Case Report

Midline Posterior Fossa Mature Teratoma in a Child

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ABSTRACT

Teratomas are germ cell tumors commonly composed of cell types derived from all of the three germ layers. Intracerebral teratomas typically present in midline or paraxial lesions located in the pituitary stalk or the pineal region. Teratoma in posterior fossa is a rare entity. We reported a case of midline posterior fossa mature teratoma in a 3-month-old child.

KEYWORDS: Child, mature teratoma, posterior fossa

INTRODUCTION

Teratomas are congenital tumors that contain tissues derived from all the three germ layers: ectoderm (e.g., skin and hair), mesoderm (e.g., adipose tissue), and endoderm (e.g., respiratory structures and digestive organs).[1] Histologically, these lesions are classified into mature and immature types.[1] Teratoma commonly presents during infancy or in childhood. The estimated incidence of intracranial teratoma is 0.3%–0.6% of all intracranial neoplasm.[2] Teratomas in the posterior fossa are extremely rare.[2] Herein, author presented a case of midline posterior fossa teratoma.

CASE REPORT

Three-month-old female child admitted in neurosurgery department with drowsiness, vomiting, and refusal to feed of few days duration. On examination, she had bulging anterior fontanel and dilated scalp veins; she was comatose, with intermittent decerebrate posturing. A nonenhanced computed tomography scan revealed a posterior fossa mass with obstructive hydrocephalus with significant periventricular oozing. An urgent ventricular tap was performed in the emergency room to relieve intracranial pressure. The patient regained sensorium over next few hours. She was planned for definitive surgery after magnetic resonance imaging (MRI) of the brain.

MRI of the brain was suggestive of midline posterior fossa tumor that was variegated intensity on T1W and T2W and heterogeneous enhancing multiloculated cyst in contract-enhanced T1W [Figure 1]. In view of tumor and mass effect, patient was taken up for surgery. An external ventricular drain was placed before craniotomy. Midline suboccipital craniectomy with gross total excision of tumor was performed. Intraoperatively, tumor was firm with few areas of harder consistency. The tumor was mildly vascular with a well-defined plane of cleavage from the surrounding brain parenchyma. Patient tolerated the procedure well and recovered uneventfully [Figure 2]. The child required ventriculoperitoneal shunt for persistent ventriculomegaly after few weeks. Patient had shown gradual improvement and discharged home. At 2-year follow-up, the child has achieved normal developmental milestones.

Histopathological examination showed features of mature teratoma. H and E stained section reveals mature elements of all three germ cell layers, stratified squamous epithelium, hair follicles, intestinal epithelium, cartilage, and bone [Figure 3].

DISCUSSION

Intracranial teratomas are commonly located in the midline with estimated incidence of 0.3%–0.6% and have male predominance.[2] Incidence is higher in pediatric population. These tumors are mainly found in the suprasellar and pineal region; sometimes it is found in the cerebellopontine angle or fourth ventricle. Teratoma in posterior fossa is rare. First teratoma located in midline posterior fossa was reported in 1912.[3] In contrast to teratomas, epidermoid cysts

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are the lesions derived from a single layer, that is, the ectoderm (e.g., keratin), whereas dermoid cysts are the ones derived from two layers, the ectoderm and the mesoderm. The presence of adipose tissue in these latter tumors constitutes the main difference between them.[4]

Histologically, teratomas are classified into mature and immature types. Mature teratomas are benign and contain well-differentiated tissues. Immature teratomas are composed of undifferentiated fetal tissue and it has a tendency of malignant transformation, so it is also called as malignant immature teratoma.

Distant metastasis is common in malignant immature teratomas. Lung is the most common site of distant metastasis, and cerebrospinal fluid pathway spread has also been found. Teratomas commonly present during infancy or in childhood.[4] In our case, it was mature midline posterior fossa teratoma.

On reviewing the literature, Noudel et al.[5] reported case series of intracranial pediatric teratomas in which 5 of 14 patients found to have mature teratomas. However, all of them were located in pineal or sellar region but none of them in posterior fossa.[5] Kong et al.[6] reported 6 cases of teratomas from case series of 36 patients. Out of six cases only one was located in posterior fossa.

Intracranial teratoma has three types of presentations. In the first type, patients are still born or die immediately after birth. These tumors are quite large and replace brain tissue with teratomatous tissue. In the second type, child is born with enlarged head and tumor is relatively small at birth. In the third type, child is normal at birth but rapid enlargement of head occurred during early weeks of life. These patients harbor the smallest tumor.[7]

Teratomas are difficult to treat as these tumors are presented in younger age and quite large at the time of presentation. Complete surgical excision is the treatment of choice. Radiotherapy and chemotherapy are recommended in immature teratomas but use of radiotherapy is restricted in young patients. The prognosis of teratoma is poor irrespective of treatment.[8,7] Our patient recovered well and discharged home after surgery.

**CONCLUSION**

Midline posterior fossa teratomas are rare, but it should be included in differential diagnosis. Surgery is the treatment of choice, and complete surgical resection
is necessary to offer potentially curative therapy to the patient.

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**Conflicts of interest**

There are no conflicts of interest.

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