Oral Mature Teratoma: Report of a Rare Disease

Oral Matür Teratom: Nadir Bir Hastalık

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ABSTRACT

Teratomas arising from the oral cavity are rare. Most cases appear in the midline and paraxial locations; the sacrococcygeal region being the most common site (40% to 60%). Only 10% of teratomas originate from the head, neck and central nervous system. Oropharyngeal teratomas represent 2% of all teratomas. They develop in neonates and are associated with feeding difficulty and airway obstruction. Although some tumors consist of immature elements, teratomas of the head and neck are mature in nature, and surgical excision is the most effective treatment. We present a neonate with a polypoid mass originating from the anterior hard palate and protruding out of the mouth. The mass, which was completely excised, was histologically identified as a mature teratoma.

Keywords
Teratoma; palate, hard; feeding behavior

ÖZET

Oral kavite teratomları oldukça nadirdir. Hastaların çoğunda orta hatta ve paraksiyal lokalizasyonda olup sakrokoksigeal bölge en sık yerleştiği bölgedir (%40 ile %60). Teratomların sadece %10'u baş, boyun ve santral sinir sisteminden gelişir. Orofarengeal teratomlar tüm teratomların %2'sini oluşturur. Yenidoğanlarda beslenme zorluklarına ve hava yolu obstrüksüyonlarına sebep olurlar. Bazıları immatür hücreler içerse de baş ve boyun teratomları matür karakterde olup eksizyon en önemli tedavi yöntemidir. Bu çalışmada sert damağın ön kısmından kaynaklanan ve ağız dışına taşan polipoid kitlesi olan bir yenidoğan olguyu sunuyoruz. Tamamen çıkarılan kitlenin histolojik sonucu matür teratom olarak rapor edildi.

Anahtar Sözcükler
Teratom; sert damak; beslenme davranışı

Çalışmanın Dergiye Ulaştığı Tarih: **14.03.2009** Çalışmanın Basıma Kabul Edildiği Tarih: **22.05.2009**

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INTRODUCTION

Teratomas are derived from multiple tissues foreign to the organ or site from which they arise. They are the most common extragonadal germ cell tumors of childhood and best defined as benign tumors that contain all three germinal layers: ectoderm, mesoderm and endoderm.[1-3] Teratoma is a rare lesion that occurs in approximately in 1:4000 births.[4] Most cases appear in the midline and paraxial location; the sacrococcygeal region is the most common site (40% to 60%).[4] Two to 10% of cases are found in the head and neck region,[4,5] especially at the cervical and nasopharyngeal regions. Although there is a 6:1 female to male preponderance in other sites, they occur equal in both genders in the head and neck region.[6] Of the head and neck teratomas, pure oral presentation is rare.

We report a neonate with a teratoma originating from the hard palate and causing feeding problems. Local Ethics Committee approval was obtained.

CASE REPORT

A 3300-gr male infant born of a 32-year-old mother by vaginal delivery after 37 weeks of gestation presented with a pedunculated mass protruding out of his mouth (Figure 1). The mother had not had ultrasonographic examainations during her pregnancy because of sociocultural reasons. On initial examination, the baby was only two hours old and the mass was protruding outside of the mouth. The baby was not able to suck his mother's breast. The mass was 5x4 cm in diameter and originated from the anterior hard palate, near the midline. The mass was attached to the oral cavity with a peduncle 15 mm in diameter. It hampered feeding without causing respiratory distress. Nasal and nasopharyngeal endoscopy was normal. There were no associated abnormalities. We did not obtain fine needle aspiration biopsy, computerized tomograpy or magnetic resonance imaging. Our preoperative diagnosis was epulis. An excisional biopsy was planned. The mass was totally excised and bleeding was controlled under local anesthesia on the first day. After the surgery, the hard palate remained intact. We did not use a feeding tube because the baby started to suck his mother's breast after the surgery and he was discharged from the hospital on the second postoperative day. Histopathologic examination revealed a mature teratoma (Figure 2). The patient showed no signs of recurrence in a follow up period of one year.

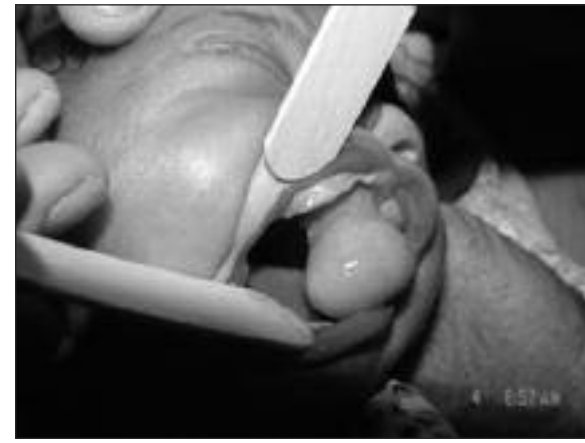

Figure 1. The neonate with oral teratoma.

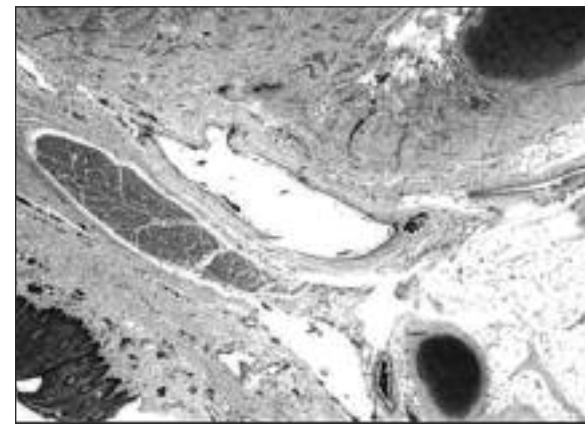

Figure 2. Histopathologic examination showed a benign tumour which included skin, epidermal appendix, cartilaginous tissue, lipomatous structure and muscle tissue (HE X40).

DISCUSSION

Congenital teratomas, which occur in 1 of 4000 live births, are diagnosed predominantly in female neonates.[4,7] Most cases appear in the midline and paraxial locations, the sacrococcygeal region being the most common site (40% to 60%).[4] Only 10% of teratomas originate in the head, neck, and central nervous system.[8] Oropharyngeal teratomas represent 2% of all teratomas.[9,10] Teratomas arising from the oral cavity are rare in the newborns; only 13 cases have been reported in the literature.[11]

Teratomas usually develop from primordial germ cells and are composed of tissues foreign to the site of involvement. There are at least three hypotheses for the origin of teratomas: parthenogenesis, incomplete twinning, and totipotent somatic cell origin.[12] Although a teratoma is clinically defined as having three embryologic

layers, recent classifications also include monodermal types. Head and neck teratomas are classified into four types: dermoid which contains epidermal and mesodermal elements is the most common type; teratoid which consists of the ectoderm, mesoderm, and ectodermal elements is poorly differentiated; true teratoma which contains all three germ cell layers differentiates into a recognizable early organ; and epignathus which is highly differentiated into recognizable organs or limbs is very rare and associated with a high mortality rate. It is suggested that the lesion developed by pinching off the remnants of three germinal layers considered to be marginated in the normal migratory pathway as the structures of the head and neck developed. Malignant transformation of teratoma occurs in approximately 20% of cases and is more common in adults. Germ cells in malignant teratoma are bizarre collections of the epithelial and mesenchymal tissues. The manifestation of these neoplastic lesions is aggressive local infiltration; many metastasize.[13]

The prenatal diagnosis of tumors of the head and neck is very important. Early diagnosis enables a carefully planned delivery during which an open airway can be maintained to improve perinatal outcome.[14] It should be kept in mind that a large obstructing mass may be fatal, even if it was diagnosed prenatally.[15] Because airway obstruction can cause severe complications, Cesarean section is the preferred method of delivery for infants with an obstructing mass of the head or neck. Immediately before the termination of fetomaternal circulation, these infants should undergo intubation by the oral or nasal route. Some neonates with a huge obstructing mass may require tracheostomy in the delivery room.[14]

Children with pure oral teratomas seem to have less dramatic respiratory behavior compared to children with other head and neck presentations. When oral teratomas grow, they tend to protrude outside of the mouth, rather than posteriorly toward the oropharynx. Because newborns are obligate nasal breathers, obstruction of the oral compartment is relatively less important and usually causes less urgent feeding problems.[12] In addition, children with oral teratomas have been reported to have associated abnormalities including cleft palate, cystic hygroma,[16] and other multifocal teratomas.[17,18] In our case, no associated anomalies were found.

Although radiologic imaging with ultrasonography, computed tomography, or magnetic resonance imaging and testing for the level of α-fetoprotein are helpful for identifying teratomas, the diagnosis must be based on histologic examination. In our case, preoperative diagnosis was epulis, but histopathology revealed a mature teratoma. Surgical excision is the treatment of choice for oral teratomas. A review of the literature indicated that total resection could be performed in most patients, and recurrence after that procedure was rare. Congenital teratomas are generally mature in nature.[8]

Oral teratomas are extragonadal germ cell tumors that occur very rarely during infancy and childhood. Children with oral teratomas have less severe symptoms when compared to those with oropharyngeal, nasopharyngeal and cervical teratomas. Although all reported pure oral teratomas are benign, there is a risk of malignant change, and long-term follow-up is mandatory even if the tumor is totally excised with free margins. The prenatal diagnosis of tumors of the head and neck is very important. Early diagnosis enables a carefully planned delivery during which an open airway can be maintained to improve perinatal outcome. It should be kept in mind that a large obstructing mass may be fatal, even if it was diagnosed prenatally. Therefore, all mothers must have ultrasonographic examainations during their pregnancy.

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