An unusual posterior fossa tumour in a young child

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DESCRIPTION

A 23-month-old previously healthy boy presented to our hospital with a 10-day history of progressive early morning vomiting and sleepiness. Neurological examination revealed an encephalopathic child with impaired upgaze without papilloedema or other focal neurological abnormalities. Emergency MRI demonstrated a large homogeneously enhancing tumour (figure 1A,B) that was T2 isointense to grey matter (figure 1C), without reduced diffusivity on apparent diffusion coefficient image sequences (figure 1D). Gross total excision of the tumour was obtained (figure 1E) and the pathology demonstrated papillary fronds of columnar cells consistent with a diagnosis of choroid plexus papilloma of the fourth ventricle (figure 2). Postoperatively the patient developed fixed leftward gaze preference and worsening hydrocephalus requiring placement of a ventriculoperitoneal shunt.

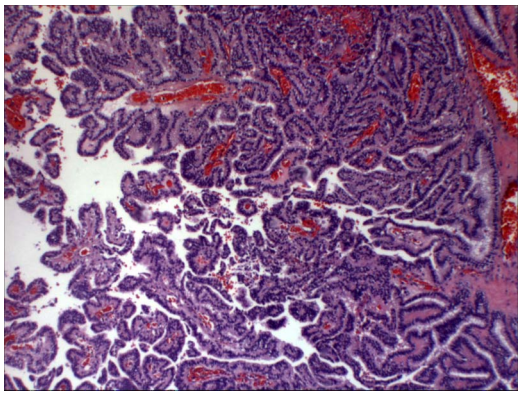

Figure 2 H&E stained surgical specimen showing papillary fronds with uniform columnar cells lining fibrovascular cores consistent with a diagnosis of choroid plexus papilloma. The cells have oval nuclei with minimal pleomorphism and only rare mitoses without evidence of cytologic atypia or necrosis (40× magnification).

DISCUSSION

Choroid plexus papilloma represents 2–6% of all childhood brain tumours and up to 20% of tumours in children under 1 year of age.[1] Usually arising from the lateral ventricles in childhood and the fourth ventricle in adulthood, choroid plexus papilloma can be curative following complete neurosurgical correction.[2] The homogeneous contrast-enhancing features and lack of reduced diffusivity on MRI help distinguish choroid plexus papilloma from more malignant posterior fossa tumours of childhood. Our case highlights the MRI

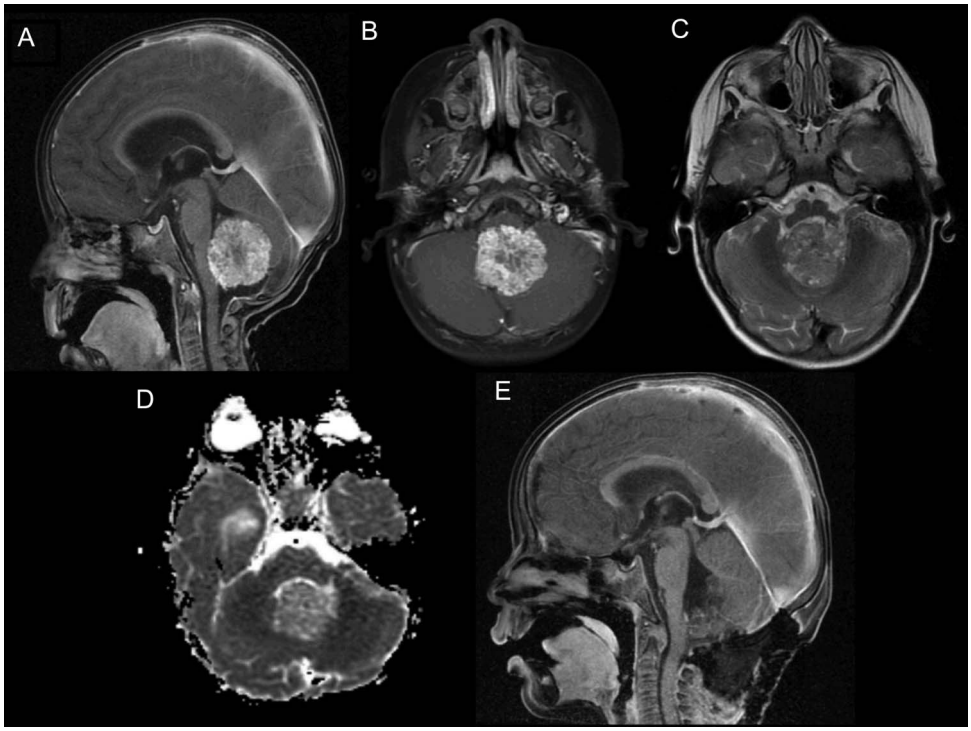

Figure 1 (A, B) Post-gadolinium MRI sequences showing a homogeneous fourth ventricular neoplasm with associated obstructive hydrocephalus. The mass is T2 hypointense compared with grey matter (C) and does not demonstrate reduced diffusivity on apparent diffusion coefficient sequences (D). (E) Postoperative MRI is consistent with a gross total resection.

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features of choroid plexus papilloma of the fourth ventricle and is important to consider in the differential diagnosis of posterior fossa tumours in young children.

Learning points

- Choroid plexus papilloma is a rare tumour of childhood that may arise anywhere within the ventricular system and cause obstructive hydrocephalus.
- Choroid plexus papilloma of the fourth ventricle is rare in childhood and may radiographically mimic more common homogeneously contrast-enhancing posterior fossa brain tumours.

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