Case report:

**Congenital nasopharyngeal teratoma with cleft palate in neonate**

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**Abstract:**

Congenital Germ cell tumours are uncommon. The most common site of teratoma is in the sacrococcygeal region. Teratoma arising from the Head and Neck comprises less than 10% of reported cases and of these cases nasopharyngeal lesions are rare. We present a case of nasopharyngeal teratoma with large cleft palate with parietal meningomyelocoele. Herewith we presented a case of 3 days old female child with sudden onset breathlessness with cyanosis and convulsions. On examination baby with birth weight 2.3 kg with cleft palate with right parietal meningomyelocoele with Bony hard mass which was irregular in shape measuring about 5x3x2cm arising from nasopharyngeal cavity extending towards posterior pharyngeal wall causing obstruction to airway and choking. Emergency Tracheostomy done, distress relieved, cyanosis relived. Child had aspiration pnemonitis with severe respiratory distress hence baby put on ventilator.

**Introduction:**

Teratoma of Head and Neck are rare less than 10% of all cases of which nasopharyngeal teratoma is very rare. Teratoma are benign tumours containing cells from Ectodermal, Mesodermal and Endodermal layers and they involve at least two of the above layers. Although they are rare neoplasms composed of tissue elements derived from germinal layers of the embryo, they may originate anywhere along the midline and clinical behavior varies significantly by size and site. Nasopharyngeal teratoma usually benign but may cause considerable morbidity and mortality because of their location. Nasopharyngeal teratomas represents one of the most unusual causes of respiratory distress during the neonatal period. Mostly evaluated by CT scan, MRI can provide additional vital information. MRI provides better soft tissue characterization than CT scan.

**Case presentation:**

3 days old female child with sudden onset breathlessness with cyanosis and convulsions. On examination baby with birth weight 2.3 kg with cleft palate with right parietal meningomyelocoele with Bony hard mass which was irregular in shape measuring about 5x3x2cm arising from nasopharyngeal cavity extending towards posterior pharyngeal wall causing obstruction to airway and choking. Emergency Tracheostomy done, distress relieved, cyanosis relived. Child had aspiration pnemonitis with severe respiratory distress hence baby put on ventilator. Clinically Baby stabilized after 5 days with gradual weaning from ventilator. Surgical resection of bony hard mass done with repair of cleft palate in a single stage. Biopsy report suggestive of mature teratoma. Clinically child improved and discharged on 28th day of life with closure of Tracheostomy. Now accepting breast feeds well,
has gained weight, without any signs of distress and radiological resolution of Pneumonitis.

MRI Neck-
Approximately 5x2.7x2cm sized lobulated mass lesion in nasopharynx inseparable from soft palate, adenoid region and mainly right sided tonsillar fossa region causing severe narrowing to nearly obliteration of nasopharyngeal and partly oropharyngeal passage with central fat containing areas, cystic areas and few calcific areas suggestive of neonatal nasopharyngeal teratoma.

Histopathology report-
Gross examination- Specimen consists of a single polypoid nodular mass of grey tan tissue measuring 3.5x2.5x2cm. cut specimen shows grey tan appearance with a piece of bone measuring 1.5x1.5x1cm.

Microscopic Examination- Section show a polypoid mass composed of fibrocollagenous and fibroadipose tissue lined by mature epidermis with skin adnexal structures like sebaceous glands, sweat glands and hair follicles or by pseudostatified columnar epithelium. E/O surface ulceration covered with acute inflammatory exudates is seen. The deeper tissue shows mucous glands, mature bone, mature cartilage, adipose tissue showing foci of fat necrosis, congested blood vessels and diffuse mononuclear cell infiltration. There is no e/o malignancy in the sections examined.

Impression – Mature Teratoma

Discussion:
Teratomas are Benign tumours containing cells from ectodermal, mesodermal, endodermal layers. They occur in about 1 in every 4000 births and most commonly in the sacrococcygeal region, followed by the ovaries. Congenital epignathus teratomas are rare embryological neoplasms localized in the region of Head and Neck. An epignathus is found in approximately 1:35000 to 1:200000 live births, accounts for 2-9% of all teratomas(1). Epignathus refers to teratoma of oropharyngeal region.

Teratoma arising from Head and Neck region comprises less than 10% and of these nasopharyngeal lesions are rare(2). Teratomas are congenital tumours that are composed of tissues derived from all three embryonic germ cell layers.

Four basic histological classifications are generally recognized-
1)Deroid cysts (Epithelial lined with skin, elements, composed of ectodermal and mesodermal cells)
2)Teratoid cysts (all 3 germ layers but poorly differentiated)
3)True Teratoma (all 3 germ layers differentiated into specific tissues or organ)
4)Epignathi (oral tumours with developmental fetal organs and limb, very rare, with high mortality rate)(3)

Another pathological variation of epignathi is fetus in fetus, which may be considered to be incomplete twining of monozygotic twins at a primitive stage when axial development begins(4) Teratomas are true neoplasms originating from pleuipotent cells and composed of tissues from all three germinal layers. Hairy polyp(Dermoid) is a teratoid lesion(5). Hairy polyp of the pharynx may be associated with an ipsilateral branchial sinus (6). Teratomas are more common in females. Usually benign when they present in early childhood(7). In Nasopharyngeal teratoma the most common symptoms are upper respiratory obstruction, dysphagia and failure to gain weight. Polyhydramnios and severity of respiratory distress correlate with site and size of teratoma. Large teratoma cause hyperextension of

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the neck of the baby and lead to oesophageal obstruction, swallowing disturbances and polyhydramnios (8).

Clinical differential diagnosis of neonatal oral mass include-
- Embryonic congenital Rhabdomyosarcoma
- Retinoblastoma
- Nasal Glioma
- Heterotropic Thyroid
- Cystic Lymphangioma of oro or oropharyngeal region
- Sphenoid Meningoencephalocele (4)

Diagnosis: CT and MRI used to differentiate from other causes.

Treatment: Complete surgical excision depending on site of tumour with proper medical management.

Prognosis: Prognosis is excellent, rare chances of recurrence.

Conclusion:
Congenital Nasopharyngeal Teratomas are usually benign. Surgery is the treatment of choice, surgery should be done on emergency basis in a patient who presents with signs and symptoms of severe airway obstruction.

In our case Inspite having severe respiratory distress with aspiration pneumonitis and seizures. After surgery patient improved dramatically with no residual neurodeficit, radiological and clinical clearance of pneumonitis. Child improved clinically with accepting breast feed and gaining weight adequately.

References:
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