Case report

Supratentorial Extra-axial Ependymoma: A Rare Radiological Mimic of Cystic Meningioma

Fattah Rahiman Ghazali[1], Ahmad Hadif Zaidin Samsudin[1]

Abstract

Ependymoma is commonly an intra-axial infratentorial tumour affecting paediatric age group. We report a rare case of supratentorial extra-axial ependymoma in a teenager who presented with worsening headache and mild cognitive impairment. Previously, only few of such cases have been reported in the literature. However, in the last decade, there has been increasing number of this rare entity represented as a solid-cystic mass. In this report, we discuss the radiological features as we think it is mimicking a cystic meningioma and recommend that ependymoma should be considered in the diagnosis of a solid-cystic extra-axial lesion especially when located supratentorially.

Keywords: Ependymoma, supratentorial, extra-axial, solid-cystic, cystic meningioma.

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Introduction

Intracranial tumours are classically divided into intra-axial and extra-axial masses. Differential diagnosis of extra-axial masses depends on the involved extraparenchymal tissue, with approximately 35% of all intracranial neoplasms are benign meningioma[1]. Cystic meningioma is a variant of meningioma with intratumoral cystic degeneration or extratumoral arachnoid cyst which can be centrally or peripherally located, outside and adjacent to tumoral edge and inside the brain parenchyma[2].

Definitive diagnosis of extra-axial brain tumours is usually trivial especially in cases of common pathology such as meningioma. Rare and challenging extra-axial tumours such asneuroenteric cyst, primary leptomeningeal melanomatosis, isolated dural neurosarcoidosis, intradiploic epidermoid cyst and cavernous haemangioma of skull have been reported[3].

Ependymoma has been reported as a rare diagnosis of extra-axial tumour, especially when located supratentorially, as it is usually an intra-axial tumour commonly located at posterior fossa. Common in paediatric age group, it can present at any age depending on the location.To the best of our knowledge, to date, only few cases of supratentorial extra-axial ependymoma had been previously reported in the literature[4-8]. In our case, the radiological features of the tumour were mimicking cystic meningioma and the diagnosis of ependymoma has not been considered early due to the peculiarity.

Case Report

Our patient is a 17-year-old lady, presented with headache for 4 months duration. It was centrally located and sharp in nature with pain score of 8 over 10. It became worsening and increasing in intensity and frequency. The headache was associated with vomiting and blurring of vision. She also became slow and had difficulty in finding words. Otherwise, she had no forgetfulness, and her activity of daily living (ADL) was independent. There was no history of trivial trauma and no symptoms of infection.

At presentation, she was orientated to self, time,

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place, and person. Her speech was spontaneous however reduced in output and was impaired in fluency.Her attention and memory were intact however she had impaired calculation and abstract. Cranial nerve examination revealed bilateral abducens nerve (VI) palsy and right facial nerve (CN VII) upper motor neuron palsy. Her visual acuity was 6/9 bilaterally. She had right superior visual field defect. Otherwise no relative afferent pupillary defect. The rest of cranial nerves were intact. There was positive right pronator drift. Upper and lower limbs examination revealed normal power with intact sensation. However, there was increased tone on the right side with upgoing plantar with brisk right upper limb reflex. Cerebellar signs were negative.

Non-contrasted computed tomography (NCCT) of brain revealed a large left frontal solid mass with multiple intralesional small foci of hypodensity and perilesional hypodensity likely represent cystic components. In contrast-enhanced computed tomography (CECT), it was heterogeneously enhanced and there are multiple surrounding and traversing cortical vessels. It caused significant mass effect and midline shift to the right with subfalcine and uncal herniation. There was no tonsillar herniation or perilesional oedema, hydrocephalusor broaddural based seen (Figure 1). No skull lesion, erosion or hyperostosis was observed.

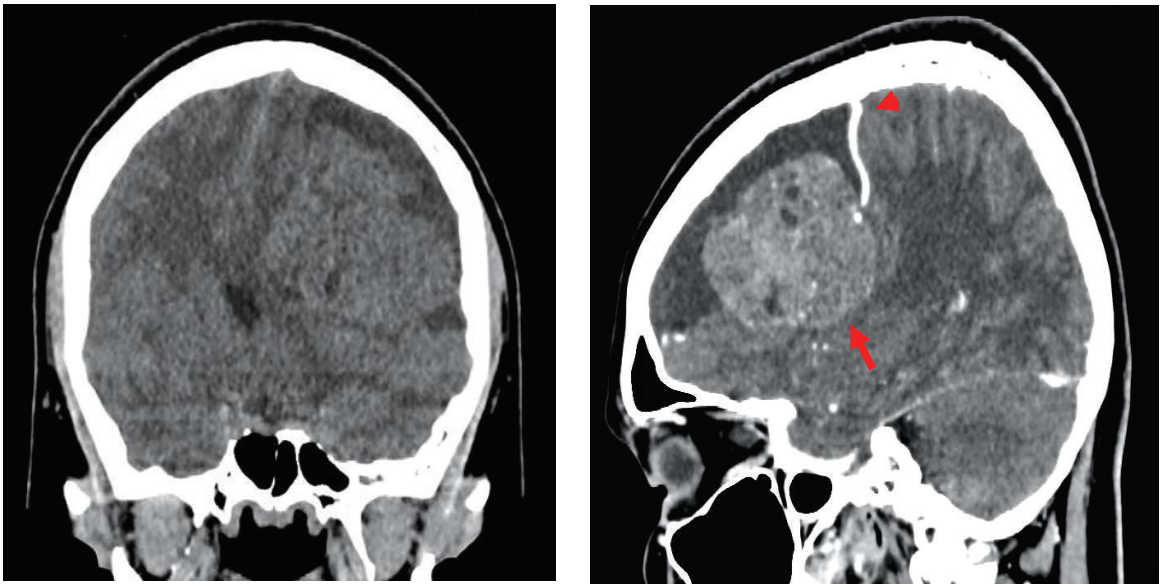

Figure 1.CT: from left: (A) non-contrasted, (B) contrast-enhanced; showing a solid-cystic mass at left frontal region with significant mass effect causing midline shift and subfalcine herniation to the right (1A). No perilesional oedema. Intralesional cystic foci, and multiple surrounding and traversing vessels were noted (1B: arrow and arrowhead, respectively).

A huge broad-based solid-cystic extra-axial mass at left fronto-parietal region seen on magnetic resonance imaging (MRI) which was done subsequently. The solid component showed heterogenous signal intensity and enhancement, restricted diffusion pattern and multiple foci of blooming artefact that represent microhaemorrhages and calcifications. Multiple enhanced septations seen within the laterally located cystic component (Figure 2). No white matter oedema. There was mass effect onto body of corpus callosum, left cerebral peduncle, left lateral and third ventricles. Fourth ventricle is preserved. Presence of midline shift to the right causing subfalcine and uncal herniation. No tonsillar herniation. Pons, medulla oblongata and cerebellum are unremarkable. On MRA, multiple vessels seen surrounding and traversing the mass. No abnormality of the circle of Willis. MRS shows reverse choline: ..creatine ratio, elevated myoinositol peak and low NAA peak. Differential diagnosis of cystic meningioma was made.

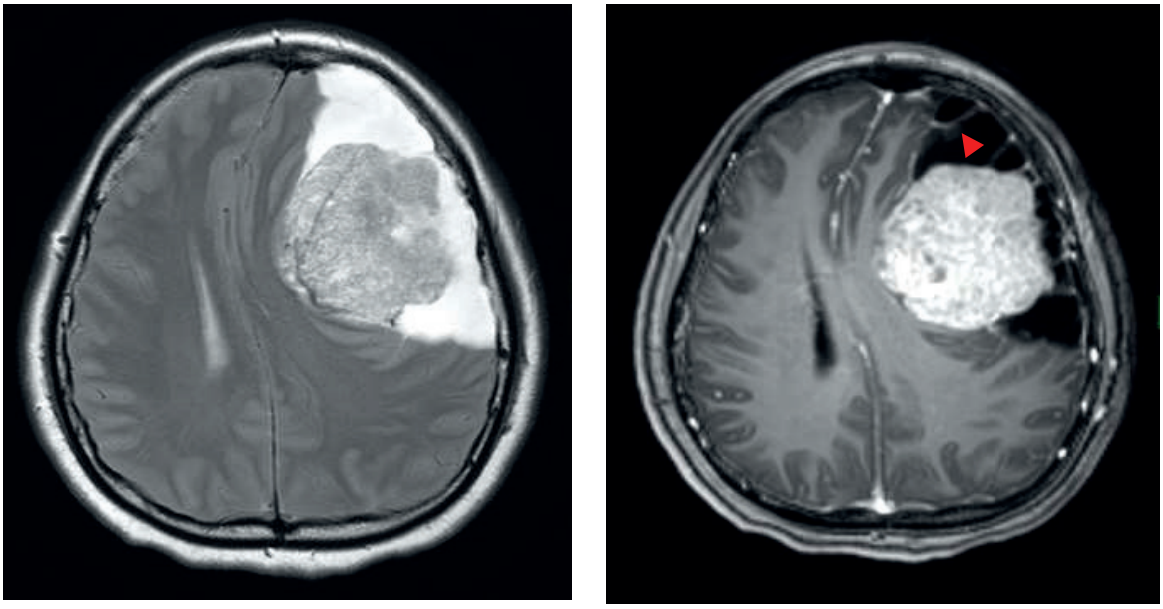

Figure 2.MRI: from left: (A) T2-weighted image, (B) post Gadolinium; showing a large solid-cystic mass involving left fronto-parietal region with significant mass effect. No perilesional white matter oedema or hydrocephalus (2A). Heterogenous enhancement of the solid component and multiple enhancing septae of the cystic component (2B: arrowhead) were noted.

The patient was referred to neurosurgery for surgical treatment. Intra-operatively, there was straw colour fluid overlying the tumour. The tumour was seen below the dura with brain and tumour border was well identified. It was a soft, suckable and pale coloured mass which was removed circumferentially. Medial part of the tumour appeared invading the brain tissue and unable to visualize proper tumour border. No bleeding was observed.

Malignant cells seen within the fluid that was sent for cytology. Histopathological examination of the tumour was consistent with ependymoma, WHO Grade II, in view of lower cell density, lower mitotic count with no microvascular proliferation or palisading necrosis identified. Post-operatively, patient made full recovery and all of her symptoms had resolved. Follow-up imaging was planned for her to look for any residual and craniospinal assessment for further treatment planning.

Discussion

Ependymoma, a glial tumour with ependymal cell differentiation, usually arises from intracerebral ventricular lining and central canal of spinal cord. It is the third most common brain tumour of neuroepithelial origin in paediatric; most frequently involve posterior fossa, followed by supratentorial and spinal cord, respectively. Traditionally considered as a single disease entity, recently it is shown to be biologically distinct entity that have different cells of origin[9]. Age of presentation varies, depending on the location of the tumour.

First known to be reported by Hanchey et al in 1976, supratentorial extra-axial ependymoma, while still being rare, is seen increasingly reported within the past decade and seems to be mimicking a meningioma[4-7]. It is thought to be arising from heterotopic fetal rests of ependymal cells[4-8].There are two molecular subgroups of supratentorial ependymoma,characterized by the fusion of transcription factor on chromosome 11 - *RELA* and *YAP1*; the later usually occurs in infancy and has a better prognosis[9].

Most of the reported cases of supratentorial ependymoma affect older children and young adult, ranging from 13 to 43 years old[4-8]. Presenting symptoms vary, depending on the location, with seizure as one of the common presentation. Our patient is a 17-year-old girl who was previously well until 4 months prior to her presentation, experienced headache and was noted to have mild cognitive impairment, owing to the location of the tumour at left fonto-parietal region.

MRI signal intensity of supratentorial ependymoma are generally heterogenous T1 hypointense and T2 hyperintense of the solid component which is usually avidly enhanced even though few lesions can have little or no enhancement[9]. Supratentorial ependymoma has tendency for extensive cyst formation due to delay clinical presentation with usually large size of the tumour at presentation. Calcifications are common findings while foci of haemorrhages may be present[9,10].

Many authors reported that their supratentorial ependymomas were meningioma-mimicker[4-7]. It is similar in our case, as the mass was a broad dural-based mass with large peripheral cystic component, mimicking a cystic meningioma. However, it was confirmed as ependymoma by the histopathological examination.In cystic meningioma, the peripherally located cystic component is thought to be due to the peripheral degeneration, arachnoid cyst, fluid secretion by tumour cells, internal haemorrhage absorption and scar tissue's loculated cerebrospinal fluid (CSF) within or adjacent to the meningioma[2].Most of the tumours were surgically resected[4-6]. Few cases reported no tumour residual or recurrence in subsequent follow-up[5]. Adjuvant chemotherapy or radiotherapy or both may be given to the patient despite of controversial and lack of data[5-8]. Our patient had resolution of her symptoms after the operation. She was given subsequent follow-up imaging to look for residual and spinal surveillance. Adjuvant therapy will be considered depending on the radiological findings.

Conclusion

An increasing number of reported cases of supratentorial extra-axial ependymoma within the past decade indicates the rising incidence of this rare entity. The radiological features that usually mimicking the meningioma, particularly cystic meningioma in our case, should raise suspicion of this entity. Therefore, we recommend that it must be considered as one of the differential diagnoses for supratentorial extra-axial solid-cystic lesion especially in older children and young adult.

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