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Epignathus Teratoma in A Newborn - A Rare Case Report .

KEYWORDS: Epignathus , Oropharyngeal , Teratoma, Germnial layers

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ABSTRACT *Oropharyngeal teratoma is a very rare form of malformation composed of cells of all three germinal layers. It arises from the palate / pharynx and protrudes from the mouth . The incidence is 1 in 35,000 - 2,00,000 births with female predominant. Classically presents inutero or neonatal period. We represent a large skin covered teratoma that arose from the hard palate and floor of the mouth in a neonate . 1 ½ hours old term girl baby delivered by naturalis to a primi mother with birth weight of 2.6 kg. Cry and activity was good with mild respiratory distress. There was a 5 X 3 cm mass protruding out of the mouth with soft to firm in consistency . Bifid tongue and complete cleft palate, fusion of mandible. Child was investigated with CT Skull & tumour markers . We planned for excision and the entire mass was excised in toto . Histopathology report came as mature teratoma . Post-operative period was uneventful.*

Case history :

One and half hours old , term , girl baby delivererd by naturalis with birth weight Of 2.6 kg. Meconium history and bladder habits normal . On examination Colour - pink, cry & activity -good , Heart rate – 142/mt, mild respiratory distress . Local examination - 5 X 3 cm soft tissue mass protruding between upper and lower lip with soft to firm in consistency . Bifid tongue with complete cleft palate. The mass was occupying entire oral cavity arises from hard palate and floor of the mouth (fig.1) Child was investigated with imaging of CT Skull (fig .2) and confirmed that the lesion was confined to the oral cavity with normal tumour markers of alpha fetoprotein and beta HCG . We planned for excision . Intra operatively (fig.3) we found that bony lesion extending into the hard palate which was removed with help of bone cutter. The entire mass was excised in toto.

Figure -1 . New born with mass in the oral cavity.

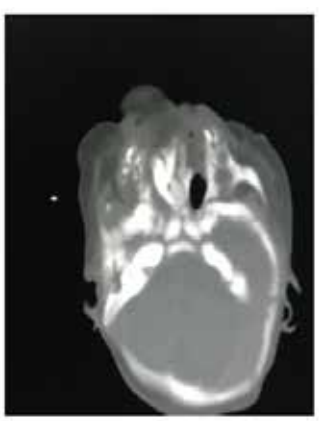

Figure-2 . CT picture of Mass.

Histyopathology Showed features of mature teratoma - lined by stratified squamous epithelium underlying adnexal structures , fat lobules , mature cartilage , salivary acini , skeletal muscle bundle and blood vessels . Cyst space lined by columnar epithelium & mature glial tissue . No immature elements. Post –operative period was uneventful . Child was started of oral feeds with palate covering pads (fig.4)

Figure-3. bifid tongue with cleft palate

figure-4. Palate covered with pad.

Discussion :

Epignathus teratoma is very rare form of teratoid tumour arises from the oropharyngeal region. In 1837 Geoffry & Hillaire described the term epignathus.

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The first case report was made by Bown – Kelly in 1918. Incidence 1 : 35,000 - 2,00,000 births with female predominant. Within this oropharyngeal origin is < 2% and nasopharyngeal origin of 60%. In 1940 , Ewing's classifies nasopharyngeal teratoma into three types.

1. Dermoid - Epidermal / Mesodermal germ layers- attached to soft/ hard palate.

2. Teratoma - Consists of all three germ layers with indifferent degree of organs.

3. Epignathus - Teratoma with high degree of organization and recognizable Structures.

Classically presents inutero / neonatal period. Very rarely young children . Pathologically it is classified under cervical teratomas and have components of all three germ layers and contain fat , cartilage , muscle. Site of origin is palato – pharyngeal region around the basisphenoid (rathke's pouch) , grow progressively and fills the buccal cavity and finally protrudes out of the mouth. The associate features are polyhydramnios and midline facial defects cleft lip & palate. Investigation shows increased maternal serum alpha fetoprotein . Antenatal ultrasonogram shows complex mass protruding from the oral cavity with polyhydramnios. For better delineation of anatomy MRI is ideal.

Excision is the ideal line of management . Prognosis is generally poor due to its location and airway obstruction. EXIT procedure is offered in selected cases.

Conclusion :

Epignathus teratoma is a very rare tumour, can cause neonatal death dueto airway obstruction and asphyxia after birth. Surgery - resection is the treatment of choice . Majority of these tumours are benign in nature. Mortality rate is high because of asphyxia secondary to airway obstruction - either immediately after birth or during birth.

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