CASE REPORT

Foetal intracranial teratoma: choosing the best time and mode of delivery

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We present a case of foetal intracranial teratoma diagnosed at 28 weeks of gestation after prior normal sonographic examinations. A multidisciplinary team, involving obstetricians, neonatologists, and neurosurgeons, suggested management. The foetus was delivered by lower segment caesarean section at 30 weeks of gestation but the neonate succumbed 3 days after delivery, 1 hour after the withdrawal of active treatment. Issues governing the timing and mode of delivery, together with the role of cephalocentesis in the management of this condition, are discussed.

Introduction

Foetal intracranial tumour is a rare disease associated with significant perinatal morbidity and mortality. Even where surgical excision is possible, neurological handicap is not uncommon. Since many cases present in the late second trimester or third trimester, termination of pregnancy is not a legal option in some countries. The timing and mode of delivery are, therefore, important issues to be discussed between the management team and the patient.

Case report

In December 2003, a 34-year-old woman, gravida 2 and para 1 (a normal vaginal delivery at full-term) booked in a university teaching hospital antenatal clinic at 13 weeks of gestation. An ultrasound examination showed a normal crown-rump length and no obvious foetal anomalies. She attended a private obstetrician for subsequent antenatal care and serial ultrasound examinations performed every 4 weeks showed normal foetal growth and morphology. At 28 weeks of gestation, a 7-cm heterogenous intracranial foetal tumour was identified on ultrasound and she was referred for further management. The finding was confirmed and normal intracranial structures were seen to be distorted (Fig 1). No arteriovenous shunt was observed inside the tumour and there was significant hydrocephalus involving both lateral ventricles, dilated to an atrial width of 3.3 cm. It was difficult to assess the third and fourth ventricles due to distortion of the normal anatomy. The biparietal diameter was 11 cm and the head circumference was 38 cm, which were both well above the 97th percentile for a term foetus. The liquor volume was normal and there were no other structural abnormalities.

Foetal magnetic resonance imaging showed a 7-cm heterogenous mass with multiple cystic components involving bilateral supratentorial regions. The cerebellum was

Key words
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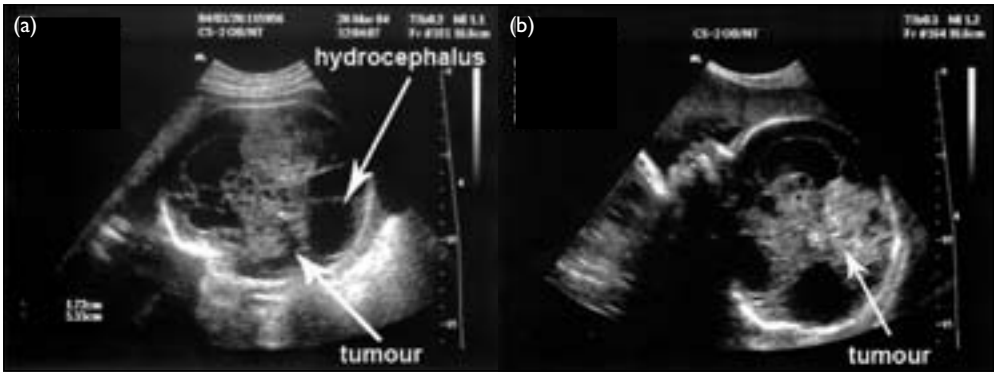

FIG 1. Obstetric ultrasound of the intracranial tumour at 28 weeks of gestation
(a) Transverse and (b) sagittal ultrasound images demonstrate the presence of an intracranial tumour and hydrocephalus

胎兒腦內腫瘤：決定最佳的生產時間及方法

本文報告一個在28孕周首先發現的胎兒腦內腫瘤的治療個案。一組由產科、初生嬰兒科和腦外科醫生集合的專家共同商議和會診。在30孕周，胎兒從下段剖腹生產。3天後，在移除主動性治療1小時後，嬰兒便去世。本文逐一探討如何決定生產的時間及方法，和頭顱穿刺術是否可行。

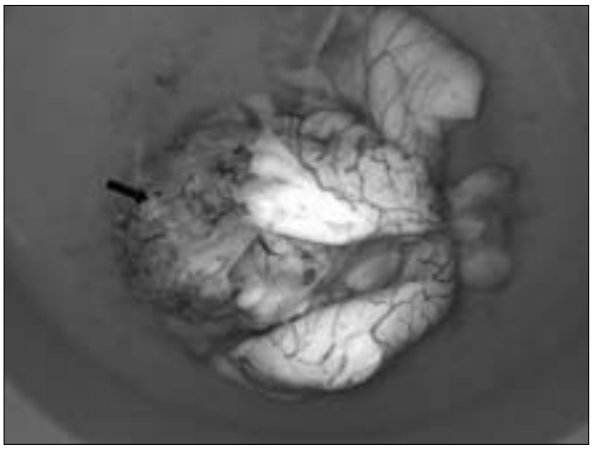

FIG 2. Macroscopic view of the intracranial tumour (arrow) occupying the left cerebral hemisphere

compressed inferiorly and normal brainstem could not be identified. There was also a marked obstructive hydrocephalus involving both lateral ventricles. There was no evidence of intracranial haemorrhage. A diagnosis of intracranial teratoma was made. The couple were given counselling by members of the foetal medicine unit together with the neonatology and neurosurgery consultants. They were counselled about the baby's very low chances of survival given the size and location of the lesion, and informed that it was very likely that brain development would be affected even if the baby survived and the tumour was resected. If the baby's condition was stable and the tumour assessed as resectable on post-delivery imaging, surgical excision would be performed. The couple understood that their baby's prognosis was poor. They were also counselled about the mode and timing of delivery and told that further delay of delivery might make delivery more difficult as the size of the tumour was expected to increase further. The use of antenatal corticosteroids to reduce respiratory distress syndrome associated with preterm delivery was also discussed. The neonatologist considered the chances of the baby being stabilised would be optimised if the delivery was done at 30 weeks of gestation after a course of antenatal corticosteroids. The parents were also counselled that while caesarean section might not improve the baby's prognosis there would be an increased risk of maternal morbidity. They were also informed that induction of labour in a preterm gestation is usually more prolonged and that the baby's large head was likely to make the delivery more difficult, leading to increased trauma to both mother and baby. Finally, it was decided that a caesarean section should be performed after one course of intramuscular antenatal corticosteroids, and the couple agreed that post-delivery assessments and surgical excision of the tumour should be done if needed.

A male baby weighing 2.54 kg was delivered at 30 weeks of gestation by lower segment caesarean section. The head circumference was 42 cm. The skull sutures were widened and the anterior fontanelle was tense. There was no dysmorphism, nor other structural abnormalities. He was intubated soon after delivery for poor respiratory effort. A computed tomographic scan of the brain showed a large supratentorial heterogenous mass containing specks of calcification and cystic components. The cerebral hemispheres were poorly formed and the cerebellum was compressed. The neurosurgeons and neonatologists considered the mass non-resectable and no other modalities of treatment were available. After further counselling, the couple elected to withdraw active treatment. The baby died 1 hour after extubation.

A postmortem examination found a severely hydrocephalic brain and a large 8-cm intracerebral tumour replacing the entire left cerebral hemisphere (Fig 2). Histological examination showed an immature teratoma.

Discussion

Congenital intracranial tumours are rare, with a reported incidence of 0.34 per million live births.[1] Teratoma is the most common type, accounting for about one third of cases.[2] In a review of 39 cases,[3] 44% were detected between 28 and 32 weeks of gestation while only 10% were detected before 24 weeks of gestation. The sudden appearance of the intracranial tumour at 28 weeks of gestation seen in this case is not uncommon, a phenomenon that may be related to rapid growth of the tumour in the second trimester.

Because of the usually extensive involvement and destruction of normal intracranial structures, the prognosis for foetal teratoma is generally poor. One series reported a 1-year survival rate of 2.4% in 42 cases with confirmed congenital intracranial teratomas without epignathus.[4] In a more recent review of cases reported since 1980, three (7.7%) babies had their intracranial tumours resected and were the only survivors at 4 years of age, at the time of the report.[3] Of these three cases, two were diagnosed with developmental delay when reviewed at 3 to 4

years of age.[5,6] It is difficult to predict resectability of these tumours antenatally. In the reported cases, one occupied the left cranium with marked distortion of the facial bones,[5] and another was based in the temporal fossa with extension to the anterior cranial fossa and occipital region.[6] Therefore, large tumour size and spread does not necessarily mean the tumour cannot be resected.

Timing of the delivery should be decided by the multidisciplinary team and the patient jointly. The decision requires balancing the risk of prematurity with the risk of further compression of any remaining cerebral tissues. Moreover, referral to a tertiary centre, further imaging studies, and repeated counselling sessions all increase the detection-delivery interval. In this case, the delivery was about 2 weeks after detection, which was comparable to the usual detection-delivery interval reported in cases detected between 28 and 32 weeks of gestation. Around 80% of these babies were delivered within 2 weeks of having their tumours detected.[3]

Selecting the mode of delivery poses a dilemma. On the one hand, it is an obstetric principle that the higher maternal risks imposed by caesarean section should be avoided if the foetal prognosis is ominous. On the other hand, the likelihood of an ever-growing intracranial tumour causing cephalopelvic disproportion and its associated morbidity cannot be underestimated. Most reported cases were delivered by caesarean section despite the poor foetal prognosis. Determining the optimal mode of delivery is problematic given the difficulty of performing a randomised controlled trial in this rare situation. In a review of 132 mandated preterm deliveries performed in a tertiary centre between 24 and 37 weeks of gestation, only four cases were due to foetal malformations.[7] The overall caesarean section rate was 70%, which was due to unfavourable cervix, unsuccessful trial of labour, non-cephalic foetal presentation, and foetal intolerance of labour. Most preterm labour induction was done for pre-eclampsia. Blackwell et al[8] showed that the successful induction rate drops from 69% between 32 and 34 weeks of gestation to 35% between 29 and 31 weeks of gestation. It may not be appropriate to apply this to foetuses with intracranial tumours or macrocephaly, where the large head might cause failure of progress of labour and even skull fracture.

Cephalocentesis, either ultrasound-guided transabdominally or intrapartum transvaginally, has been advocated as a means of facilitating vaginal delivery and thus avoiding the maternal morbidity of caesarean section in cases of severe hydrocephalus.[9,10] Its value as a means of managing very large intracranial tumours is controversial. In their review, ten Broeke et al[11] found that cephalocentesis was employed in only five cases out of 18. Although three of these had a successful vaginal delivery, the procedure may be complicated by the risk of intrauterine infection, haemorrhage, damage to any remaining cerebral tissues, and patient discomfort. Moreover, even after cephalocentesis, the foetal head size may not be reduced significantly if there is a large intracranial tumour, as in this case, that cannot be decompressed. Furthermore, as mentioned before, resectability is difficult to predict antenatally and cephalocentesis may negatively affect the prognosis.

Although some countries (for instance, the United Kingdom) permit termination of pregnancy after 24 weeks of gestation if there is evidence of severe foetal abnormality, this is illegal in Hong Kong. Only 10% of reported foetal intracranial tumours have been detected before 24 weeks of gestation. Therefore, in most cases, termination of pregnancy is not a valid option. This case report illustrates the need to achieve a fine balance between reducing morbidity while maximising the prognosis for the foetus.

In conclusion, foetal intracranial tumours carry a poor prognosis, and individualised multi-disciplinary management and decision-making involving the pregnant woman is important. Most cases are diagnosed late in the second trimester and delivered well before term. The optimal mode of delivery is controversial and the choice made needs to balance the maternal risk of caesarean section against the possibility of cephalopelvic disproportion and successful induction of labour in a preterm gestation.

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