Rare case of extranodal Rosai-Dorfman disease presenting as a middle cranial fossa tumour

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DESCRIPTION

A 16-year-old previously healthy boy presented with a 7-year history of 'black out' spells. The patient reported that the episodes occurred one to two times in a month associated with dizziness and loss of consciousness without associated urinary or faecal incontinence. At times, he would have auditory hallucinations and brief lapses in memory. His neurological examination was normal. A CT of the head performed revealed a hyperdense right middle cranial fossa mass. MRI of the brain showed an enhancing, large dural-based mass in the right middle cranial fossa with an anterior dural tail and encasement of the cavernous and supraclinoid segments of the right internal carotid artery (figure 1). An MRI of the entire spine was normal. The patient underwent near-total resection, and frozen sections demonstrated dense fibrovascular connective tissue interspersed with clusters of small cells with high nuclear-to-cytoplasmic ratios with round or oval hyperchromatic nuclei as well as scattered lymphocytes and histiocytes (figure 2). Immunohistochemistry demonstrated negative epithelial membrane antigen. S100 and CD68 positive histiocytes were identified that were CD1a negative, and CD20 and CD3 positive lymphocytes were also present. Together, this constellation of findings

Figure 2 Histopathology reveals the following results: (A) H&E 40x: clusters of lymphocytes and histiocytes; (B) H&E 100x: whorls of dense, paucicellular connective tissue with occasional spindle cells surrounded by chronic inflammatory cell infiltrate; (C) 200x: histiocytes with clear cytoplasm and dense central nuclei; and (D) S100 immunohistochemistry reveals positive staining of histiocytes and rare emperipolesis (inset).

was most consistent with the diagnosis of Rosai-Dorfman disease.¹ The patient had good clinical recovery after surgery and remains disease free for 6 years following diagnosis.

Rosai-Dorfman disease is a non-Langerhans cell histiocytic condition more frequently seen in children and young adults that typically presents with fever and painless cervical lymphadenopathy with some association with hypergammaglobulinaemia.² Recent genomic studies have demonstrated mutations involving the mitogen-activated protein kinase pathway and various other intracellular transcriptional and cell cycle regulation mechanisms.¹ Isolated extranodal disease is rare, though central nervous system involvement including dural, parenchymal and spinal lesions has been reported and oftentimes mimics meningioma on

Figure 1 CT head shows a hyperdense, extra-axial mass in the right middle cranial fossa (A) with partial calcification (B). MRI brain T1 post-contrast study shows an enhancing mass with an anterior dural tail extending along the right petrous bone, encasing the cavernous and parasellar region (C) and causing mild mass effect on the lateral aspect of the right midbrain and right mesial temporal lobe (D).

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Learning points

- ▶ Rosai-Dorfman disease is a histiocytic condition that can present with isolated extranodal intracranial involvement mimicking meningioma on neuroimaging.
- ▶ Histological hallmarks are S100 and CD68 positive histiocytes that are CD1a negative and the presence of emperipolesis.
- ▶ Prognosis is generally good for isolated cases following surgical resection.

neuroimaging.^{3–5} The management of isolated extranodal Rosai-Dorfman disease is generally surgical resection and carries an overall good prognosis.²

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