**Case Report**

**Intracranial Rosai-Dorfman Disease - a Case Report and a Review of the Literature**

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Rosai-Dorfman disease (RDD) is an uncommon, non-neoplastic benign lymphoproliferative disease characterized by prominent painless cervical lymphadenopathy with fever and leukocytosis. RDD is histologically characterized by emperipolesis, where large histiocytes become infiltrated with lymphocytes and plasma cells. Intracranial RDD is extremely rare. Only 79 cases have been reported, including the present case. Intracranial RDD is associated with headaches, seizures, and numbness caused by increased intracranial pressure. A 67-year-old Japanese woman presented with dizziness and was diagnosed with a tumor in the cerebral falx. The preoperative radiological diagnosis was meningioma. She had no lymphadenopathy. The patient underwent a craniotomy and tumor resection. The tumor consisted of lymphoid tissue with scattered lymph follicles. The infiltrating histiocytes showed emperipolesis. The histiocytes were immunoreactive for S-100 protein and CD68 and negative for CD1a, leading to the diagnosis of intracranial RDD. The postoperative course was uneventful without further therapy. The dizziness had not re-appeared and MRI demonstrated no recurrence of tumors for 7 months. Intracranial RDD shows a male predominance and occurs later in life than nodal RDD. The clinical manifestations and prognosis are variable depending on the location of the tumor and treatment. Most intracranial RDD have a benign course, but long-term follow-up is important, because recurrence has been observed.

**Keywords:** Rosai-Dorfman disease, intracranial, sinus histiocytosis with massive lymphadenopathy, emperipolesis, literature review

**Introduction**

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is an uncommon, non-neoplastic benign lymphoproliferative disease characterized by prominent painless cervical lymphadenopathy with fever and leukocytosis.1 Other clinical features of RDD are elevated erythrocyte sedimentation and polyclonal hypergammaglobulinemia.2 RDD is histologically characterized by emperipolesis, where large histiocytes become infiltrated with lymphocytes and plasma cells.3 The disease involves extranodal sites including the skin, salivary glands, orbit, liver and upper respiratory tract.2 RDD involving intracranial sites is rare, and intracranial Rosai-Dorfman without lymphadenopathy is very rare.4 Intracranial RDD presents headaches, seizures, and numbness caused by increased intracranial pressure. We report a case-study of a patient with intracranial RDD lacking lymphadenopathy and other organ involvement. We also provide a short review of the RDD literature.
Case Report

A 67-year-old woman presented with dizziness and was diagnosed with a tumor in the cerebral falx. A preoperative MRI revealed a mass (1.5 × 1.3 × 1.0 cm) in the left frontal side of the longitudinal fissure. The lesion demonstrated high signal intensity on T1-weighted images (Figure 1A) and isointensity to hypointensity on T2-weighted images (Figure 1B). On contrast-enhanced T1-weighted images, the lesion was heterogeneously enhanced and showed dural tail signs, indicating the extra-axial location (Figure 1C). There was no evidence of brain edema surrounding the lesion. The preoperative radiological diagnosis was meningioma.

Physical examination disclosed no lymphadenopathy. Hematological examination showed a white blood cell count of 6.5 × 10^3/μl (3.5-9.1 × 10^3), platelet count of 10.4 × 10^9/μl (13.0-36.9 × 10^9), platelet distribution wide of 11.5 fl (12.3-15.2), red blood cell count of 3.32 × 10^6/μl (3.76-5.00 × 10^6), Hb of 10.3 g/dl (11.3-15.2), and Hct of 30.4% (33.4-44.9). C reactive protein (CRP) was elevated at 1.67 (<0.17).

The patient underwent a tumor resection after craniotomy. The tumor was reddish and well-differentiated from the brain, and was attached to the cerebral falx. Total resection was achieved.

The tumor consisted of lymphoid tissue with scattered lymph follicles. In the sinus area between the follicles, there were pale-staining areas. There were also fibrous scar-like areas (Figure 2A). The infiltrating histiocytes contained abundant pale-staining cytoplasm and round nuclei, each with a defined nucleolus. This specific feature of engulfment of the erythrocytes and lymphocytes is called emperipolesis (Figure 2B). There were no Russell bodies or Dutcher bodies in the plasma cells.

On immunohistochemical examination, infiltrating plasma cells showed small amounts of kappa and lambda light chains. The infiltrating lymphocytes were a mixture of B and T cells, showing CD20 and CD3 positivity, respectively. The histiocytes were immunoreactive for S-100 protein and CD68, but negative for CD1a (Figure 3A, B, C). There were no meningeal cells immunoreactive for epithelial membrane antigen (EMA) in the lesions (Figure 3D). Periodic acid-Schiff and Grocott methenamine silver stain failed to identify any organisms.

Based on these histological and immunohistochemical findings, the lesion was diagnosed as intracranial RDD.
The postoperative course was uneventful and the patient was discharged 7 days after the operation without further therapy. Seven months after the operation, the dizziness had not reappeared and MRI demonstrated no recurrence of the tumor.

**Literature review**

We found a total of 79 reported cases of intracranial RDD, including the present case (Table 1).

<table>
<thead>
<tr>
<th>No.</th>
<th>Author, year</th>
<th>Age and Sex</th>
<th>Symptoms</th>
<th>Location/Size(cm)/Extracranial</th>
<th>Preoperative Diagnosis</th>
<th>Treatment</th>
<th>Prognosis</th>
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<tr>
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<td>21M</td>
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<td>-</td>
<td>R</td>
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<tr>
<td>2</td>
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<td>headaches, dizziness</td>
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<td>-</td>
<td>R+Ste</td>
<td>-</td>
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<td>-</td>
<td>-</td>
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<td>4</td>
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<td>lt petrous bone</td>
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<td>SR+Rad</td>
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<td>-</td>
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<td>-</td>
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<td>8</td>
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<td>35M</td>
<td>lt headaches</td>
<td>lt C-P angle and cavernous sinus/2.0</td>
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<td>SR</td>
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<td>9</td>
<td>Bhattacharjee 1992</td>
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<td>sellar</td>
<td>-</td>
<td>SR</td>
<td>7 M NR</td>
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<td>10</td>
<td>Shaver 1993</td>
<td>5M</td>
<td>III, IV, V, VI, VII palsies</td>
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<td>-</td>
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<td>1 M G</td>
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<td>TR</td>
<td>6 M NR</td>
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<td>22M</td>
<td>polydipsia, polyuria</td>
<td>pituitary/1.2</td>
<td>-</td>
<td>Bx+Ste</td>
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Nearly all cases occurred in the intracranial meningeal regions. Other locations included 12 cases in sellar and pituitary areas,5,11,21 5 cases in intraparenchymal areas,12-16 and 1 case in the fourth ventricle.17 Sixteen cases showed multiple intracranial lesions.11,18-20 20 cases had extracranial lymphadenopathy and other extra-nodal lesions.4,10,11,18,20,25,27-30 The sizes of lesions ranged from 1.2 to 8.0 cm.

Cases ranged in age from 2 to 78 years (mean = 38.9 years). There was a male predominance (48 males and 30 females, male:female ratio = 1.6:1). There was no difference in the age between intracranial RDD with or without extracranial lesions.

Symptoms included headaches, seizures, visual problems, and others. (Table 1)

Most of the cases were diagnosed as meningioma preoperatively. Other preoperative diagnoses included meningitis,9,45 glioma,13 and lymphoproliferative lesion.1

Treatments included total resection in 34 cases, subtotal resection in 26 cases, resection without comments in 10 cases, and biopsy sampling in 8 cases. For additional therapy after total resection, 2 cases received radiotherapy9,15 and 1 case received steroid therapy.1 After subtotal resection, 5 cases received radiotherapy22,23,35,40,44 and 4 cases received steroid therapy.21,35,50 After biopsy sampling, 6 cases were treated with steroid therapy,4,10,35,36,46 2 cases with radiotherapy18,36 and 1 case with anti-epileptic therapy,21 and 1 case with no treatment.9

Tumors recurred in 4 cases of the 33 cases with total resection9,25,30 and in 3 cases of the 26 cases with subtotal resection,27,30 regardless of postoperative therapy. There was only 1 case that resulted in death, probably not caused by RDD, that occurred 5 days postoperatively.21,47

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**Figure 3.** Immunohistochemical findings.
The infiltrating histiocytes in the sinus are immunopositive for S-100 protein (A). The reactivity varies from strong to weak. The histiocytes are also immunoreactive for CD68 (B), and are immunonegative for CD1a (C). There were no EMA-positive cells in the lesion including spindle cell elements (D). (Original magnification ×200, bar = 100 μm)
<table>
<thead>
<tr>
<th>No.</th>
<th>Reference</th>
<th>Age</th>
<th>Gender</th>
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<th>Outcome</th>
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<td>TR</td>
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<td>meningioma</td>
<td>R</td>
<td>-</td>
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<td>38M</td>
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<td>SR</td>
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<td>16</td>
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<td>24 M NR</td>
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<td>seizures</td>
<td>lt parieto-occipital</td>
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<td>SR</td>
<td>4 Y NR</td>
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<td>38M</td>
<td>headaches</td>
<td>lt parieto-occipital double/each</td>
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<td>TR</td>
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<tr>
<td>19</td>
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<td>37F</td>
<td>headache, dizziness</td>
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<td>TR</td>
<td>1 Y G</td>
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<td>20</td>
<td>Udono 1999(^{20})</td>
<td>67F</td>
<td>diplopia, headaches</td>
<td>rt frontal/-/nasopharynx</td>
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<td>headaches, visual impairment, amennorrea</td>
<td>suprasellar</td>
<td>pseudotumor</td>
<td>Bx</td>
<td>9 M Rec</td>
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<td>67F</td>
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<td>rt cerebellar</td>
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<td>TR</td>
<td>1 M NR</td>
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<td>seizures, headache</td>
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<td>1 Y NR</td>
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<td>-</td>
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<td>3 Y NR</td>
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<td>headaches</td>
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<td>SR</td>
<td>17 M NP</td>
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<td>headaches, seizures, memory problem</td>
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<td>-</td>
<td>SR</td>
<td>3 Y Rec</td>
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<td>39M</td>
<td>vertigo, visual disturbances</td>
<td>rt temporal/4.5</td>
<td>meningioma</td>
<td>TR</td>
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<td>55M</td>
<td>nasal obstruction, epistaxis</td>
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<td>TR</td>
<td>3 M Rec</td>
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<tr>
<td>30</td>
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<td>rt parietal/5</td>
<td>meningioma</td>
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<tr>
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<td>52M</td>
<td>headaches, malaise</td>
<td>lt petroclival/-/lymphadenopathy</td>
<td>meningioma</td>
<td>SR+Rad</td>
<td>15 M RE T</td>
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<tr>
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<td>SR+Ste</td>
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<td>7M</td>
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<td>rt parietal/-/lymphadenopathy</td>
<td>-</td>
<td>Bx+Ste</td>
<td>NC</td>
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<td>meningioma</td>
<td>TR</td>
<td>NR</td>
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<tr>
<td>36</td>
<td>Ture 2004(^{36})</td>
<td>6F</td>
<td>dizziness, vertigo</td>
<td>rt occipital and cerebellar hemisphere/3</td>
<td>-</td>
<td>SR</td>
<td>4 M NP</td>
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<td>60F</td>
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<td>meningioma</td>
<td>TR</td>
<td>3 M Rec</td>
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<td>Bx+Ste</td>
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<td>lt headache</td>
<td>lt frontal</td>
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<td>SR</td>
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<td>Bx+Ste+Rad</td>
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<td>suprasellar/3</td>
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<td>TR</td>
<td>3 M NR</td>
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Since RDD was first reported by Rosai and Dorfman in 1969,1 more than 750 cases have been reported.2 RDD generally affects persons 20 years of age or younger (mean age 20.6) and shows a male predominance.2, 4 About 90% of cases have painless cervical massive lymphadenopathy. Extranodal RDD has been reported in approximately 43% of cases.2 Intracranial RDD without lymphadenopathy or other organ involvement is extremely rare (0.5% of cases).4 The male predominance for intracranial RDD is the same, but the mean age is higher (2 to 78 years, mean age 38.9).

Intracranial Rosai-Dorfman disease mainly develops in dural regions. On radiographical findings, the lesions appear to be dural-based, extra-axial and enhanced with surrounding vasogenic edema.49 Therefore, the preoperative diagnosis for most cases is meningioma, other cerebral tumors or lymphoproliferative lesion. Of those with intracranial RDD, 7.6% of cases have lymphadenopathy and 17.7% show other systemic lesions. More than 70% of RDD cases have presented with only intracranial lesions.

The clinical manifestations of intracranial RDD are due to increased intracranial pressure. Most cases complain of headaches, followed by seizure and visual and hearing impairments. Those at sellar and pituitary areas may cause polydipsia, polyuria, and galactorrhea. Histology and immunostaining are essential to reach a correct diagnosis of RDD.10 Intracranial RDD needs more careful observation, since patients are reported to show more fibrosis, fewer typical histiocytes, and less emperipolesis than nodal RDD.2

Diagnosis of intracranial RDD can be difficult to differentiate from lymphoplasmacyte-rich meningioma, plasma cell granuloma, Langerhans cell histiocytosis, and plasmacytoma.26, 45, 50 Lymphoplasmacyte-rich meningioma shows infiltrating meningeal cells immunoreactive for EMA,50 which can distinguish it from RDD, in which structural cells do not express EMA. It may be difficult to differentiate...
RDD from plasma cell granuloma. Microscopic features of plasma cell granuloma reveal histiocytes that are negative for S-100 protein and do not show emperipolesis. On the other hand, the histiocytes in RDD are immunopositive for S-100 protein and emperipolesis is present, differentiating it from plasma cell granuloma. Although S-100 protein-positive histiocytes showed variable reactivity, the intensity of S-100 protein is reported to be variable by the size of the histiocytes. Langerhans cell histiocytes is positive for CD1a, but the histiocytes in RDD are negative for CD1a. Plasmacytoma shows a cytologically monotonous and monoclonal proliferation of plasma cells. The plasma cells of our cases were polyclonal and immunopositive for kappa or lambda light chain.

Intracranial RDD mimics menigioma or other intracranial tumors radiologically and clinically. Therefore, the first choice for treatment is surgical resection. Surgical treatment contributes not only to reducing increased intracranial pressure but also to confirming the diagnosis. Surgical resection was very effective in many cases. After subtotal resection, some cases respond with tumor decreases following radiotherapy and steroid therapy, but other cases showed tumor recurrence in the same region and in other regions such as the spine. The prognosis of intracranial RDD is favorable. Most patients have no recurrence after surgery, however, 14.3% of cases showed recurrence within the follow-up period of 3 months to 5 years. Therefore, intracranial RDD needs further study to facilitate discovery of additional treatment options. Our case has shown no signs of recurrence 7 months after surgery.

Intracranial RDD is very rare and the clinical course and treatment of this disease are therefore not well established. The clinical manifestations and prognosis is variable depending on the location and treatment. Most intracranial RDD are benign diseases, but long-term follow up is important.

References

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Intracranial RDD is very rare and the clinical course and treatment of this disease are therefore not well established. The clinical manifestations and prognosis is variable depending on the location and treatment. Most intracranial RDD are benign diseases, but long-term follow up is important.
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