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Pilomyxoid astrocytoma of the fornix imitating a colloid cyst

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ABSTRACT

Pilomyxoid astrocytoma is a recently described pediatric brain tumour with distinct histological features and has been shown to behave more aggressively than pilocytic astrocytoma. Pilomyxoid astrocytomas usually involve the hypothalamic/chiasmatic region with imaging features similar to pilocytic astrocytoma. We report a case of pilomyxoid astrocytoma in the fornix with imaging features imitative of colloid cyst.

1. Introduction

Pilomyxoid astrocytoma (PMA) is a recently described pediatric brain tumour. PMAs were previously classified within the pilocytic astrocytoma (PA) category. However, PMA has distinct histological features and has been shown to behave more aggressively than PA. Majority of previously reported PMAs involved the hypothalamic/chiasmatic region with similar imaging features of PA. We describe a case of PMA in the fornix with an atypically imaging features imitative of colloid cyst.

2. Case report

A 6-year-old girl presented with progressive headache and vomiting of 1 week with 2 episodes of generalized tonic clonic seizures. She was brought to the Emergency Department in a confused state with Glasgow Coma Scale of 13 (E4M5V4). Magnetic resonance imaging (MRI) of the brain revealed a homogenous, well circumscribed, oval mass in the anterior aspect of the third ventricle. This mass was isointense on T1-weighted, hyperintense on T2-weighted and FLAIR images. There was no enhancement with intravenous Gd-DTPA. There was acute obstructive hydrocephalus with obstruction of the foramina of Monro bilaterally. The third and fourth ventricles were not dilated (Figs. 1 and 2). Computed Tomography (CT) scan showed a slightly hyperdense lesion (Fig. 3). Initial diagnosis of colloid cyst was made based on the clinical presentation and imaging findings. The patient underwent a stereotactic endoscopic procedure with the purpose to decompress and excise the cyst as well as to relieve the hydrocephalus. Intraoperatively, the lesion was found to be solid and lobulated, occupying the anterior part of the third ventricle measuring about 1.5 cm × 2.5 cm. Biopsy was performed along with septum pellucidotomy and placement of extra-ventricular drain to relieve the hydrocephalus. Patient subsequently underwent craniotomy for excision of the tumour, where a gel-like tumour arising from the fornix was excised. A residual rim of tumour was left behind.

Histopathological examination of the specimen showed piloid astrocytes disposed in a fibrillary to myxoid background. The tumour cells were uniform with round to oval nuclei and hair-like cytoplasmic processes. There were no Rosenthal fibers, eosinophilic granular bodies, mitosis or endothelial proliferation (Fig. 4). The diagnosis of pilomyxoid astrocytoma WHO Grade 1 was made.

Post-operative recovery was complicated by transient diabetes insipidus with hypocortisolism and bilateral subdural collections requiring surgical drainage. There were also neurological complications where the patient developed generalized spasticity and cognitive impairment. Patient was discharged 2 months later. A follow-up MRI was scheduled about 4 months after discharge to assess the residual tumour and plan further management, but unfortunately the patient defaulted follow-up.

3. Discussion

Pilomyxoid astrocytoma (PMA) was introduced as a clinicopathological entity by Tihan et al. in 1999 [1]. Similar to pilocytic astrocytoma (PA), PMA may occur anywhere along the neuraxis and

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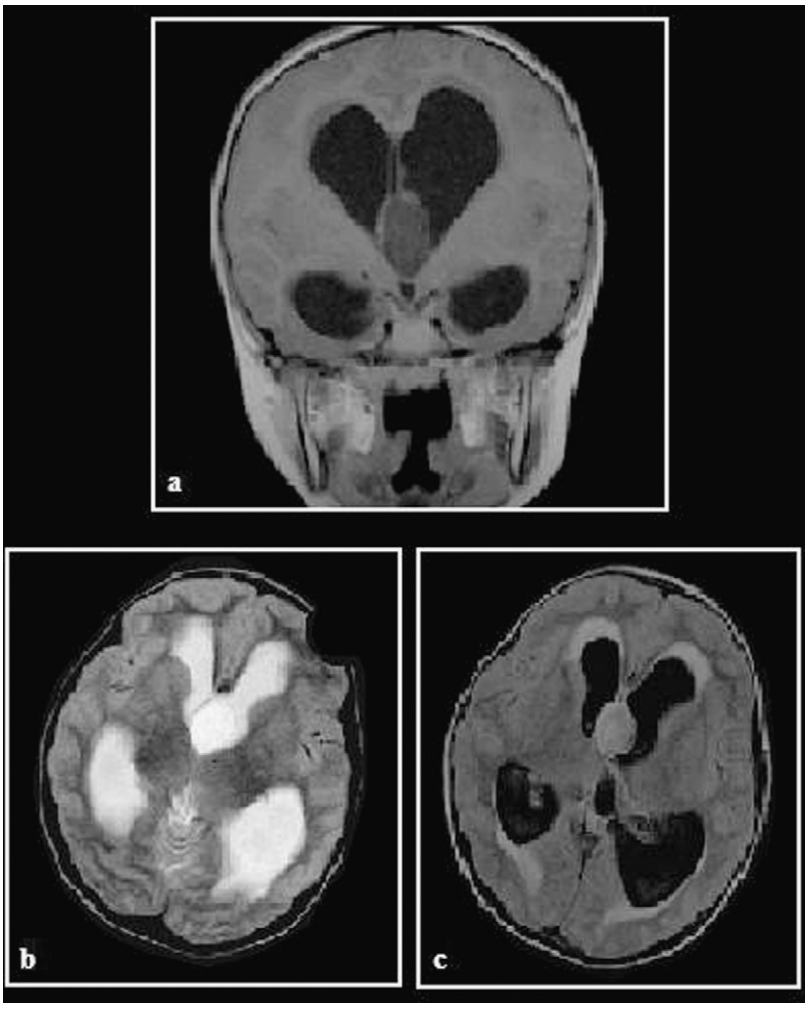

Fig. 1. MRI of the brain (a) T1-weighted in coronal view, (b) T2-weighted in axial view and (c) FLAIR in axial view. There is a well defined, oval mass anterior to the third ventricle. It is homogenously isointense on T1-weighted and hyperintense on T2-weighted as well as FLAIR sequences. There is associated acute supratentorial obstructive hydrocephalus.

afflict individuals throughout childhood [2]. PMA, however, exhibits predilection for the hypothalamic/chiasmatic region and tends to affect infants and younger children [1,3]. There also has been a report of spinal cord tumours in older children with histological features identical to those of PMA [4].

The clinical presentations of PMA parallel those of other pediatric brain tumours, and include failure to thrive, developmental delay, altered consciousness, vomiting, feeding difficulties and generalized weakness [1]. Gait abnormalities, dysmetria and nystagmus may be present in posterior fossa tumours. Neck stiffness

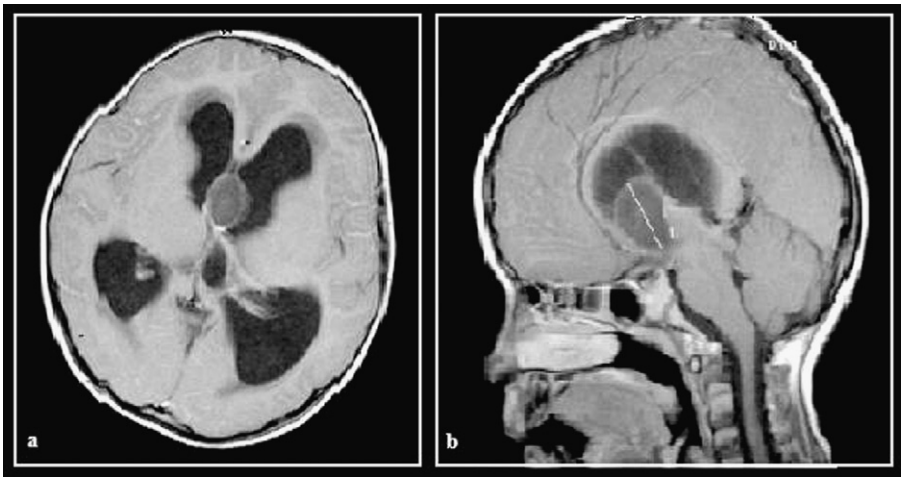

Fig. 2. Gadolinium-enhanced spin-echo sequence MRI of the brain in (a) axial and (b) sagittal sections show no significant enhancement of the mass.