Isolated plasma cell granuloma of the meninges

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Sir,

A 77-year-old female with hypertension, presented with right partial motoric seizures. Neurological examination showed right hemiparesis. Brain MRI revealed extensive post contrast interhemispheral dural, pachimeningeal and leptomeningeal enhancement of the left cerebral hemisphere (Figure 1), which were initially interpreted as meningeal carcinomatosis. However CSF analysis performed on two occasions, with normal cell count, glucose and protein levels, did not show malignant cells. Chest and abdominal CT scan and mammography were normal. Brain and meningeal biopsies were performed and histological findings (Figure 2) were consistent with plasma cell granuloma. Following this bone marrow biopsy, skeletal X-rays survey, peripheral blood cell count, renal function, calcium and protein electrophoresis were normal. The patient was treated with corticosteroids without any neurological sequel and significant improvement of MRI findings.

This case emphasizes how the diagnosis of leptomeningeal carcinomatosis remains challenging, as neither CSF cytology nor MRI is adequately sensitive for the diagnosis. Plasma cell granulomas are uncommon, benign inflammatory masses of unknown etiology.[1] They are histologically characterized by polyclonal plasma cells, large histiocytes with lymphophagocytosis and fibrosis.[1] There are only few reports of isolated CNS plasma cell granuloma in the literature.[1] Based on similar histological features, plasma cell granulomas, Rosai-Dorfman disease, and idiopathic hypertrophic pachymeningitis are now believed to form a spectrum of inflammatory or reactive conditions with, probably, the same unknown etiology.[2,3]

Rosai–Dorfman disease, or sinus histiocytosis, with massive lymphadenopathy, is a benign idiopathic histiocytic proliferative disease with pathognomonic histological and immunohistochemical characteristics. Extranodal involvement occurs in 43% of cases[4], and by December 2008 111 cases of Rosai–Dorfman disease involving the central nervous system had been reported in the literature.[5] The main histopathological differentiation between plasma cell granuloma and Rosai–Dorfman disease is a careful appraisal of the morphological features together with the absence of emperipolesis in the S100 positive histiocytes.[5] It has also been demonstrated that some cases of idiopathic hypertrophic pachymeningitis show the histopathological features of meningeal plasma cell granulomas and occasionally show pseudo-tumoral thickening of the dura.[3] As all three conditions are rare, their radiological features can easily be confused with meningeal carcinomatosis and histological features can be easily confused with other diseases like plasmacytoma or lymphoplasmacyte-rich meningioma.[6]

In any of these instances, brain and meningeal biopsy and careful histological examination is crucial in reaching the correct diagnosis.

References

1. Brandsma D, Jansen GH, Spliet W, Van Nielen K, Taphoorn MJ. (2003) The diagnostic difficulties of meningeal and intracerebral plasma cell granulomas--presentation of three cases. J Neurol 250:1302-6.
2. Govender D, Chetty R. (1997) Inflammatory pseudotumour and Rosai-Dorfman disease of soft tissue: a histological continuum? J Clin Pathol 50:79–81.
3. Deprez M, Born J, Hauwaert C, Otto B, Reznik M. (1997) Idiopathic hypertrophic cranial pachymeningitis mimicking multiple meningiomas: case report and review of the literature. Acta Neuropathol (Berl) 94:385–389.
4. Foucar E, Rosai J, Dorfman R. (1990) Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Review of the entity. Semin Diagn Pathol 7:19–73.
5. Adeleye AO, Amir G, Fraifeld S, Shoshan Y, Umansky F, Spektor S. (2010) Diagnosis and management of Rosai-Dorfman disease involving the central nervous system. Neurol Res 32:572-8.
6. Sato K, Kubota T, Kitai R, Miyamori I. (2006) Meningeal plasma cell granuloma with relapsing polychondritis. Case report. J Neurosurg 104:143-6.

Figures

Figure 1. Brain MRI showing extensive post contrast interhemispheral dural enhancement, as well as pachimeningeal and leptomeningeal enhancement of the left cerebral hemisphere with involvement of subarachnoidal spaces of the left hemisphere convexities. On FLAIR images hyperintensity of subcortical white matter is seen. A and B) FALIR transversal images; C and D) T1 postcontrast transversal images; E) T1 postcontrast coronal image; F) T1 postcontrast sagittal image.

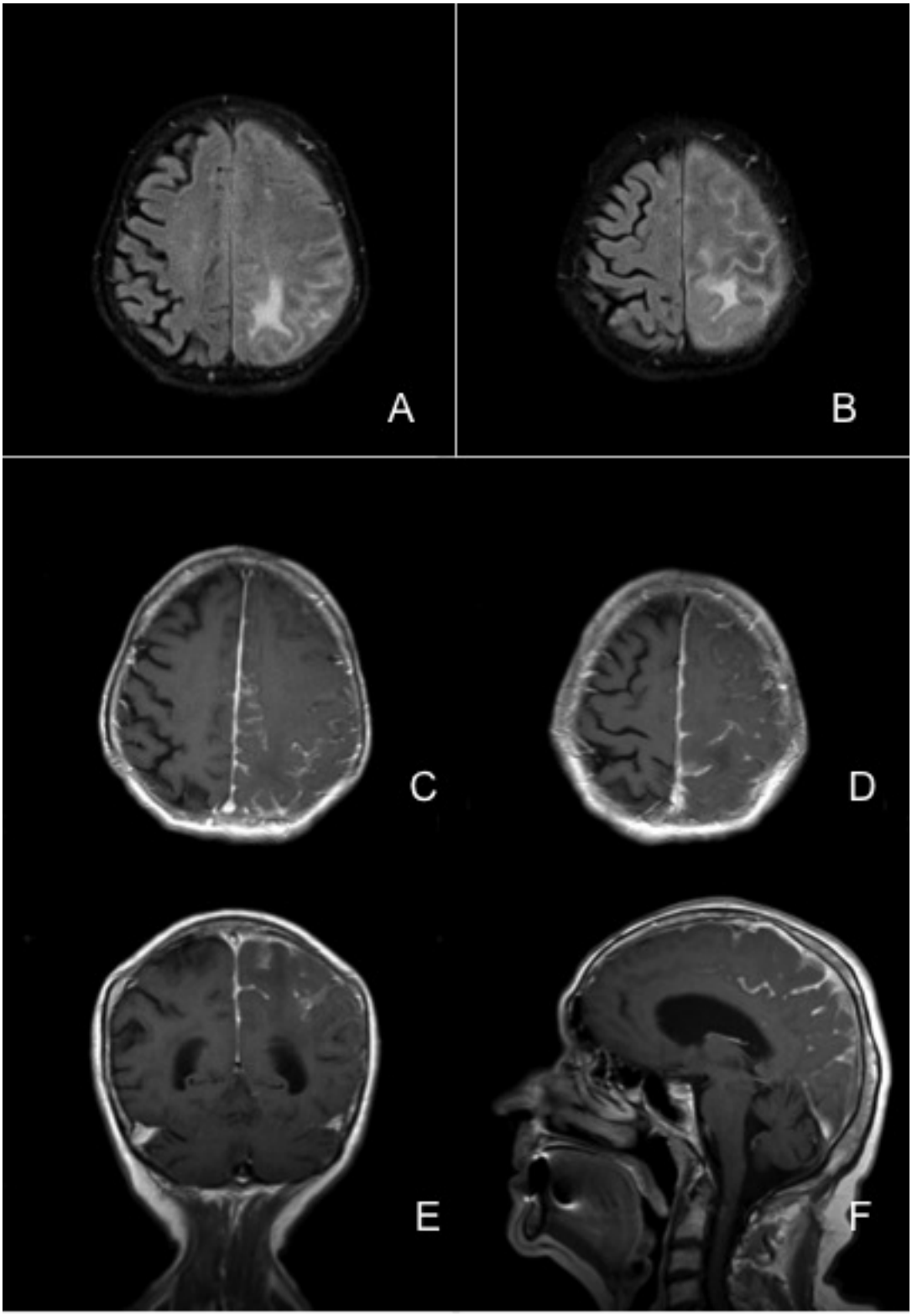
A
B
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Figure 2. A) Biopsy specimen showing normal brain surface with an inflammatory infiltrate composed of plasma cells, lymphoid aggregates, individual histiocytes and leukocytes on the surface (HE, x4); B) Plasma cells without significant atypia predominate (HE, x40); C) Immunohistochemistry on CD138 confirmed plasma cells (x20); D) Immunohistochemistry on CD56 was negative (x20); E) and F) Immunohistochemistry on kappa and lambda light chains showed polyclonal plasma cell population (x20).