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Rosai-Dorfman disease of Meninges: Report of 2 cases with review of literature

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Abstract

Rosai-Dorfman disease (RDD) also called Sinus histiocytosis with massive lymphadenopathy is a rare benign histiocytic proliferative disorder predominantly affecting lymph nodes. Extra nodal sites include central nervous system, head and neck region, eyes, upper respiratory tract and skin. Involvement of the central nervous system (CNS), especially in the absence of nodal disease is rare. We are reporting 2 cases of RDD in the meninges. Both cases are presented with intractable headache and seizures. Clinically and radiologically, both were diagnosed as meningioma. Histopathologically both turned out as Rosai-Dorfman disease. IHC studies confirmed the diagnosis.

Keywords: *Central nervous system; Emperipolesis; Histiocytic proliferation; Rosai-Dorfman disease.*

Introduction

Rosai Dorfman disease (Sinus histiocytosis with massive lymphadenopathy) is a rare benign histiocytic proliferative disorder. Over 650 cases have been reported since 1969.[1] Isolated intracranial RDD is extremely rare[1]. Only 54 cases have been reported in CNS[2]. Our cases are dura based nodular mass which simulated meningioma clinically. These cases are being reported because of its rarity and the chance for a wrong diagnosis, clinically and radiologically. The correct diagnosis is important because of good response to surgical treatment.

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Case report

Case 1

Fourty five year old male presented with intractable headache and seizures for a period of 3 months. Clinical examination showed signs of raised intracranial tension and meningeal irritation. All his routine investigations were within normal limits. CT scan showed a single temporal mass enhancing lesion suggestive of meningioma. [Figure 1]. MRI Brain showed multiple extra axial intense homogeneously enhancing lesion over right parietal, frontal & temporal convexity without impinging on cerebral parenchyma and bone, suggestive of meningioma/ neurofibroma. Grossly it was a well circumscribed nodular yellowish mass with dural attachment. [Figure 3]. Histopathology showed an inflammatory lesion composed of histiocytes and mononuclear cells. The histiocytes were large with abundant eosinophilic cytoplasm, vesicular nuclei and prominent nucleoli .A few atypical histiocytes were present. [Figure 4]

Many of the histiocytes showed emperipolesis. [Figure 5]. Plasma cells & lymphocytes were seen intermixed with the histiocytes. IHC showed strong S 100 positivity in the histiocytic population. [Figure 6]

Case 2

Second is the case of 50 year old male patient with history of late onset seizures of 6 month duration. Clinical examination showed signs of raised intracranial tension and meningeal irritation. All his routine investigations were within normal limits. CT scan showed a homogeneous lesion with dural attachment measuring 2.4X3 cm in left parietooccipital region with surrounding odema, suggestive of meningioma. [Figure 2]

Fig 1. CT scan brain of case1 showing a mass lesion diagnosed as meningioma

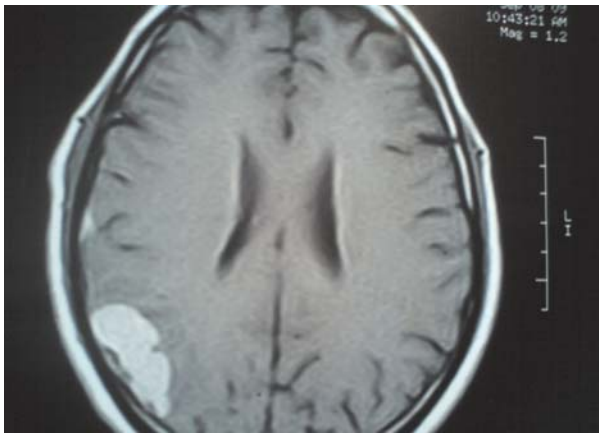


Fig 2. CT scan brain of case 2 showing the lesion diagnosed as meningioma

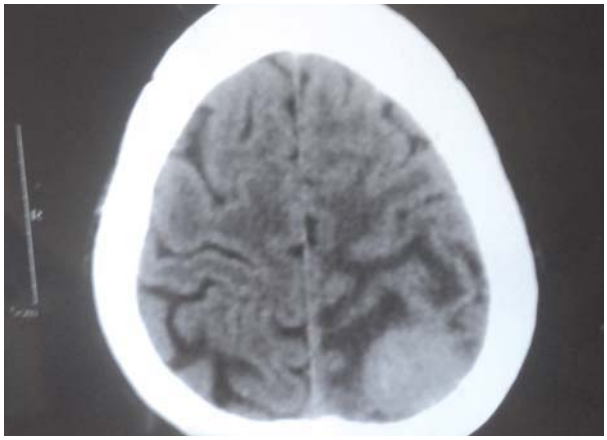


Fig 3. Gross - well circumscribed nodular yellowish mass with dural attachment

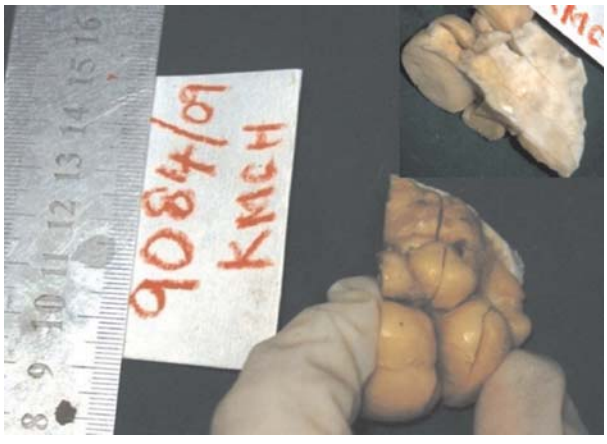


Fig 4. Histiocytes showing emperipolesis (400X)

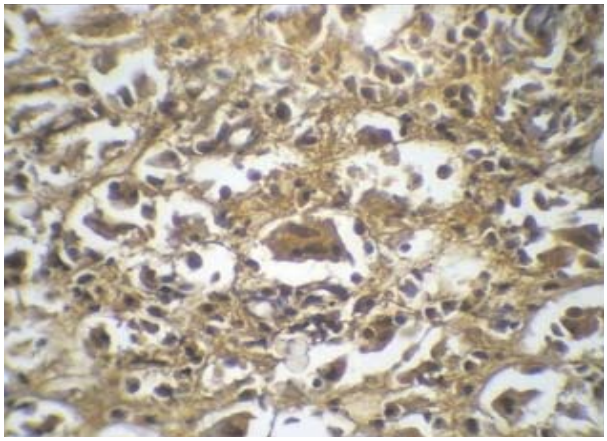


Fig 5. S100 positivity

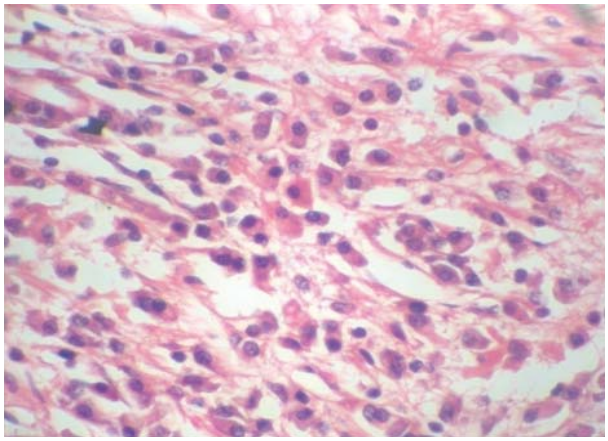
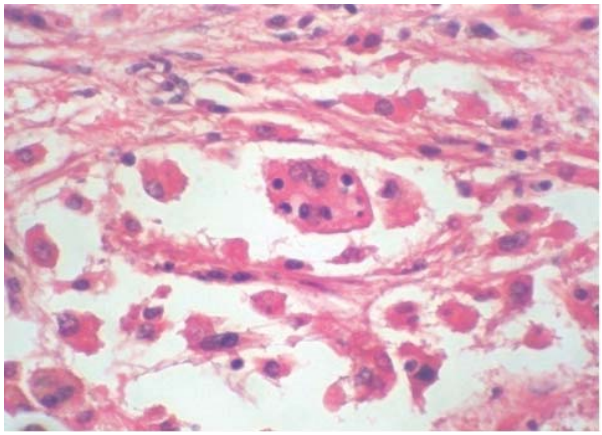


Fig 6. Higher power showing dense infiltrate of mononuclear cells & macrophages (400X)



Discussion

Rosai-Dorfman disease (Sinus histiocytosis with massive lymphadenopathy) was first described in 1969 as a benign lesion with systemic symptoms and lymphadenopathy[1,2]. Rosai-Dorfman disease is rarely found intracranially[1-3]. Only few cases are reported in the literature. Since it mimics meningioma clinically and radiologically, histopathological examination is the tool for correct diagnosis[2-5].

The present case was also clinically and radiologically diagnosed as meningioma. But histologically, it was an inflammatory lesion with plenty of histiocytes, many of which showing emperipolesis, suggesting a diagnosis of Rosai-Dorfman disease which was confirmed by IHC studies. Literature review showed a number of cases of RDD in different parts of body. Very few cases are reported in Central nervous system. Reported cases in CNS are less than 50 in number. In almost all cases excision was performed with the diagnosis of meningioma[2,4]. Final diagnosis was established in histopathological examination[4-6]. Light microscopic examination of H&E slides alone is sufficient to arrive at a diagnosis. IHC markers are performed for confirmation[3-5]. Usual marker is S100 which shows strong positivity-both cytoplasmic & nuclear positivity. Our cases also showed strong S100 positivity.

Differential diagnoses to be included are granulocytic sarcoma, inflammatory myofibroblastic tumor. These can be ruled out by the histomorphological appearance.

Correct diagnosis of a case of RDD, is lifesaving to the patient since complete excision of the lesion is curative and the pathologist should be aware of these mimickers for accurate diagnosis. Awareness of occurrence of RDD in a non-nodal location is necessary to avoid misdiagnosis. Our patients have remained asymptomatic for the last 2 years with no evidence of any recurrence or nodal disease till date.

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