INTRODUCTION

Epignathus tumour is an extremely rare type of congenital teratoma arising from the oral cavity. It causes severe airway obstruction and is, therefore, associated with high neonatal mortality.\(^1\) It is associated with polyhydramnios and elevated serum alpha fetoprotein levels. Accurate prenatal diagnosis is of great value for counselling and appropriate management, including EXIT procedure, where appropriate.\(^2\)

We present a case of stillborn baby with epignathus and polyhydramnios.

CASE REPORT

A 28-year-old woman presented in emergency, with history of gestational amenorrhoea of approximately 5 months. She had labour pains and leaking for the past 5 hours. She was gravida 2 and para 1. The previous pregnancy was full term and a healthy baby boy was delivered by normal spontaneous delivery. She had no previous antenatal checkups during the present pregnancy. She did not have any significant medical or surgical history. She gave history of fever during the 4th month of her pregnancy, for which she took some medicines; however, she could not recall their names. Otherwise her pregnancy was unremarkable.

On examination, she was 26 weeks pregnant and was in advanced 1st stage of labour. Her blood pressure, pulse and temperature were within normal limits. Emergency ultrasound showed an alive, active foetus with unstable presentation. Biometry showed an estimated gestational age of 26 weeks and 5 days. There was a large mass measuring 10 x 10 cm anterior to the foetal neck. It had well defined lobulated margins and a heterogeneous echotexture containing hyper echoic as well as large anechoic, cystic areas. There was no lateral or posterior extension of the mass. The foetal spine was normal. There were no signs of foetal hydrops. No additional structural abnormality was identified. The liquor was, however, grossly increased.

The patient was treated after maintaining intravenous line by ringolactate infusion. She was given antibiotics and analgesics. She spontaneously expelled a fresh...
stillborn female baby in the second stage of labour, 2 hours after admission. The mass visualized on ultrasound was seen coming out of the baby's mouth, which was wide open due to the mass (Figure 1). The mass was attached via a 3-cm long stalk superiorly at the junction of the soft and hard palate. The baby was otherwise normal. The mass was friable and bled to touch. It had mixed solid and cystic areas on gross examination of the cut section. The baby and the mass together weighed 2.8 kgs. The mass was 900 g in weight. Histopathology of the mass showed that the outer wall was composed of skin appendages and neural tissue. The cystic spaces were lined by columnar cells. The intervening areas were occupied by fibrofatty tissue. There was no evidence of malignant cells. Mature teratoma was thus diagnosed.

**DISCUSSION**

Congenital teratomas are rare tumours with an incidence of 2.5:10000 live births. Epignathus is a rare oropharyngeal teratoma arising from upper jaw, palate, and sphenoid bone. It has an incidence ranging between 1:35000-200000 live births. It shows a female predominance. Its mortality ranges from 80% to 100%. The child in our case was a stillborn female.

Jordan and Gauderer proposed a classification of cervical teratomas based on birth status, age at diagnosis, and the presence or absence of respiratory distress. This patient fell into Group I, according to this classification.

Epignathus may occur as an isolated abnormality or associated with other congenital abnormalities like cleft palate (commonest), bifid tongue and nose, and Pierre Robin syndrome (cleft palate, glossoptosis and micro-retrognathia). Association with meningo-encephalocele has also been described. Duplication of cranial base, mandible and pituitary gland have also been noted with it. This case was not associated with any additional malformation. These tumours are almost always benign. Too et al. however, reported a case of malignant epignathus teratoma. Some elements of these tumours may become malignant after incomplete removal.

Benign mature teratoma was diagnosed on the histopathological examination of the resected tumour in this case. Epignathus occurs more frequently in young mothers. In this case, the mother was 28 years of age. These lesions are non-familial. The tumour is usually associated with polyhydramnios, secondary to swallowing difficulty. Placental oedema may be associated with it because of foetal cardiac decompensation, secondary to extensive vascularisation of the tumour. Pre-eclampsia has also been described with it. Maternal serum alpha fetoprotein levels are always elevated in this condition. Most cases of epignathus arise from the sphenoid bone. Some arise from the hard and soft palates (as seen in this case), the pharynx, the tongue, and the jaw. From the site of origin, the tumour grows into the oral or nasal cavity or intracranially. It can lead to death soon after birth due to severe airway obstruction.

On prenatal ultrasound, the presence of a mass related to the foetal mouth showing mixed echotexture with soft tissue echogenicity, cystic components, and calcification, associated with polyhydramnios, suggest the most possible diagnosis of epignathus. Use of 3 dimensional ultrasound can further confirm the relation of the mass with the foetal mouth. Kaido et al. have described the use of colour Doppler ultrasound to assess the tumour vascularity. They suggested that a hypovascular tumour is associated with favourable prognosis, decreasing in size naturally in the prenatal period. After the initial diagnosis, ultrasound and MRI can be used to assess the relationship of tumour with the surrounding structures, its extension, and any complications caused by it.

When the epignathus is not associated with additional congenital anomalies, the patient can be operated on placental support (OOPS procedure). The delivery is planned with the cooperation of obstetricians and surgeons. The baby is delivered by caesarean section with immediate ablation of the tumour using EXIT technique (ex utero intrapartum treatment). The uterus is kept relaxed with the intact uteroplacental flow. The foetus is supplied by oxygen via umbilical cord until the tumour is removed.

Alternatively, especially if there is suspicion of intracranial extension of the tumour or associated primary lesions of the central nervous system on the antenatal investigations, after delivering the baby by caesarean section, the umbilical cord and the foeto-placental circulation are left intact to allow oxygenation of the foetus until a rapid examination and endotracheal intubation or tracheostomy is done, as necessary. After securing the airway, the umbilical cord is clamped and the baby is transferred to the neonatal intensive care unit. The surgery can be done subsequently once the baby is stable. The diagnosis can then be further confirmed by plain X-rays and CT scan. Radical disfiguring surgery is contraindicated in the neonate as it may result in the impairment of speech and deglutition. A purely endoscopic transpalatal endonasal approach can be used as an alternative to the conventional transfacial approach.

Foetal surgical removal of the tumour is another possible treatment option. Although it has not been reported for treating the epignathus, it has been successfully used for the treatment of sacrococcygeal teratoma in addition to other congenital defects, like diaphragmatic hernia. Open as well as minimal access techniques of foetal surgery can reduce pre- and postnatal mortality in patients with this tumour. However, further studies are
required to determine their role in the patients with epignathus teratoma.

REFERENCES