Pilomyxoid astrocytoma of the fornix imitating a colloid cyst

Nur Adura Yaakupa, Kartini Rahmata, Norlisah Mohd Ramli, N. Vairavan, Vicknes Waran, K.T. Wong

Department of Biomedical Imaging, Faculty of Medicine, University of Malaya, 50603 Kuala Lumpur, Malaysia
Department of Neurosurgery, University of Malaya, 50603 Kuala Lumpur, Malaysia
Department of Pathology, University of Malaya, 50603 Kuala Lumpur, Malaysia

Pilomyxoid astrocytoma (PMA) 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...
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 paediatric 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.
Histologically, PMA composed of highly monomorphous and piloid cells in a markedly myxoid background. Tumour cells are often arranged around vessels, in an angiocentric pattern that remotely resembles the perivascular rosettes seen in ependymomas. There is lack of Rosenthal fibers and only rarely are eosinophilic granular bodies seen, both of which are characteristics of PA. Mitotic figures are rare [1–5].

The imaging features of PMA appears similar to PA and quite unlike colloid cyst. Initial review of 13 PMA MR images by Burger and co-workers [3] reveals all the tumours to be well circumscribed without evidence of peritumoural oedema or parenchymal infiltration. Although PMA may occur throughout the neuraxis, most of these tumours (76.9%) are located in the hypothalamic/chiasmatic and/or originate from the midline. A majority of them (84.6%) are solid, with the remainder showing minimal cystic component. PMA occasionally causes obstructive hydrocephalus (38.5%) and only rarely shows imaging evidence of central necrosis (7.7%). On T1-weighted MRI, 92.3% of the tumours are isointense, with the others being slightly hypointense. With contrast administration, PMA tends to enhance homogenously [2]. There are also observations of substantial cerebrospinal fluid (CSF) dissemination in PMA [3]. On proton magnetic resonance spectroscopy imaging (MRSI), it has been shown that PMA demonstrated decreased concentrations of total choline (Cho), creatine (Cr) and N-acetyl aspartate (NAA). In contrast, proton MR spectra of PA showed elevated Cho and decreased Cr and NAA signals [5]. Therefore, MR spectroscopy may prove useful in distinguishing PMAs from PAs, by detecting the low metabolite concentrations in PMAs.

The lesion in our patient, however, demonstrated an unusual MR appearance. Although the tumour returned the expected signals of PMA on the spin-echo and inversion recovery sequences, it did not show significant enhancement. Combined with its well circumscribed, oval morphology and location, it was initially diagnosed as a colloid cyst and treatment strategy concordant with the working diagnosis.

Colloid cysts are benign, rare intracranial lesions that account for 0.5–1.0% of brain tumours, commonly occurring in the third to fifth decades of life [6]. They are almost always located in the anterior aspect of the third ventricle. They typically present with progressive headache secondary to obstructive hydrocephalus [7]. They are also a recognized cause of sudden death [6]. Radiologically, these lesions show variable appearances. Most colloid cysts are oval or rounded. On CT scan, most are slightly hyperdense compared to the brain, but some may be hypo- or isodense. They may show thin rim enhancement after administration of iodinated contrast media. On MRI, they may occasionally show intracystic fluid levels or central and peripheral components in the lesion. About 50% of colloid cysts are hyperintense on T1-weighted images and the rest are either hypo- or hypointense with respect to the brain. On T2-weighted images, most colloid cysts are hypointense to the brain. Cysts that are hypointense on T2-weighted sequences may be difficult to visualise using fluid-attenuated inversion recovery images (FLAIR). The variability of the MR signal pattern is dependent on the cholesterol and protein content of the cyst [6].

Limited clinical experience makes it difficult to generate conclusive prognostic data regarding PMA. A preliminary investigation on the long-term clinical outcomes of 21 patients with hypothalamic PMAs had shown a significant local recurrence rate of 76% and mortality rate of 33%. There is also substantial CSF dissemination (14%) seen in PMA [3]. In a study by Horn et al. [7] on the treatment options of colloid cyst, the rate of recurrence/residual tumour depends on the type of surgical approach. A higher rate of residual tumour is seen in patients underwent endoscopic resection compared to open craniotomy (47% vs. 6%), albeit no recurrence was seen on intermediate follow-up. In another study of a cohort of 78 patients harbouring symptomatic colloid cysts, 34% presented with acute deterioration with an overall mortality rate of 12% [8]. Given the significant rate of CSF dissemination in PMA, it is important that imaging of the spine is performed to complete the assessment of this tumour.

4. Conclusion

This report describes a pilomyxoid astrocytoma at a site different from previously reported and also with unusual radiological appearance which imitates a colloid cyst. This case highlights that the differential diagnosis for colloid cyst should include PMA. This is important as PMA demonstrates an aggressive behaviour with propensity for CSF dissemination. MRI of the spine should be done once diagnosis is obtained.

Conflict of interest

None declared.
References


