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Neuropathology Education

Tumor arising in the periventricular region

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CLINICAL COURSE

A 32-year-old immunocompetent female patient, 8 months pregnant, underwent three complex partial seizures beginning with visual hallucinations and secondarily generalized. She had been in good health until then. The pregnancy was normal and no high blood pressure was noted. Clinical examination revealed a left nystagmus, a right sensory hemibody deficiency and a right pyramidal syndrome. MRI showed an isolated intraparenchymal lesion in the left parieto-occipital region with an enhancement of the left lateral ventricle floor (Fig. 1a,b). The biological assessments, including hemogram, ionogram, CRP, liver enzymes and β2 microglobulin, were all normal. An elevated erythrocyte sedimentation rate was noted. CMV, HBV, HCV, and Lyme serologies were negative. Previous HSV, EBV, and varicella zoster virus infection markers were found. CSF examination revealed hyperproteinorachia (0.95 g/L) without cellular reaction. No tumoral cells were found. Thoracic and abdominal CT was normal. After the brain biopsy, a course of corticosteroids was started. The patient did not present with any other seizures. Five years after the first epileptic manifestations, follow-up MRI demonstrated resolution of the enhancing lesions (Fig. 2) and the patient was in good health with 10 mg of corticosteroids daily.

PATHOLOGICAL FINDINGS

Microscopically ventricular and periventricular specimens consisted of a mixed lymphoplasmacytic inflammatory infiltrate containing sheets of large and foamy histiocytes. Focally some of these histiocytes engulfed a large number of lymphocytes and plasma cells, corresponding to emperipolesis (Fig. 3a). Some elements were negative: no viral, bacterial or fungal inclusions, parasitic infection, tumoral cells or granuloma were found. By immunohistochemistry, histiocytic cells were positive for S100 protein (Fig. 3b), as well as for CD68 (KP1), CD31 (granular and membranous immunoreactivity) (Fig. 3c) and vimentin. Histiocytes were negative for GFAP and CD1a.

DIAGNOSIS

Rosai-Dorfman disease (RDD), also called sinus histiocytosis with massive lymphadenopathy.

DISCUSSION

Rosai-Dorfman disease is a rare benign histiocytosis. First described by Destombes in 1965,[1] the disease was defined as an idiopathic histiocytic lymphoproliferative disorder in 1969 by Rosai and Dorfman.[2] The term Rosai-Dorfman disease may be preferred to sinus histiocytosis with massive lymphadenopathy because extra-nodal involvements occur in about 40% of patients, with or without lymphadenopathy.[3] CNS involvement is rare with 80 cases reported to-date. The mean age of intracerebral RDD is about 38 years, above the 21 years reported by Foucar *et al.* in 1982 which concerned all RDD patients.[4] Involvement of the suprasellar region remains rare and can only concern the pituitary gland.[5] Four cases of intraparenchymal brain location of RDD were reported[6–9] but only three[7–9] presented isolated intraparenchymal lesions without dural attachment. One case of intraventricular location was documented, in the floor of the fourth ventricle.[10]

The characteristic microscopic features of RDD are infiltration of histiocytes, B and T lymphocytes and

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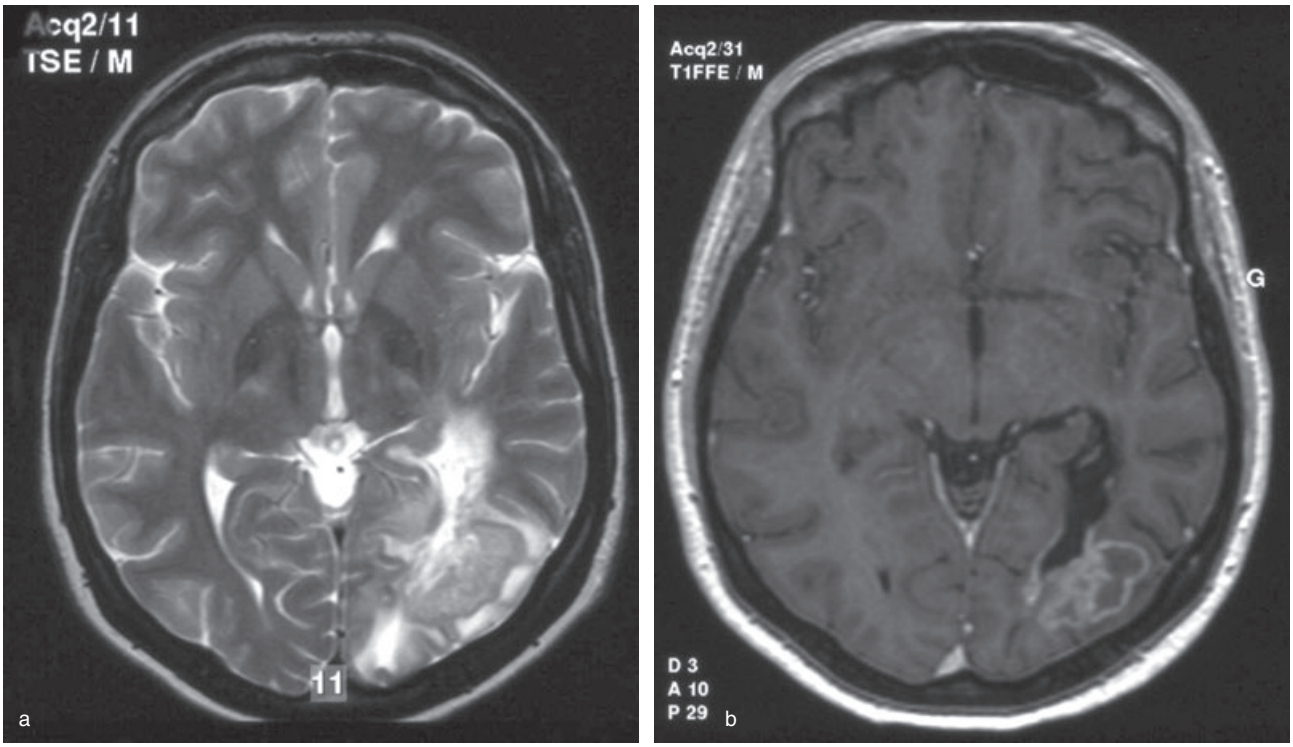

Fig. 1 Initial T2-weighted axial MRI showing a left periventricular hypersignal (a) and gadolinium T1-weighted axial MRI showing the ependymal enhancement (b).

plasma cells. Emperipolesis is typical of RDD of the leptomeninges but is not seen in 30% of cases.[11] On immunocytochemical studies, expression of CD68, CD31, α1 antitrypsin and α1 antichymotrypsin are positive both for mononuclear cells and dendritic cells, but a positive expression of S100 is found only for dendritic cells. CD1a is not expressed by any cells in RDD. As elucidated by Purav in a recent series including 10 cases of RDD of the CNS, the diagnosis of RDD is entirely based on histopathology and immunohistochemistry.[12]

In the classical form of RDD, the prognosis is usually good but the clinical course remains unpredictable. No consensus is established for the treatment of intracranial RDD. Clinical follow-up may be sufficient when lesions are not massive and do not involve vital organs. The most efficient treatment for meningioma-like lesions is surgical resection. Steroid agents could be considered as a conservative therapy, avoiding invasive procedures.

In summary, RDD is a rare entity which may only affect the CNS. Most of the intracranial lesions reported are dural-based. When the location is exceptionally intraparenchymal without dural attachement, the lesion can be mistaken for a glial neoplasm. Diagnosis is made via biopsy and steroids can be proposed when surgical excision is not possible.

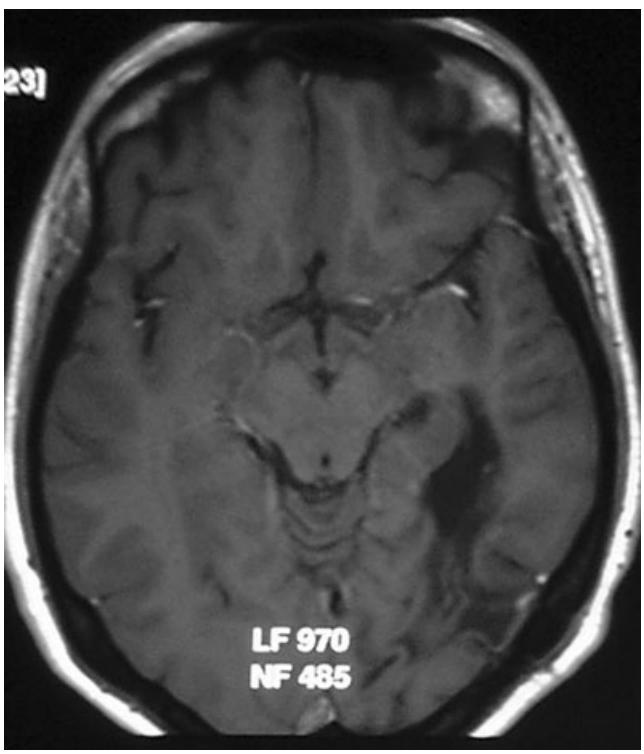

Fig. 2 After 5 years of cortcosteroid therapy: the gadolinium T1-weighted axial MRI shows resolution of the ependymal enhancement.

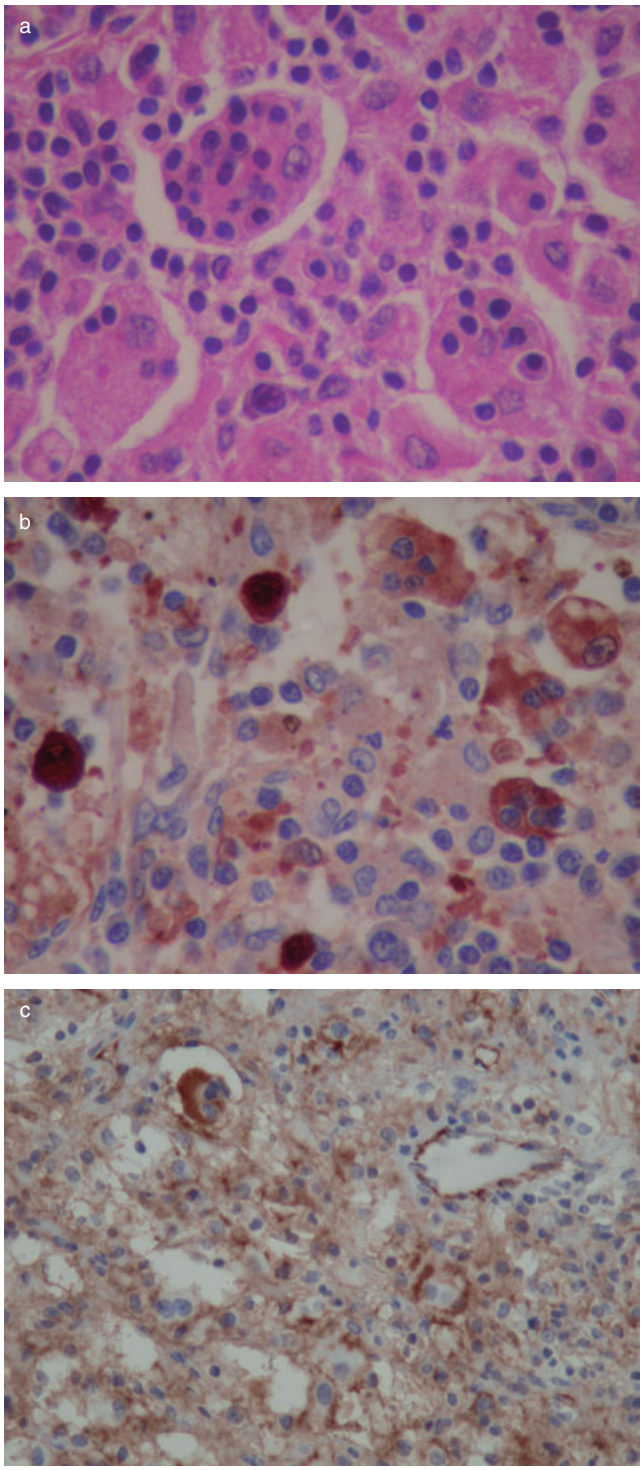

Fig. 3 (a) Histiocytes engulfing several small lymphocytes (emperipolesis). HE. (b) The large histiocytic cells are positive for S100. (c) Histiocytic cells with emperipolesis show a membranous and/or intracytoplasmic immunoreactivity for CD 31.

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