Parietal             | Total excision             | N/A                      |
| Ghosal N et       | 2007 | one   | 26/M    | Parietal convexity   | Excision                   | N/A                      |
| Mitetic et        | 2008 | one   | 8/NA    | Nucleus Lentiiformis | STB                        | N/A                      |
| Raj Kumar et      | 2008 | one   | 36/M    | Frontal convexity    | Total excision             | 4 months, no progression |
| Wan et al         | 2008 | one   | 43/M    | Suprasellar          | N/A                        | N/A                      |
| Peev NA et        | 2008 | one   | 47/F    | Cerebellar           | Total excision             | 3 years, no progression |
| Mingyi et         | 2008 | one   | 71/M    | Bifrontal            | Excision                   | N/A                      |
| Bing F et         | 2009 | one   | 32/F    | Parieto-occipital( intraparenchymal) | Biopsy/steroids | 5 years, resolution. |

**Abbreviations for the table**
- M: Male  F: Female  N/A: Not available  CP: Cerebello Pontine  STB: Stereotatic Biopsy  ST: Subtotal  SRS: Stereotactic radiosurgery
and the lesions are angiographically avascular. On MR spectroscopy the lesion may mimic a granulomatous lesion, as seen in case-3. Udono et al have reported these lesions to have low signal intensity on MRI, T2-weighted imaging (27). Sze and Zimmerman suggested that the low signal intensity on T2-weighted MR imaging might reflect the presence of free radicals produced by macrophages during active phagocytosis (26). The differential diagnosis includes lesions such as meningioma, histiocytosis X, Wegener’s granulomatosis, sarcoidosis, tuberculomas, gliosarcomas, Hodgkin’s disease, plasma cell granulomas, and in suprasellar lesions also includes germinoma, hemophagocytic lymphohistiocytosis, and metastasis. Radiologically and histologically they can mimic granulomatous lesions, as seen in all our three cases where they have received ATT at some point of time. Thus histological and immunohistochemical confirmation is essential for a definitive diagnosis of RDD. On microscopic examination a polymorphous infiltrate of histiocytes, lymphocytes and plasma cells in a fibrous stroma can be seen. In RDD two subsets of histiocytes, differentiated by size, are present (3,14). The large histiocytes typically exhibit emperipolesis, which is an active penetration of one cell by another (9). In RDD the large histiocytes contain well preserved lymphocytes and are usually S-100 positive (14). The medium-sized histiocytes may not exhibit emperipolesis, are representative of a histiocyte at an earlier stage, and are typically S-100 negative.

In RDD with intracranial involvement, the treatment of choice is surgery. Complete resection, subtotal resection, and biopsy of the lesion have been attempted. One should aim for complete resection of the lesion as leaving residual tumour has lead to recurrence of the disease (19). From our analyses of the 49 previous case reports (table 1), we found where the lesion has been totally excised from areas such as the CP angle, convexity dura, cerebellum, parasagittal, and sphenoid wing, on follow up there was no recurrence or progression of the disease. In areas like the cavernous sinus, petroclival, suprasellar region, or where there are multiple lesions and only subtotal excision could be achieved, there are reports of both, progression and no progression of the disease on follow up. However, where the lesion was only biopsied, there was definite progression of the disease, even after adjuvant therapy.

Adjuvant therapy has included irradiation, chemotherapy and steroids. Petzold et al found intracranial tumour recurrence with symptoms in 14% of 29 patients with a mean follow-up of 10.1 years. They concluded that a five year follow-up with brain imaging was essential and advocated local low-dose radiation to treat patients with subtotal resections (19). Gamma knife radiosurgery to the residual lesion, and for the other intracranial lesions has been advocated (7,24). Horneff et al, have advocated the combined use of low-dose methotrexate and 6-mercaptopurine therapy for four months to ensure remission followed by 6-mercaptopurine for a total of two years (8). McPherson et al, administered corticosteroid agents following surgery, leading to a marked resolution of both the remaining surgically untreated lesions and the balance of the patient’s symptoms. This report represents the first case of the resolution of intracranial RDD following corticosteroid therapy (16). Similarly Bing et al also found resolution of the lesion on five years follow up with treatment with steroids (5). Patients may also develop hydrocephalus at a later date, which may require a CSF diversion. Sang et al have reported a patient with posterior fossa RDD who developed hydrocephalus 5 months after craniectomy and excision of the lesion (23). The prognosis is variable. In patients with non-intracranial RDD, the clinical course tends to be protracted and unaltered by treatment, and the disease will resolve spontaneously. Some have complete resolution of the adenopathy whereas, in others, the condition persists but is asymptomatic (15). RDD with involvement of the CNS appears to have a benign prognosis. However in one study, operative mortality has been reported to be approximately 4% (19)

**Conclusion**

Rosai-Dorfman disease is a well recognized clinicopathological entity. The disease may be nodal or extranodal. It may present with only
extranodal involvement of the skin, orbit, upper respiratory tract, testes, or rarely CNS. Rosai-Dorfman disease should be considered in the differential diagnoses of both dural-based and intraparenchymal lesions of the CNS, where dural based lesions mimic the radiological appearance of a meningioma, and intraparenchymal lesions may mimic granulomatous lesions. Thus, a definite diagnosis relies on the histological pattern and immunohistochemical characterization of the lesions. Surgical excision of the lesion is the treatment of choice. In cases with subtotal tumour resection or recurrence of the lesion, adjuvant therapy with local low dose radiotherapy and steroids can be considered. Prognosis is benign especially in the absence of nodal disease.

**Abbreviations**

RDD Rosai-Dorfman Disease  
SHML Sinus histiocytosis with massive lymphadenopathy  
ESR Erythrocyte sedimentation rate  
CT Computerised tomography  
MRI Magnetic resonance imaging  
CNS Central Nervous system  
ATT Anti-tubercular therapy

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Rosai-Dorfman disease is an uncommon idiopathic histiocytic disorder of lymph nodes and extranodal sites with rare central nervous system manifestation. The authors report three cases and summarise the existing literature. Overall, less than 50 cases have been described in world literature.

Its rarity makes it difficult to generalise and make definitive recommendations with respect to management strategies. However, based on prevailing data, a case should be made for total resection if this can be achieved with low morbidity rates. Where it is not possible to achieve a gross total resection, it is observed that many cases remain stable for a considerable period. Close monitoring can therefore be exercised with radiation therapy or possible radiosurgery administered at time of disease progression. This treatment strategy is similar to the management of various benign brain tumours.

More extensive collection of data and long term follow-up will be useful to elucidate the natural history and refine management strategies.

Comments

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