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Epignathus: More than a Mouthful - A Case Report and Review of Literature

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Abstract
We report a case of antenatal diagnosed epignathus which was managed on emergency basis in view of premature rupture of membranes. Child underwent an emergency tracheostomy in the immediate post-natal period and successful excision of epignathus tumour. Post procedure tracheostomy was closed after 2 weeks, and child recovered well. Biopsy was suggestive of mature teratoma.
Keywords: *epignathus, neonatal surgery, teratoma, emergency tracheostomy, antenatal diagnosis.*

Case Report

A mother presented with 20 week antenatal ultrasound showing soft tissue thickening lower lip? teratoma (12mm), a repeat ultrasound showed a 21x25mass arising from the edge of the mandible, with mandibular base partially displaced -?congenital foetal epulis. Mother was on regular antenatal checkup and was being planned for an elective CS with an EXIT procedure.

But, in view of Preterm premature rupture of membranes (PPROM) mother had to be taken up for emergency C section. Call was received from Labour room for assistance in airway management (i.e. tracheostomy; if necessary), parallel OT and paediatric anaesthesia team and neonatology team was present. After C section, cord was clamped and baby had a weak cry, a large soft tissue mass was noted arising from the mouth (Figure 1). In view of weak cry, bag and mask ventilation was attempted, child developed bradycardia and cyanosis. Chest compressions were initiated, and two shots of adrenaline were given. Endotracheal intubation was not feasible due to large mass protruding from mouth. Emergency tracheostomy was proceeded with using 3mm uncuffed tube under local anaesthesia and monitored anaesthesia care. Child was stabilized, a UV line was placed, and child shifted to NICU. Child underwent excision of epignathus under GA on post-natal day

1. A ~5×4x4cm sized tumour with variable consistency – soft and areas of bony hard consistency was excised. Tumour had a wide based which was arising from Lt side soft palate and extending to tonsillar pillar, base bony in consistency with ill-defined margins (Figure 2). After excision a complete cleft palate was noted with a large defect and deformed mandible with pseudomacrostomia.

Figure 1 Large epignatus tumour, post tracheostomy picture

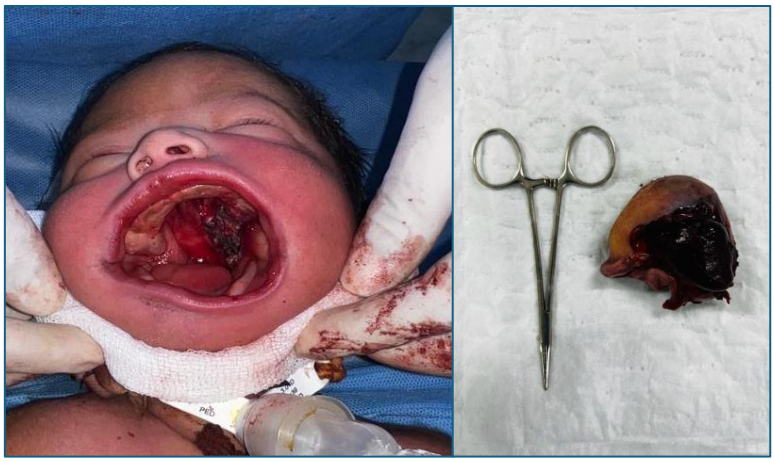

Figure 2 Post excision view of palate with resected specimen

Child underwent tracheostomy closure on POD 14, and orotracheal intubation, which was uncomplicated. But child self-extubated on post op day 1, child had minimal tracheostomy site infection which was managed conservatively, child also developed late onset neonatal sepsis (enterococci) on PND 14 and was managed with IV antibiotics. Child was started on OG feeds by post op day 4. Pedodontics opinion was sought in favour of mandibular remodelling devices, and strapping of lips was advised. Patient gradually was transitioned to expressed breastmilk feeds and then direct feeds. MRI of the brain and facial bones showed no residual lesions, no significant intracranial abnormalities, a large midline cleft palate, hypoplastic mandible with retrognathia, and normal bilateral TM joints. Biopsy was suggestive of a mature teratoma. On post op

review at 3 and 6 months, child has good thrive and was feeding well (Figure 3). Patient underwent elective cleft palate repair and surgical repair of macrostomia at 14 months and 20 months of age respectively with good cosmetic and functional outcome (Figure 4).

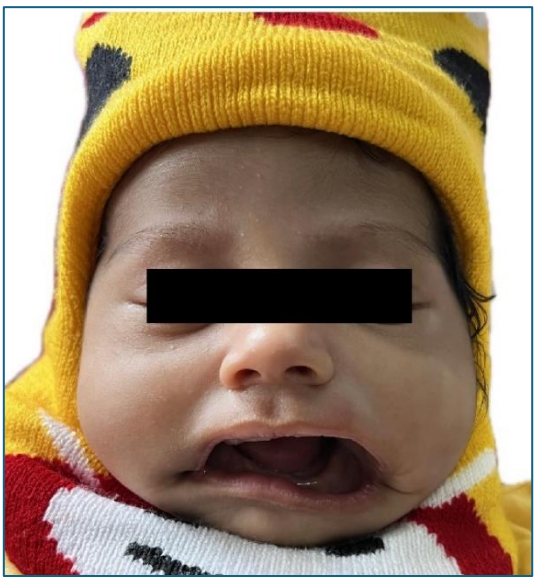

Figure 3 Patient at 3 months follow up.

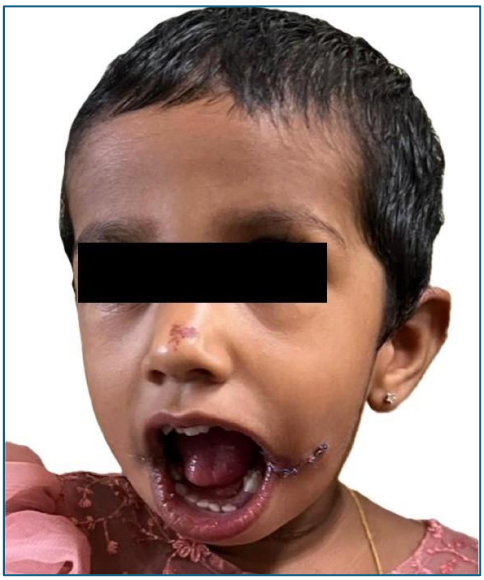

Figure 4 Patient at 20 months follow up.

Discussion

Epignathus (epi – up/above + Gnathus - Jaw) term for teratomas protruding through the mouth. It is rare with an incidence of 1 in 200,000 births. It Accounts for 2–9% of all teratomas. It's known to arise from hard or soft palate in the region of Rathke's pouch. In the antenatal period the large tumour prevents foetal swallowing and leads to polyhydramnios, resulting in premature rupture of membranes. Other antenatal features include raised amniotic AFP and pre-eclampsia. 3D ultrasound and foetal MRI can aid in better delineation of the lesion. Epignathus is usually benign, with rare reports of malignancy[1]. Intracranial extension has been reported. Child presents with a mass protruding from mouth and

makes it challenging for the child to swallow and breathe and, in this situation it is fruitless to attempt oro-tracheal intubation, nasotracheal intubation has been attempted in the past, may be difficult to find appropriately sized tubes, and may also cause bleeding from the tumour. Treatment plan should include an elective Caesarean Section and securing airway via tracheostomy. The tumour resection is deferred once baby is stabilized after ruling out intra-cranial extension. EXIT procedure[(2)] would involve performing caesarean section under deep general anaesthesia and delivering the head and neck, to allow access for tracheostomy before clamping the cord. Epiganthus is associated with - Pierre robin syndrome, cleft palate, bifid tongue, and bifid uvula. Differential diagnosis for this condition includes oropharyngeal rhabdomyosarcoma, giant epulis, cystic lymphangioma – oral/nasopharyngeal and heterotopic thyroid tissue. Further management includes cleft palate repair, mandibular moulding devices for deformed mandible and speech therapy.

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