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REVIEW ARTICLE

Giant Congenital Intracranial Immature Teratoma: A Case Report and Review of Literature

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Abstract Congenital intracranial tumour constitute only 0.5–1.5% and are usually associated with poor prognosis. New advancement in imaging may help in early diagnosis. We presented a case of giant fetal intracranial tumour diagnosed antenatally at 28 weeks of gestation in sonography. The magnetic resonance imaging confirmed a large hetrogenous intracranial mass with bilateral ventriculomegaly. Because of the large size of the tumor and associated macrocephaly, the baby was delivered by caessaran section. On delivery, fetal head size was 44.5 cm with wide suture and left facial nerve was involved. Ventriculoeritoneal shunt was inserted on 4th postnatal day, however complete surgical resection of tumour was not performed because of parents refusal. Neonate had rapid increase in head size and expired on postnatal day 16th. The histopathological findings of mass demonstrated a congenital immature teratoma. Neonate expired on 16th postnatal day. Antenatal diagnosis of intracranial tumour is possible with advanced imaging modalities, however an early diagnosis before second trimester is difficult. Prognosis is generally poor and depends on size of tumour, histology and timely intervention.

Keywords Congenital teratoma · Fetal intracranial tumour · Fetal teratoma

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Introduction

Intracranial tumours of foetus are rare entity and constitute $< 1\%$ of all paediatric tumours [1]. Many case present with spontaneous in utero intracranial haemorrhage, hydrocephalus, macrocephaly, dystocia, and stillbirth. Regardless of their ability to grow and invade surrounding areas, the overall prognosis for congenital intracranial tumors is not good, with a global postnatal survival of only 28%.

Case Report

A 22 year primigravida consulted the antenatal clinic at 33 weeks of gestation for an abnormal 28 week ultrasound that revealed an echogenic mass in the fetal brain (56 mm × 39 mm) (Fig. 1) with bilateral ventriculomegaly, a reduced cortical thickness (1–2 mm) with minimal peripheral vascularity. No other anomalies were identified. Her investigation included TORCH serology and glucose tolerance test were normal. Fetal magnetic resonance imaging (MRI) at 38 weeks confirmed a lobulated solid cystic mass (89 × 59 mm) in right lateral ventricle, abutting the midline falx and displacing it to opposite side (Figs. 2, 3). There was also mass effect on third ventricle with obstructive upstream hydrocephalus and thinned out cerebral parenchyma. A female term baby weighing 2750 g was born through caesarean section in view of cephalopelvic disproportion. At birth, the baby did not need any resuscitation and had normal Apgar's of 8 and 9 at 1 and 5 min respectively. On examination at birth, the baby had a large head (44.5 cms; more than 2SD). The cranial sutures were wide with a bulging anterior fontanel. Baby was unable to close left eye and with absent nasolabial fold on left side,

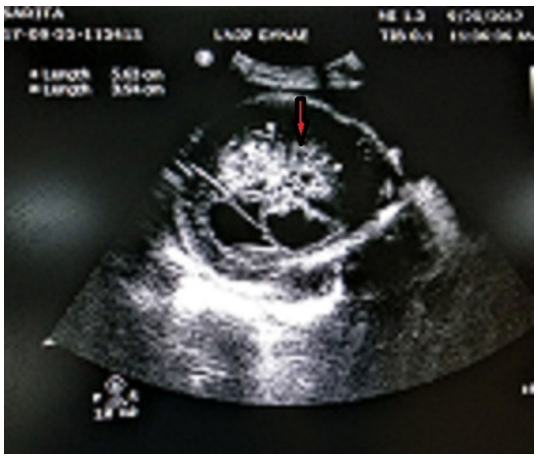

Fig. 1 Antenatal USG of fetal cranium

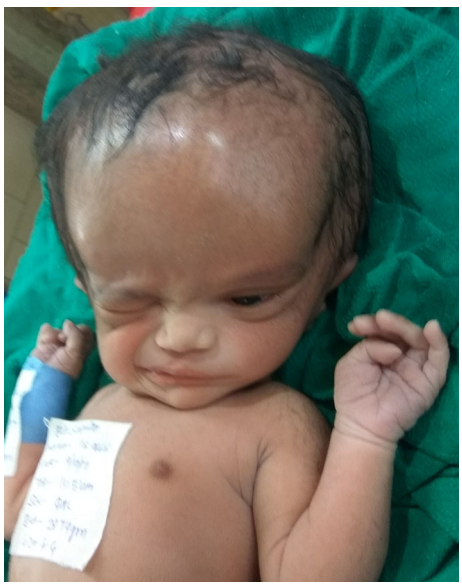

Fig. 2 Enlarged head of the baby with facial palsy at birth

which was suggestive of involvement of left facial nerve (Fig. 4). She had normal tone and symmetrical movements of the extremities.On postnatal day 2 of life, cranial ultrasound and contrast enhanced computed tomography confirmed antenatal findings.

The neurosurgery team was consulted and a craniotomy with removal of the mass was discussed with the parents. However, as the parents declined the consent for surgery, an early ventriculoperitoneal shunt was inserted on 4th postnatal day in view of increasing intracranial pressure. After the shunt placement, baby had an uneventful course intially, but had further rapid increase in the head size and expired on 16th postnatal day. The ultrasound guided postmartum biopsy of the intracranial mass was done after parental consent by the neurosurgeons. Autopsy was not performed due to parent's denial. Histopathology of tissue

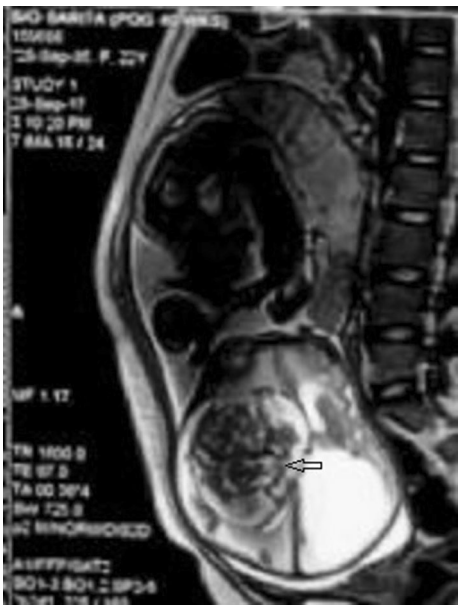

Fig. 3 Fetal MRI (Saggital view)-Heterogenous intracranial mass with displaced falx

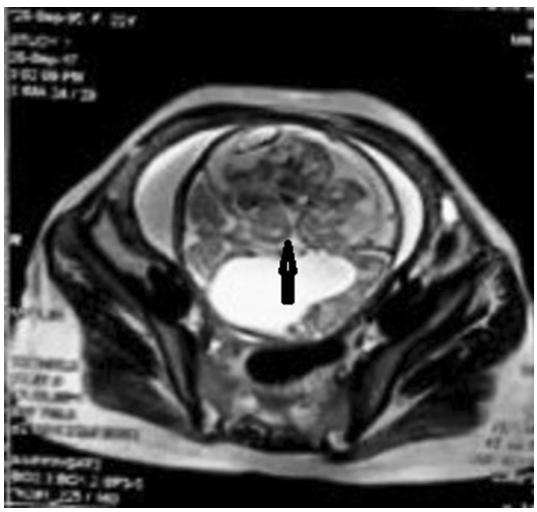

Fig. 4 Fetal MRI (Axial view)

from intracranial mass confirmed an immature teratoma (type 2).

Review of Literature and Discussion

Congenital intracranial tumours are rare and only few reports exist. Even though most of them are usually diagnosed in the late second or third trimester, an early diagnosis is uncommon and difficult. There are a few reports where the intracranial tumors have been diagnosed as early as 14 weeks of gestation [2]. The intracranial tumor should be suspected when a mass-occupying lesion, with cystic or

solid areas is seen within the fetal head or when there is a change in shape or size of the normal anatomic structures. These usually present as a rapidly growing heterogeneous mass associated with polyhydroamnios, macrocephaly, ventriculomegaly and at times intracranial calcifications. Hydrocephaly and macrocephaly results in secondary dystocia by cephalopelvic disproportion and delivery by vaginal route is usually not possible in 60% of the cases.

MRI is valuable in differentiating between tumours and haemorrhage and to delineate the extent of lesion. Cassart et al. [3], had retrospectively analysed 27 cases of fetal intracranial tumour by clinical findings and imagings. USG and MRI imaging appeared complementary in the antenatal assessment of these lesions and most were diagnosed after 20 weeks of gestation [3].

The definite diagnosis requires histologic examination. Histologically, the most common tumour subtypes include teratoma (26.7–36.5%), astrocytoma (9.5–28.9%), primitive neuroectodermal tumour (8.4–26.7%), choroid plexus papilloma (6.7–15.4%), and glioblastoma multiforme (3.1–14.6%) [4].

Teratomas are the most common ectopic tumour which contains multiple tissues from more than one mesoderm. They can be classified into: (1) mature teratomas (2) immature teratomas (3) malignant teratomas.

Foetal intracranial teratomas are usually more than 5 cm in diameter and frequently fill the cranial cavity completely. Because of the huge size of the tumours, it is often impossible to locate the exact site of origin. Unusual cases of a huge immature congenital teratoma with an extension through neck, pharynx and orbit and trigeminal nerve has been reported. In our case, facial nerve was involved.

The clinical management of these types of intracranial lesions is unclear, due to their low incidence and incomplete understanding of their natural history. Many cases have been reported in literature and primarily include single case report. Surgical excision may be curative, but it is only limited to the smaller benign intracranial teratomas. Fukuoka et al. [5] suggested that chemotherapeutic treatment diminished the aggressiveness and haemorrhagic nature of the tumor and allowed a second surgery to complete resection of tumor. Radiotherapy is not recommended in neonates especially for immature teratomas [6].

Prognosis of teratoma depends on the time of diagnosis, size of the teratoma and timely intervention. The prognosis is usually fatal because the lesions are usually extensive and grow intensively and destroy regular cerebral structures. Neonates with demonstrable cerebral parenchyma have the potential to improve following early management of hydrocephalus and surgical resection. Surgical resection is usually accompanied by developmental delay even if successful. An early diagnosis is therefore important, for timely counselling and an early termination of pregnancy to avoid obstetric complications.

Conclusion

Understanding of natural history and true incidence is important. Advancement of modern USG machines and timely scanning during pregnancy allow early detection of these tumours during fetal period. MRI helps to delineate the extent of lesion but definitive diagnosis can only be confirmed after birth by histology. As prognosis in neonates is generally poor and depends on histologic type, size of the tumour and timing of diagnosis. Early detection reduces patient discomfort by timely termination of pregnancy and prenatal counselling and to detect the potential lesion for surgical resection.

Compliance with Ethical Standards

Conflict of interest None.

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