A rare case of Epignathus

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The term Epignathus describes a rare benign teratoma (poorly organised tissues derived from elements of three embryonic germ cell layers) that originates from orophanynx associated with serious airway compromise upon birth. The tumour grows from palate, mandible or base of the skull and protrudes through the mouth. Pedunculated Epignathus consists of structures confined to head and neck attached either to nasopharynx in the

region of the basiphenoid or dorsal aspect of palate and ptergyoid plates (Rakhe's pouch). This occurs predominantly in females and clinical picture includes dyspnea, suffocation difficulty in swallowing, sucking and vomiting leading to high mortality.

Case report

Patient 'S', 23 years old G_2P_1 was admitted to Vanivilas Hospital, Bangalore with the history of pain in abdomen and amenorrhea of 6 months duration. She was married for 3 years. There was no H/o consanguinity. She had L.S.C.S. for her first baby for breech presentation. Her cycles were regular

and was immunised elsewhere. There was neither H/o fever, drug intake, consumption of alcohol and chewing tobacco nor there was family H/o twins or anomalous baby in both families.

On examination, she was moderately built and nourished. All her vital parameters were within normal limits. Per abdomen examination showed 36 weeks gestation and a fluid thrill. Foetus was presenting by breech. It was difficult to localise the FHS. A diagnosis of

polyhydramnios was made and an ultrascan undertaken. Her haemoglobin was 12.1 gms% RBS 72mg%. VDRL - Non-reactive. Urine was clear for both sugar and albumin. Hbs Ag was '-ve'. BT and CT were within normal limits. Ultrascan showed polyhydramnios and a dead foetus of 25 weeks gestation. FL 4.6 mm, BPD 67 mm. There was a large solid and cystic exophytic lobulated mass measuring about 11.3 x 8.1 cm arising

from foetal frontal nasal region. There was calcification in the mass. There was oedema of foetal scalp. Placenta was grade 'O' and fundal. Initially, it was difficult to diagnose the Epignathus as it was mimicking a dysmorphic twin.

Labour was induced with cerviprime and oxytocin was given for accentuation of pains. Patient delivered a dead baby after 8 hours. MRP was done as the placenta was adherent.

The dead born foetus was male which is rare in Epignathus and

it was weighing 2.5~kgs. The complex mass was 11.3~x~8.1~cm and it was arising from posterior part of hard palate. There was tuft of hair in the mass and also a part of small intestine was present. X-ray of the mass showed 12~teeth.

Prenatal diagnosis is important for genetic counselling, obstetric management and in some cases neonatal surgery to save the life of such a malformed baby.

