Case Report

Congenital pharyngeal teratoma associated with malposed palatine teeth

(A case report)

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Abstract

Pharyngeal teratomas are rare. We present a mature solid teratoma (so called "hairypolyp") involving naso- and oro- pharynx in a female infant who presented with a gradually enlarging mass at the roof of the mouth since birth. The pharyngeal mass was protruding into the mouth through a palatine defect present posteriorly which was removed completely. Subsequently she developed malposed anterior palatine teeth and by 15 months of age three of them were extracted. No residual or recurrent tumor wasdetected by CT scan.

Key words: **Teratoma, Hairy polyp, Pharynx, Malposed teeth, Palate**

Introduction

Teratomas are the most common congenital tumors. They are true neoplasms originating from pluripotent cells and are composed of tissues from all three germinal layers. Teratomas of head and neck are exceedingly rare and about 10% of teratomas are found in this area; nasopharynx and cervical region being the most common sites (1-6). Even if histologically benign, nasopharyngeal teratomas can cause considerable morbidity and mortality because of their location (6, 7). We describe a case of congenital mature teratoma of naso- and oro- pharynx protruding into oral cavity who subsequently developed malposed teeth in anterior palate in addition to the normally placed anterior teeth. To the best or our knowledge, this association is not previously reported.

Case Report

A 45-day-old girl, the product of full term pregnancy born to a 24-year-old mother (G1 P1) by Cesarean section with birth weight of 3000 grs. The prenatal and postnatal courses were

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uncomplicated and no mass was detected at birth. She was noted to have a mass at the roof of the mouth (Figure 1) a few days after birth with gradual enlargement causing impaired feeding and respiration. She was referred to Mofid Children's Hospital. Examination revealed hypertelorism and defective palate with a large 5-cm mass protruding through the defect. CT of head and neck demonstrated a soft tissue tumor in oro- and naso- pharynx with a focus of calcification resembling a tooth (Figure 2). CT of brain was normal and no other abnormalities were noted. The mass was completely removed.

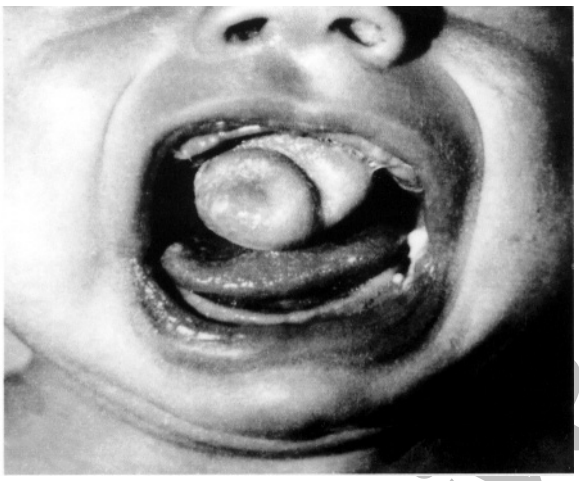

Figure 1: Gross appearance: a mass protruding from palate

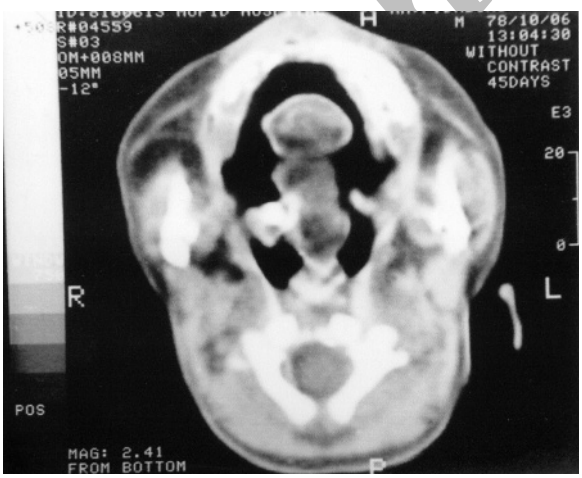

Figure 2: CT scan

The resected tumor was a 5.5x2x1.5 cm elastic-firm tan tongue-like tissue with smooth surface covered on one side with numerous fine and coarse hair (oral side). A 1.5x1.5x0.7 cm nodule was present at the periphery containing a fully developed single-root tooth as well as a transparent 1cm mucin-containing cyst. Microscopic examination showed a disorganized combination of mature mucin secreting glands, neural tissue, adipose tissue, skeletal muscle, and bone covered with skin (Figure 3). No immature component was present. The patient's parents refused to do genetic studies.

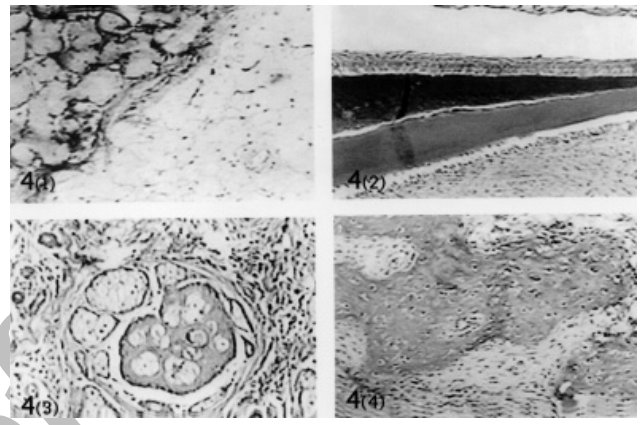

Figure 3: Microscopic appearance (H&E, X 200):
(1) mucin secreting glands and mature fat
(2) tooth structure
(3) skin adnexae
(4) bone trabecula

Postoperative course was uneventful with improvement of respiratory and feeding problems. When she was referred to the hospital at age 15 months, in addition to her normally placed anterior teeth, she had already developed

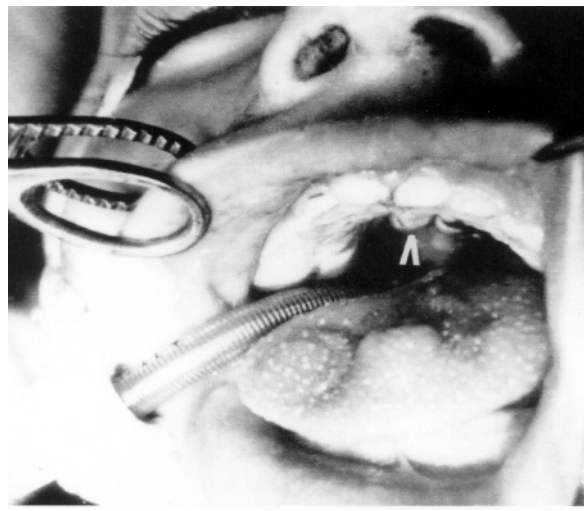

Figure 4: Malposed palatine tooth (arrowhead)

two malposed teeth in the palate which were extracted in the small city where she lived. On physical exam she was well nourished and well developed. The palatine defect was reduced in size dramatically and another tooth was present in anterior palate (Figure. 4). Nasal passage was patent. CT scan revealed no evidence of residual or recurrent tumor.

Discussion

A female infant with congenital mature pharyngeal teratoma is presented who subsequently grew malposed palatine teeth. Teratomas are the most common congenital tumors. About 50% occur in sacrcocoxygeal region. Ovaries, testes, mediastinum and neck are other common sites (6). Head and neck teratomas are relatively rare comprising about 10% of teratomas (1-7). In oral cavity, they are usually congenital and extend through cleft palate from the pituitary area via Rathke's pouch. Some rare reported locations of teratoma include nasal cavity (3), tonsil (8) and auricle (9). Teratomas are more common in female (1, 2, 4, 10). When present during early childhood, they are usually benign (1, 5). As most reported cases, our patient is female and the tumor is benign.

Tharrington et al presenting a case of nasopharyngeal teratoma in his review of 850 patients with teratoms, only one had nasopharyngeal and two had nasal-nasopharyngeal tumors (2, 6). So-called hairy polyp (dermoid) is a teratoid lesion and was first described by Brown-Kelly in 1918 (11). It is described in nasopharynx and oropharynx (12) as a solid polyp lesion covered by skin with hair and sebaceous glands and consists mainly of fibro adipose tissue, vascular tissue, foci of smooth and striated muscle, bone or cartilage and glandular tissue (12). In our case the tumor was composed of a fully developed single-root tooth and disorganized mixture of mature mucin secreting glands, neural tissue, adipose tissue, skeletal muscle, and bone covered with skin. In infants, nasopharyngeal teratoms can present with a variety of signs and symptoms, but most are related to upper airway obstruction (3, 7, 8, 10, 13). Other modes of presentation include dysphasia and failure to gain weight or simply an obvious mass (6). Our case was noted to have and oral mass causing respiratory and feeding problems. Teratomas have higher incidence of maternal polyhydramnios, preterm birth, need for emergency airway management and associated congenital abnormalities (7). Associated congenital anomalies reported in the literature include inguinal hernia, umbilical hernia, hydrocele, cleft palate, lobulated tongue, lingual hamartoma and pituitary duplication (7, 14-16).

This case is interesting, because there is no previous report on association of congenital pharyngeal teratoma and malposed palatine teeth; the tooth growth is most likely due to misplaced tooth buds. Since nasopharyngeal teratomas can cause considerable morbidity and mortality because of their location (6, 7), careful examination of the newborns may reduce the chances by early detection and surgical intervention, particularly when the intra oral tumor is too small to cause any signs or symptoms at birth, as in our case.

knowledgement

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