Congenital Teratoid Tumour of Nasopharynx With Cleft Palate

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Abstract

A case of congenital nasopharyngeal teratoid tumour presenting since birth projecting into oral cavity through the palatal cleft & gradually increasing in size thus causing breathing & feeding difficulty in a four months old female child is presented here for its rarity. A brief review of literature and management of such patients is also discussed here.

Teratoid tumours are defined as the "Masses arising from pluri potential cells and are composed of wide diversity of tissues foreign to the site where they arise". They are tridermal in origin and contain fat, muscles, acinous glands, tooth, cartilage, bone and are lined by various forms of epithelium and nervous tissue.

Teratoid tumours of nasopharynx form a rare but well recognised group of tumours. They arise close to the midline, commonest sites being sacrococcygeal region, mediastinum, retroperitoneal tissue, cranium and neck.

Till 1978 (Choudhary et. al.) 113 cases of nasopharyngeal teratoids were reported.

The case presented here is associated with a cleft palate which is not yet reported in the literature.

Case Report

A four months old female child was referred to ENT department for a mass occupying most of the oral cavity gradually increasing in size and causing breathing and feeding difficulty since birth. Baby was first born female child of a normal full term pregnancy and labour. Birth weight was 2.2 kgs. Baby had breathing difficulty since birth and also a noticeable mass filling the oral cavity protruding through a large defect in the palate. Baby had undergone CT Scan head (at Nagpur) on third day after birth but it was normal and did not reveal any intracranial connection of the mass.

On ENT examination baby was found to have hypognathism, mouth breathing and an infant feeding tube in right nostril. There was a big mass occupying oral cavity going to posterior pharyngeal wall and superiorly coming from nasopharynx through big defect in the hard and soft palates. Bifid uvula could be seen on either sides of the mass. Breathing was laboured. Weight of the baby was 3 kg at 4 months age. Haemogram showed anaemia so baby received 100 ml of blood transfusion. Urine examination was normal. C.T. Scan of nasopharynx showed palatal cleft occupied by a soft tissue shadow with 2 dens radio opaque structures. (Fig 1).

A provisional diagnosis of nasopharyngeal teratoid was done and excision was planned. After general anaesthesia and smooth orotracheal intubation, mouth gag used for cleft palate surgery was applied to expose the mass. Portion of the mass near posterior pharyngeal wall was hard while anterior portion near alveolar margin was very soft. Mass had a wide base without any pedicle. The anterior portion was first aspirated to rule out presence of any CSF but it was a dry tap. Posterior hard portion was snared out and rest of the mass was removed completely in piecemeal by dissection. Bleeding was not troublesome. A large palatal defect became evident after the removal of mass. Postoperative period was uneventful except for first 24 hours when orotracheal tube was kept in place as baby was getting breathing difficulty and bradycardia after tube removal due to tongue fall.

From 5th day onwards oral feeding was started using a dental plate to occlude the palatal defect. The histopathology report turned out to be teratoid of nasopharynx. (Fig. 2 and 3).

Fig. 1. C.T. nasopharynx showing soft tissue mass with dens radio opaque shadow in it and cleft in the palate.

Discussion

Teratoid tumours are not the true neoplasms but developmental anomalies in totipotential cells which proliferate, differentiate and form disorganised and disorderly conglomerates of various tissues (Choudhary et. al. 1978). Ewing (1940) recognised 4 different varieties of teratoids.

1. Dermoid or Hairy polyp :- These are bidermal in origin and commonly present at birth and are rarely seen in grown ups. (Resta et. al. 1984).
2. Teratoid Tumours.
3. Teratoma.

Both these have a tridermal origin and the mass contains fat, muscle, cartilage, bone, teeth and acenous glands.

According to Payling Wright (1954) teratoid masses arise due to developmental error in early embryogenesis and are usually apparent at or immediately after birth and grow more or less concurrently with the bearer.

Teratomas are however true tumours (Willis 1953) or neoplasms composed of multiple tissues of kinds foreign to the part in which they arise. Gonads are the prime sites for teratomas.

4. Epignathi :- These tumours form well formed organs and limbs of a parasitic foetus.

The teratoid tumours usually present at or soon after birth. However Badrawy et. al. (1973) reported it at 5 years of age and Didesta et. al. could report it in 14 year old female and even in 71 year old male by Resta et. al. Females are more commonly affected than males (Calcaterra 1969).

The clinical features depend upon size and location of the tumour which include dyspnoea, suffocation, feeding difficulty, nasal obstruction. Foxwel and Kelham (1953) reported large tumour protruding through the mouth. G.S. Kohli et. al. (1987) reported

Fig. 2. Photomicrograph shows well differentiated stratified squamous epithelium & appendage. (H & E x 16).

Fig. 3. Photomicrograph shows mature Cartilage.
(H & E x 16).

a pedunculated hairy polyp causing respiratory distress
in a new born.

The treatment depends upon the severity of symptoms and whether it is pedunculated or sessile. Excision is the treatment of choice. Pedunculated ones are easily avulsed, while the sessile ones with broad base can be completely removed although piecemeal without much bleeding as in our case.

The other aim of treatment is maintenance of airway. Tracheostomy as far as possible should be avoided as it interferes with the development of larynx and at times decanulation at very early age is difficult.

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