CASE REPORT

Immature Brain Teratoma of An Infant In A University Hospital: A Case Report

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ABSTRACT

Teratomatous tumours of the head are rather uncommon. We report a 3-month-old child who presented with acute signs of raised intracranial pressure, needing craniectomy and subtotal tumour removal. The patient was diagnosed as intracranial immature teratoma grade 3, from the pathological study and elevated alpha-fetoprotein (AFP). Managing brain teratoma posed a challenge to the managing team due to the location of the tumour, the unavailability of standardized chemotherapy protocol and the dilemma of commencing adjuvant chemotherapy in a very young infant. The tumour was partially removed due to its critical location. And chemotherapy was delayed until the patient achieved 7 months of age. After four rounds of chemotherapy, the patient remained in remission for more than three years follow up.
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INTRODUCTION

Teratomas are germ cell tumours which contain of one or more of the embryonic germ layers, as well as tissues foreign to the site of origin. Complete surgical resection is the mode of therapy for patients with pure mature teratoma. However, due to the higher risk of relapse in children with teratoma, adjuvant chemotherapy would be required. The main risk factors for disease relapse are incomplete tumour resection and the immaturity of the tumour. The head is an uncommon site for teratomas. Mature teratomas are usually seen in younger children. The total excision of the tumour depends on the proximity of the lesion to the vital structures, especially in the illustrated case.

CASE REPORT

We report a case of an infant who presented to Hospital Universiti Sains Malaysia at 3 months of life with 4-days history of convergent squint, unresponsiveness to visual stimulation, irritability and weak cry. There was no history of vomiting or seizure. Upon examination, there was a neck deviation to the left side, bulging anterior fontanelle, and the presence of convergent squint. Fundoscopy examination revealed bilateral papilloedema. Patient's weight was 6.1kg (50th centile) and occipitofrontal circumference was 44cm (more than 97th centile). There was no facial asymmetry, and neurological examination of all limbs were intact. Magnetic resonance imaging (MRI) of the brain showed posterior fossa mass with acute hydrocephalus (Figure 1). An emergency right ventriculoperitoneal shunt was then performed, followed by craniectomy with subtotal removal of the tumour. Approximately, 80% of the tumour mass was surgically removed. The incomplete resection of the tumour was due to its proximity to the brain stem. Intraoperatively, the tumour appeared to be highly vascularized with mixed consistency, from soft to firm in nature. The mass consisted of solid and cystic components and was well encapsulated, except that the area was adjacent to the brain stem. The biopsy result showed an intracranial immature teratoma (grade 3) which was derived from all three germinal layers (Figure 2a and 2b). Numerous nodules of fetal cartilage, woven bone tissues and glands lined by respiratory epithelium and intestinal epithelium were seen. Areas of glial tissue, nerve smooth muscle, melanocytes, fat cells, squamous cell epithelium, and transitional epithelium were also seen. Choroid plexus-like tissue resembling the cerebellum was also present. Numerous immature

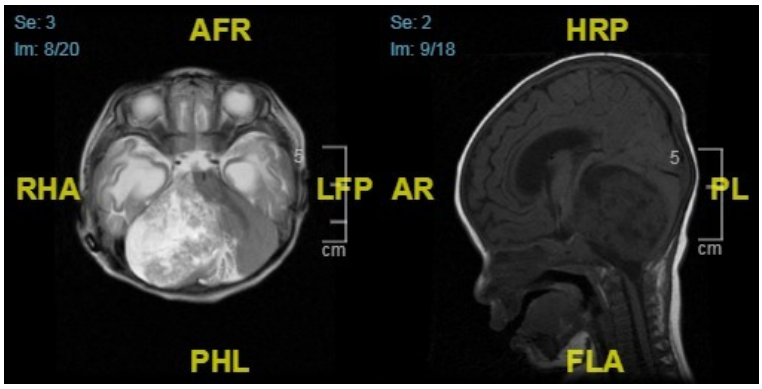

Figure 1: Large intra-axial posterior fossa tumour seen on initial MRI imaging with obstructive hydrocephalus

neuroepithelial structures and primitive cells with round small nuclei and rosette formation were identified.

There was a dilemma whether to treat this patient aggressively or conservatively due to early age at the time of presentation. He was well post-operatively. Initial AFP was 147.6ng/ml (normal for 3 months of age < 88ng/ml). Post operative AFP showed a decrease trend (from 46.1 to 15.4ng/ml). In view of the immaturity of the tumour and the inability to remove the tumour completely, he was commenced on ICE protocol. This chemotherapy regime comprised of ifosfamide, carboplatin, and etoposide. Patient completed 4 cycles of chemotherapy at 3 weekly intervals and showed a good recovery. CT scan post chemotherapy showed complete resolution of the tumour. However, due to logistic reason, patient defaulted follow-up. He turned up at 4 years of age with normal development milestone. Neurological examination revealed a tendency for mild wide base gait more on the left side. His head circumference was 51.5cm (at 75th centile). Eye examination showed bilateral mild alternating exotropia with good vision. CT scan confirmed that patient has remained in remission. His serum AFP levels were normal (less than 8.5ng/ml). He was also seen by a psychologist who found that he is now functioning at an average level of cognitive abilities, consistent to his chronological age.

DISCUSSION

The immature teratoma is a rare type of tumour worldwide, especially when it was found in the brain. Of all teratoma, about 3% occur in the head and neck region (1). Teratoma is a germ cell tumor arises from ectopic pluripotent stem cells and contained one or three embryological layers, and commonly occurred in the first year of life (2). For immature teratoma, the tumour possessed anaplastic immature features and considered a rare form of teratoma. Our patient showed signs of increased intracranial pressure such as irritable, squint, decrease visual response, left torticollis, and papilloedema. With the short presenting history, it is most likely that he has a congenital intracranial teratoma. Congenital brain tumours are tumours that are present within the 60 days of life (3). Hydrocephalus, polyhydramnios, and macrocephaly are common complications of the congenital brain tumours. We have negative maternal antenatal history for this case. His neurological examination was normal and in the majority of the cases, the neurological signs were

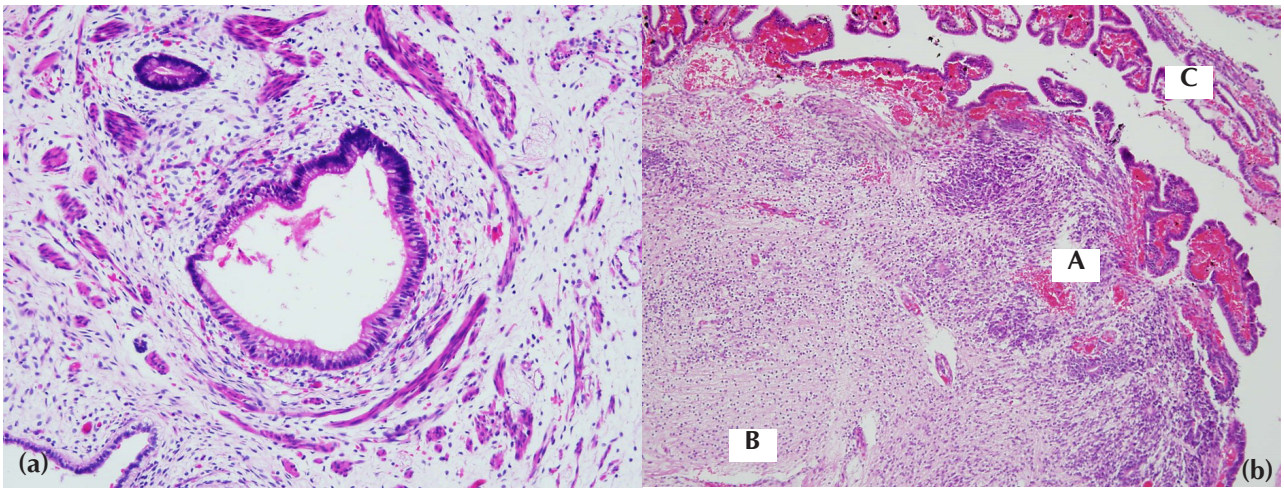

Figure 2: (a) Photomicrograph shows colonic tissue in an immature teratoma composed of colonic mucosa and its underdeveloped submucosa and muscular layer from the patient. (b) Photomicrograph shows the immature elements of this teratoma consist of neuroepithelial tissues (A). Glial tissue with many oligodendrogliocytes (B) and choroid plexus-like tissue (C) is also seen.

absent despite the presence of macrocephaly and hydrocephalus.

The high levels of alpha-fetoprotein (AFP) are often associated with endodermal sinus tumours or a mixed germ cell tumours with endodermal sinus tumour component. AFP is produced mainly by the yolk sac, liver, and gastrointestinal tract (GIT). Immature teratoma may contain primitive cells from these three organs. Intracranial immature teratoma with elevated AFP levels are uncommon (4). The high levels of AFP in our patient is possibly from the component of primitive GIT tissue. There was no yolk sac or hepatoid cells presence in this patient. In this case, AFP level showed a downward trend following operation. This is also expected as the child grow older. His AFP started to normalize following the completion of chemotherapy.

Imaging, like computerized tomography (CT scan) and MRI could be used to define the tumour extension and its relationship with the adjacent structures. Although MRI provides a clearer view of the tumour's size, particularly in the posterior fossa, CT is superior to MRI in detecting brain calcification. In at least two-thirds of the cases recorded, intracranial teratoma occurred supratentorially, with infratentorially occurring less frequently (5). On CT imaging, an isodense or low-density mass mixed with cystic components may be seen.

For patients with pure mature teratoma, complete surgical excision is an acceptable treatment option. To lower the likelihood of relapse, it is hypothesised that immature teratoma would require adjuvant chemotherapy. Chemotherapy has previously been suggested for cerebral immature teratomas with elevated AFP levels. Decision for chemotherapy and surgical resection would depend on the expected prognosis of the patient – related to his age, site of the tumour, completeness of the tumour removal, and the degree of immaturity of the tumour.

We postponed chemotherapy following incomplete surgical resection to prevent the anticipated adverse outcome to the patient. Complete resection of the tumour is only possible if the lesion is well-defined or capsulated. Recurrence in a completely excised mature teratoma is low, whereas incidence of recurrence in immature teratoma is high.

CONCLUSION

The management of complex and challenging immature teratoma in the infratentorial region, would require a multidisciplinary effort in ensuring that appropriate decision is made, which include surgery and chemotherapy. This decision will not only bring forth longer survival but also minimizing the potential sequalae from the intervention performed.

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