Case Report

Rosai-Dorfman disease mimicking a sphenoid wing meningioma

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A 40-year-old male presented with a single generalized tonic-clonic seizure. MRI revealed an enhancing, dural-based, left lateral sphenoid wing lesion suggestive of a meningioma. At microsurgical excision, the lesion was firm and relatively avascular. The histopathological report revealed S-100 positive histiocytic proliferation with lymphophagocytosis (emperipolesis) characteristic of the Rosai-Dorfman disease. The case and its management are discussed.

Key Words: Rosai-Dorfman disease, microsurgery, meningioma, CNS, extranodal sinus histiocytosis with massive lymphadenopathy

Introduction

The first report of sinus histiocytosis with massive lymphadenopathy (SHML) has been attributed to Destombes who biopsied a 24-year-old man in 1959. This was originally described as a lipid storage disorder (*adenitis avec surcharge lipidique*) developing after inflammation.[1] A decade later, Rosai and Dorfman recognized the clinical syndrome caused by this rare, benign, lymphoproliferative disorder characterized by lymphophagocytosis (emperipolesis) for which they coined the term SHML.[2] This disease entity predominantly affects children and young adults who present with painless, bilateral, cervical lymphadenopathy. Nearly 25% to 43% of all patients have at least one site of extranodal involvement, which has prompted the use of the terminology "Rosai-Dorfman disease."[3-5] Isolated central nervous system (CNS) manifestations are extremely rare with only 49 cases reported previously in the literature to the best of our knowledge.[4-14]

Case Report

A 40-year old male, presented with a history of generalized tonic-clonic convulsions. There was no neurological deficit. Investigations revealed a relatively large mass in the region of the lesser wing of the sphenoid bone simulating a meningioma (Figures 1 and 2). The patient underwent a left pterional osteoplastic flap craniotomy and a near total excision of the lesion was performed. The tumor was well-defined, extra-axial, grayish yellow, waxy, relatively avascular and poorly suckable. It was arising from a broad dural base around the lesser sphenoid wing. There was another lesion of a similar nature

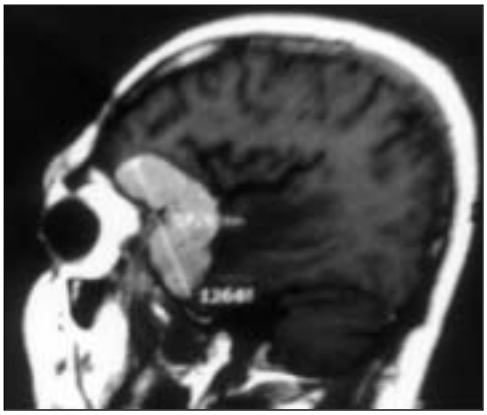

Figure 1: Contrast enhanced left parasagittal MR image revealing an enhancing, extra-axial left lateral sphenoid mass lesion straddling the floors of the anterior and middle cranial fossae

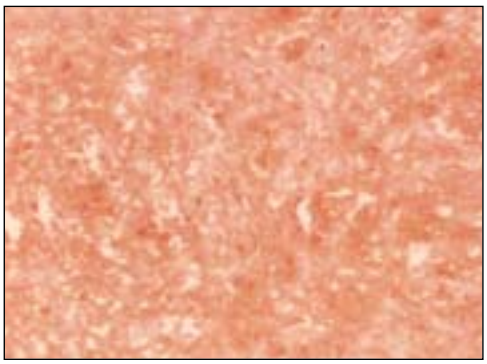

Figure 2: Histiocytic infiltrate with emperipolesis. The site of lymphocytic phagocytosis by the histiocyte (H/E, x40)

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arising from the greater wing of the sphenoid in the left infratemporal fossa. This was biopsied separately. Histology showed that the mass was extranodal SHML (Rosai- Dorfman disease).

At follow-up after 12 weeks, a contrast enhanced CT scan of the head did not reveal any residual or recurrent mass lesion. The CT of the thorax and abdomen done at the same time did not indicate any lymphadenopathy. The patient was seizure-free.

Discussion

CNS Rosai-Dorfman disease most commonly involves patients between 22 and 63 years of age, with a definite male predominance. The mean age at presentation is 41 years much as in our case.[6,11] The commonest presentations in the largest single series to date are headaches, seizures, numbness, and paraplegia though Petzold et al in their meta-analysis of 32 cases have described a 25% incidence of visual symptoms, which were the presenting feature in 19% of all cases.[11,13] Approximately 75% of all cases are intracranial with 90% involving the leptomeninges. Purely intraparenchymal lesions have also been reported.[8,11] The commonest findings on imaging suggest dural-based, extra-axial, enhancing masses with perilesional cerebral edema. Thus, the disease closely mimics meningiomas clinically and radiologically.[4,6,8,10,13] The differential diagnosis includes Wegener's granulomatosis, sarcoidosis, Hodgkins disease, plasma cell granuloma, inflammatory pseudotumor and Langerhans histiocytosis. Rosai-Dorfman disease is of uncertain pathogenesis. The large pale histiocytic proliferation may represent an autoimmune pathology or a reaction to an as yet unidentified infectious agent.[6] The histiocytes test positively for the S-100 protein and CD68 (KP1) but negatively for CD1a.[6,11] Emperipolesis, signifying the phagocytosis of lymphocytes, is characteristic of Rosai-Dorfman disease but is present in only 70% of cases.[6]

Andriko et al followed up 10 cases of surgically treated CNS disease over a mean of 15 months and found that nine patients had no evidence of disease progression. One patient died of operative complications 5 days after biopsy.[11] The operative mortality after surgery is thus nearly 4% (Two out of a total of 50 cases).[13]

Though the decision to follow patients after subtotal excision with serial imaging alone is the norm, Hadjipanayis et al have recently reported the use of stereotactic radiosurgery after partial microsurgical excision of a petroclival lesion extending into the left cavernous sinus.[12] Petzold et al found intracranial tumour regrowth or recurrence of symptoms in 14% of 29 patients with a mean follow-up of 10.1 years. Of those patients described as 'stable,' only 52% had undergone brain imaging at follow-up. They concluded that a five-year follow-up with brain imaging was essential and advocated local low-dose radiation to treat patients with subtotal resection or recurrence.[13] Horneff et al have advocated the use of high-dose steroids, methotrexate and 6-mercaptopurine for the treatment of complicated SHML after noting a seven-year disease-free survival in a three-year-old girl. Initial steroid monotherapy resulted in relapse and chemotherapeutic treatment using etoposide was ineffective. Their protocol involves combined low-dose methotrexate and 6-mercaptopurine therapy for four months to ensure remission followed by 6-mercaptopurine for a total of two years.[14]

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