

# Intracranial Rosai-Dorfman syndrome mimicking meningioma

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**Abstract:** Rosai-Dorfman disease (RDD) is a rare and idiopathic histioproliiferative disorder characterized by painless cervical lymphadenopathy, fever and weight loss. Intracranial involvement is extremely rare.

A case of central nervous system (CNS) involvement in RDD disease is reported in a 36-year-old male who presented with generalised tonic clonic seizures. Magnetic resonance imaging study revealed an enhancing mass in the right anterior third parasagittal region simulating with meningioma.

A Simpson grade II excision was done in view of a clinico-radiology suggestive of meningioma. Histopathology examination confirmed RDD. Extranodal involvement is reported at various sites, but CNS involvement remains extremely rare. The clinical presentation and differential diagnosis of RDD are discussed. (p70-72)

**Key words:** Rosai-Dorfman Disease, meningioma and central nervous system

## Introduction

In 1969, Rosai and Dorfman described the entity of sinus histiocytosis with massive lymphadenopathy, a benign disorder characterised by an unusual proliferation of histiocytes.<sup>15</sup> It is generally associated with bilateral, painless, cervical lymphadenopathy, fever, weight loss, leucocytosis, elevated erythrocyte sedimentation rate and hypergammaglobulinemia. The majority of patients are younger than 20 years of age.<sup>16</sup> Extranodal involvement comprises nearly 40% and predominant organ involvement includes bone, skin and soft tissue, upper respiratory tract, salivary glands, eye and orbit, digestive system, testes, heart, female genital system, thyroid, and pituitary gland.<sup>4</sup> Central nervous system (CNS) involvement is extremely rare.<sup>1</sup> Intracranial involvement is usually in the form of a dural based lesion that mimics meningioma.<sup>8</sup> Three intraparenchymal cases and a single intraventricular case have been previously reported.<sup>5,7,11,12</sup> Nearly 50 cases have

been reported in the world literature to the best of our knowledge. We are reporting a case of solitary intracranial lesion mimicking a parasagittal meningioma.

## Case Report

A 36-year-old male presented with a single episode of generalised tonic clonic seizures 1-year back. Patient had been given treatment for neurocysticercosis following contrast computed tomography (CT) scan of head before attending the outpatient department of our institute. He had received a course of albendazole and was receiving phenytoin for seizure control. Patient came to us when he developed mild and continuous headache of 1-month duration. His neurological examination including fundus was unremarkable. Routine investigations including x-ray chest were normal. Contrast CT head (1-year previously) revealed a hyperdense enhancing extra-axial mass attached with right frontal dura with dural enhancement medially and laterally to the lesion. The oedema was present posterior to lesion in right frontal cortex.

Magnetic resonance imaging (MRI) study revealed an enhancing mass of size  $4.3 \times 3.2 \times 1.8$  cm in the anterior third convexity dural region on right side. Mass was appearing to be extra-axial having broad based dural attachment with grade II perifocal oedema in right frontal cortex adjacent to lesion posteriorly (Fig. 1). Considering the diagnosis of meningioma the patient was taken for surgery and right frontal craniotomy was done and complete resection was achieved. Tumour was yellowish grey, firm, vascular with poor plane of cleavage from

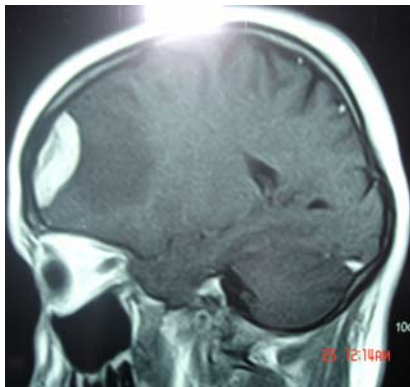
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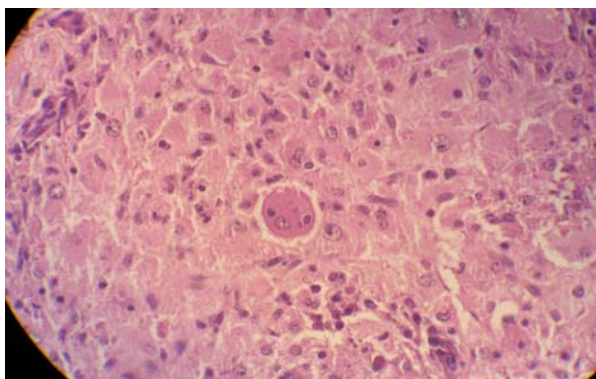
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surrounding parenchyma and attached with dura. The involved dura was excised. Consistency of tumour was similar to meningioma. A duraplasty was required after excision of mass.

Microscopic examination revealed diffuse sheets of histiocytes with round to oval, vesicular nuclei and abundant foamy cytoplasm, admixed with lymphocytes and plasma cells. Some histiocytes were seen with mononuclear inflammatory cells engulfed within their cytoplasm (emperipolesis). There were no microorganisms, necrosis or granuloma formation. Immunohistochemical studies demonstrated positive reaction to S100 and CD68 proteins among the histiocytes. The lymphoid tissue was a mixture of T- and B- cells. The features were consistent with Rosai-Dorfman disease (RDD) (Fig. 2).



**Figure 1** - MRI study revealed an enhancing mass of size 4.3 × 3.2 × 1.8 cm in the anterior third convexity dural region on right side. Mass was appearing to be extra-axial having broad based dural attachment with grade II peritumoral oedema in right frontal cortex adjacent to lesion posteriorly



**Figure 2** - Microscopic examination revealed diffuse sheets of histiocytes with round to oval, vesicular nuclei and abundant foamy cytoplasm, admixed with lymphocytes and plasma cells. Some histiocytes were seen with mononuclear inflammatory cells engulfed within their cytoplasm (emperipolesis)

At 4 months follow-up the patient was asymptomatic with normal physical and neurological examination. Seizures were controlled on phenytoin. Repeat MRI did not reveal any residual.

## Discussion

Isolated intracranial RDD has been documented in 49 previous case reports.<sup>6</sup> As compared to the more common systemic diseases it tends to involve the older age group (mean 39.4 years).<sup>4</sup> Clinically CNS lesions are usually present with cephalgia, seizure or cranial nerve deficits. The location can include suprasellar region, convexity, parasagittal region, cavernous sinus, and petroclival region.<sup>8</sup> The most of previously reported intracranial cases of RDD displayed dural attachment.

The aetiology of RDD is still unknown. Studies of monokine expression suggest derivation of histiocytic cells from activated macrophages that produce interleukin - 1 $\beta$  (IL-1 $\beta$ ), tumour necrosis factor-  $\alpha$  (TNF- $\alpha$ ).<sup>3</sup> Levine, et al suggested that human herpes virus 6 and to a lesser extent Epstein Barr virus may be involved in the aetiology.<sup>9</sup> Serological evidence of Epstein Barr virus has been found in about 50% cases of RDD, but increased serological titres may be the result of non-specific host immune response and not the cause of the disease.<sup>2</sup>

Radiologically and at operation, the appearances of RDD and meningioma are remarkably similar.<sup>2</sup> The diagnosis is based on histopathological appearances of RDD characterized by an infiltrate of lymphoplasmacytic cells and histiocytes of varying size. The large histiocytes often show emperipolesis (lymphophagocytosis). On immunohistochemical examination, these are positive for S100 protein and negative for CD1a, a marker of Langerhan's cell histiocytosis.<sup>19</sup>

The differential diagnosis includes Langerhan's cell histiocytosis, lymphoplasmacyte rich meningioma, lymphoproliferative disorders, plasma cell granuloma and infectious disease. Langerhan's cell histiocytosis may also present as a dural based lesion. It differs from RDD in that it usually has extensive eosinophilia, does not exhibit emperipolesis, and the histiocytes contain Birbeck's granules on electron microscopy and are immunoreactive for CD1a. Lymphoplasmacyte-rich meningioma or granuloma may be seen identical to meningioma on CT, MRI and during surgery. However, on microscopy areas of typical meningotheial, transitional or fibrous meningiomas are seen with extensive lymphocytic and plasma cell infiltrates.<sup>10</sup> Lymphoproliferative disorders like aggressive lymphomas may also contain phagocytic histiocytes which are S100 positive. Aggressive lymphomas exhibit frank malignant cytological features on microscopic examination. Plasma cell granulomas have a polymorphous cellular

infiltrate consisting of lymphocytes, plasma cell, histiocytes and foamy macrophages but unlike RDD S100 positive histiocytes and emperipolesis are not seen.<sup>14</sup> Intracranial infections including mycobacterial and fungal can be excluded by special stains and cultures.

Intracranial RDD is generally considered to be benign although three deaths have been reported.<sup>6</sup> It has usually been treated with surgery and patients with complete resection experienced no recurrence.<sup>8</sup> Adjunctive treatments have included chemotherapeutic agents, irradiation and steroids but their efficacy is not clear.<sup>13,17,18</sup>

### Conclusion

Although intracranial RDD is a rare lesion of CNS, it should be included in the differential diagnosis of a dural mass that clinically and radiologically resembles meningioma. Total surgical excision alone appears to be the adequate treatment modality, however long term follow-up is required to determine the prognosis and natural history of this disease.

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## ERRATUM

The Editorial office wishes to apologise for a typesetting error in Vol. 11(2), October, 2007 edition in the Clinical Study 'Multiple factor analysis of prognosis in lumbar disc herniation', by Kumar, et al., page 62, Fig. 1 should read L5 - S1.

The author would also like to correct , Table 5, page 62 - Unsatisfactory outcome should, of course, read (n = 16).