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Neuropathology Education

Bilateral intraventricular mass in a child

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CLINICAL COURSE

An 8-year-old boy presented with a history of holocranial headache for 4 weeks which was mild to moderate in intensity. There was history of intermittent vomiting for the same duration. The patient was drowsy for the last 10 days. Non-contrast CT scan of head showed a space-occupying lesion in lateral ventricles with hydrocephalus. MRI showed a large lobulated well-defined heterogeneous mass lesion filling up both lateral ventricles at the posterior and temporal horns, bilaterally reaching up to the level of the foramen magnum. The mass lesion was hypointense on T2-weighted images (T2W) and isointense to grey mater on T1W images. On contrast images there was intense homogeneous enhancement seen at the lesion (Fig. 1). Right fronto-temporo-parietal craniotomy with decompression of the tumor was performed. Perioperatively, the tumor was yellowish in color, firm, not able to be aspirated and minimally vascular. Clinical diagnosis of choroid plexus tumor was considered.

Pathological findings

Microscopic examination revealed multiple fragments of a tumor. The tumor was composed of sheets of round to oval to spindle-shaped cells with finely granular eosinophilic cytoplasm (Fig. 2A). A significant number of cells contained foamy cytoplasm (Fig. 2B). Many interspersed xanthomatous Touton giant cells were present (Fig. 2C). Scattered lymphocytes, plasma cells and eosinophils were seen. No mitosis or necrosis was seen. Normal choroid plexus was identified at one place (Fig. 2D).

On immunohistochemistry, CD68 (Fig. 2E) and fascin (Fig. 2F) were diffusely positive in histiocytic cells, including xanthomatous cells. Scattered cells showed weak cytoplasmic reactivity for S-100. GFAP and CD1a were negative. Ki-67 labeling index was low.

DIAGNOSIS

Juvenile xanthogranuloma (JXG)

Serum cholesterol and lipid profiles of the patient were within normal limits. No skin lesions were present.

DISCUSSION

Involvement of CNS by JXG is rare.[1–4] It occurs mostly in skin with a predilection for the head and neck region.[5] An extensive literature search revealed less than 20 previously reported cases of JXG involving the brain.[1–4] JXGs of CNS are often associated with seizures, ataxia, increased intracranial pressure, developmental delay and other neurological deficits.[2] Because these lesions usually affect infants and children, these are termed as juvenile xanthogranulomas. It is classified under non-Langerhans cell histiocytosis which is derived from dendrocytes/dendritic cells.[6] Histopathologic examination shows dense infiltrates of histiocytes with variable numbers of multinucleated giant cells, including Touton giant cells. Admixed inflammatory cells comprising eosinophils, lymphocytes and plasma cells are often seen.[5]

Due to presence of eosinophils, there is diagnostic difficulty in differentiating JXG from Langerhans cell histiocytosis (LCH). It becomes important to differentiate JXG from LCH because they have clinically significant differences in prognosis. LCH often possess histiocytes with nuclear irregularity, folding and grooving. Immunohistochemically, cells of LCH are diffusely and strongly positive for S-100 and CD1a.[7]

Other differential diagnoses for a tumor in the pediatric age group and at the intracranial location are lipidized astrocytic tumor, Rosai Dorfman disease (RDD) / sinus

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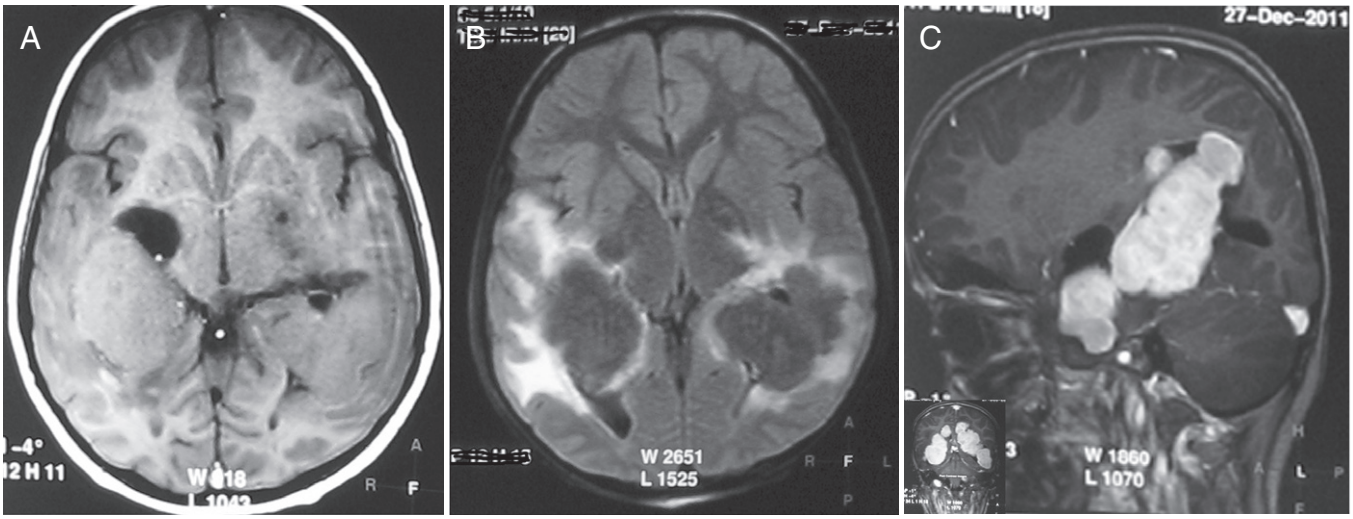

Fig. 1 MRI shows a large lobulated, well-defined, heterogeneous mass lesion filling up both lateral ventricles at posterior and temporal horns, bilaterally reaching up to the level of the foramen magnum (A, B). The mass lesion is hypointense on T2-weighted images (T2W) and isointense to grey mater on T1W images. On contrast images there is intense homogeneous enhancement seen at the lesion (C) (Inset left lower corner).

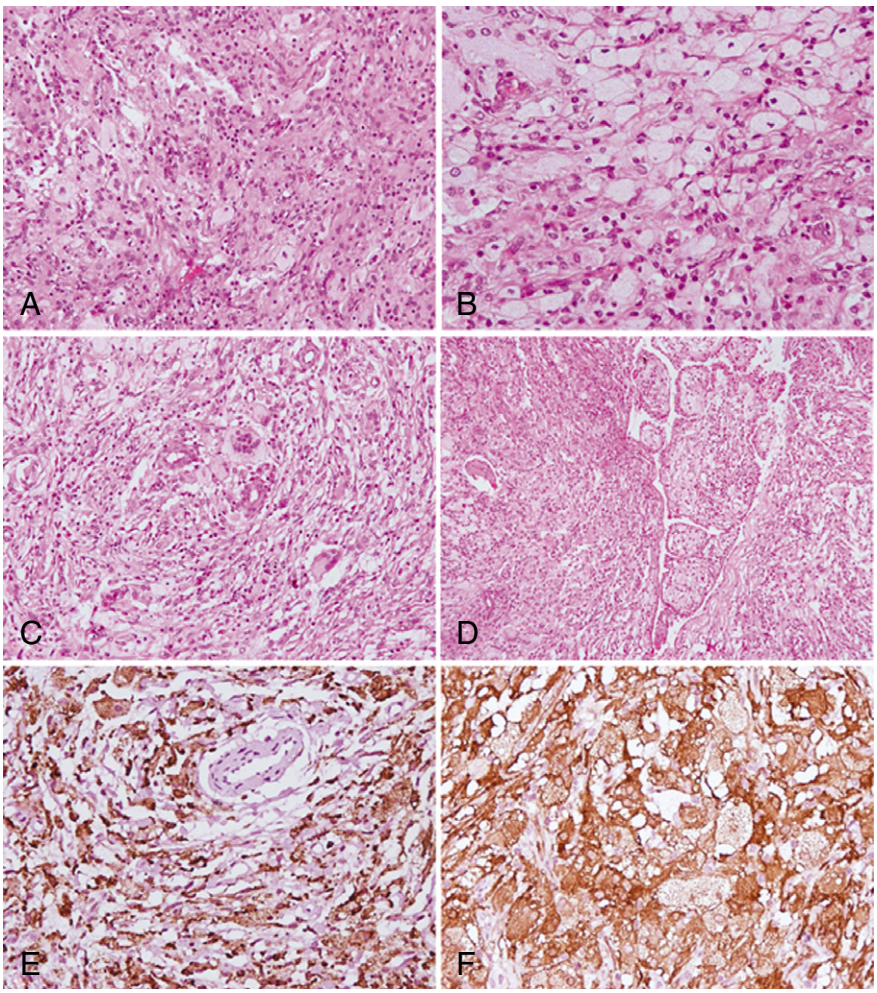

Fig. 2 A: Photomicrograph shows sheets of round to oval cells with abundant foamy to eosinophilic cytoplasm. HE ×100, B: Higher magnification to show large foamy histiocytes and interspersed inflammatory cells. HE ×400, C: Xanthomatous Touton giant cells are admixed. HE ×400, D: Normal choroid plexus is infiltrated by histiocytes and inflammatory cells. HE ×100, E and F: CD68 and fascin are strongly positive in histiocytic cells, respectively.

histiocytosis with massive lymphadenopathy, hemophagocytic histiocytosis and xanthomas.

Astrocytic tumor shows gliofibrillary background at least at some places. In addition, absence of GFAP immunostaining and expression of histiocytic markers help in ruling out the possibility of gliomas. Pleomorphic xanthoastrocytoma (PXA) occurs in children and usually consists of lipidized foamy cells with fascicular and storiform arrangement of neoplastic cells. But as the name suggests, PXA shows pleomorphic astrocytes with bizarre cytologic features. The presence of eosinophilic granular bodies in PXA is an additional important diagnostic feature. Extranodal RDD rarely occurs in the brain. Histologically, it is characterized by large histiocytes, which often show viable hematopoietic cells (mostly lymphocytes) within the cytoplasm, a phenomenon known as emperipolesis. On immunohistochemical examination, cases of RDD are positive for S-100 protein.

Congenital or acquired hyperlipidemic disorders may result in the development of intracranial xanthomatous masses. There was no systemic abnormality of lipid metabolism in our case.

A common form of non-neoplastic xanthomatous brain lesions is xanthogranuloma of the choroid plexus. These lesions are usually found incidentally at autopsy. Morphologically, choroid plexus xanthogranulomas are characterized by cholesterol clefts and a densely vascularized granulomatous tissue. Although our patient had tumors in both lateral ventricles, the histological appearance clearly differed from that of choroid plexus xanthogranuloma.

The histogenesis of primary intracranial JXG is unclear, but an origin from macrophages/histiocytes located in the meninges and the choroid plexus seems most plausible.[2]

The prognosis of patients with cutaneous JXG is excellent, with the majority of lesions showing spontaneous regression.[5] Based on the few case reports of intraventricular and dura-based JXGs, surgery seems to be the first line of treatment in symptomatic patients.[1–4] In the current case, surgery was performed for diagnostic purposes and for decompression in order to reduce intracranial pressure. The patient is currently under follow-up and free from symptoms.

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