Congenital Large Oropharyngeal Immature Teratoma

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Abstract

Epignathus is a congenital tumor and a rare type of teratoma. It is associated with midline abnormalities. In our case the teratoma was of oropharyngeal origin, extending to cervical region, inhibiting the growth of maxillary and mandibular bones and tongue. No treatment could be applied. Although there are few cases of epignathus reported to be treated with ex-utero intrapartum treatment procedure, there are no reported cases in the literature with totally undeveloped bone and soft tissue of lower and upper jaws as in our case.

Keywords: Epignathus, oropharyngeal mass, maxillo facial deformities.

Konjenital büyük orofaringeal immatür teratom

Epignathus konjenital bir tümördür ve nadir görülen bir teratom türüdür. Orta hat anomalileri ile ilişkilidir. Olgumuzda teratomun orofarenks kaynaklı olup servikal bölgeye kadar uzanım gösterdiği, maksiller ve mandibular kemiğin ve dilin gelişimine engel olduğu saptandı ve tedavi yapılamadı. Literatürde eksutero intrapartum tedavi prosedürü ile tedavi edilebilen az sayıda epignatus olgusu bulunmasına rağmen olgumuzda olduğu gibi alt ve üst çeneye ait kemik ve yumuşak dokunun tamamen gelişmediği bir olguya rastlamadık.

Anahtar Sözcükler: Epignathus, orofaringeal kitle, maksillo fasiyal deformiteler.

Background

Teratomas are congenital germ cell tumors that contain tissues of variable maturity and have a known malignant potential which is unpredictable from their histological features or stage of development. Teratomas occur in approximately one in 4000 live births, show a female preponderance, and have an 18% risk of other congenital malformations, some of which can be incompatible with life.[1] The most common sites of origin of teratomas in children are the sacrococcigeal region, gonads and mediastinum. Epignathus teratoma represents a rare group of congenital neoplasms in the head and neck. It has an incidence of 1/35.000-1/200.000 births. The clinical presentation of this tumor varies depending on size and location in the oronasopharynx.[2] In utero a tumor occluding the foetal airway may not cause many significant problems because the foetus is perfused by the umbilical cord and placenta, but it may impede foetal swallowing leading to the accumulation of amniotic fluid or 'maternal polyhydramnios'.[1] Epignathi that arise from the palate or pharynx and protrude

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from the mouth result in life-threatening airway obstruction and usually cause asphyxiation shortly after birth.[3] The large oropharyngeal teratoma can cause airway obstruction and neonatal mortality.

Case

The patient was a 20-year-old healty woman pregnant to her first child with a gestational age of 25 weeks. Questionning the family history it was learned that all the family members were healthy. Foetal heart activity was observed in three dimensional ultrasonography. Foetal measurements were compatible with 25 weeks. Amniotic fluid index was increased. A mass lesion of 73x76 mm, originating from the facial and neck region, containing solid and cystic components was observed (Figure 1). The mass lesion was just below the eye level of the foetal face so the nose and the mouth could not be observed and foetal magnetic resonance imaging (MRI) was performed. In the MRI a7x8x11 cm smooth lobulated contoured, septated multicystic mass lesion with a fibrous capsule occupying the anterior maxillofacial and cervical region was detected. The mass lesion was unrelated with the cervical spinal canal and and intracranial area (Figure 2). Bilateral orbital development was normal. Bone and soft tissue of lower and upper jaws were not developed. The family was informed about the situation. The surgical procedures to be performed after the birth because of the nondevelopment of maxillary and mandibular bone and soft tissue due to the teratoma were consulted with plastic and reconstructive surgerions and paediatric surgeons and the family was informed. As a result of the consultations it was learned that a functional jaw would be unable to be reconstructed in this case and the extrauterin intrapartum treatment procedure after birth would be difficult. As the mass lesion extends to cervical region respiratory and digestive tracts were completely closed and the baby would face breathing and nutrition problems. Family was informed about these problems. The pregnancy was terminated via vaginal route with the family's decision. The autopsy of the foetus revealed that the mass lesion with a dimension of 7x8x11 cm was occupying the whole oropharynx and was unrelated with any of the oropharyngeal structures but the mass was extending to cervical region. Cervical tissue and organ development was normal. The upper border of the mass was adjacent to the lower border of the orbital bone but it was unrelated with the sphenoid bone. Maxilary and mandibular bones and the tongue were absent, there were some cartilage remnants which were thought to be belonging to the jaw (Figure 3). The mass was shown in Figure 4 macroscopically. The mass was diagnosed as immature teratoma histopathologically (Figure 5).

Discussion

Teratomas are composed various tissues of ectodermal, endodermal and mesodermal origin.

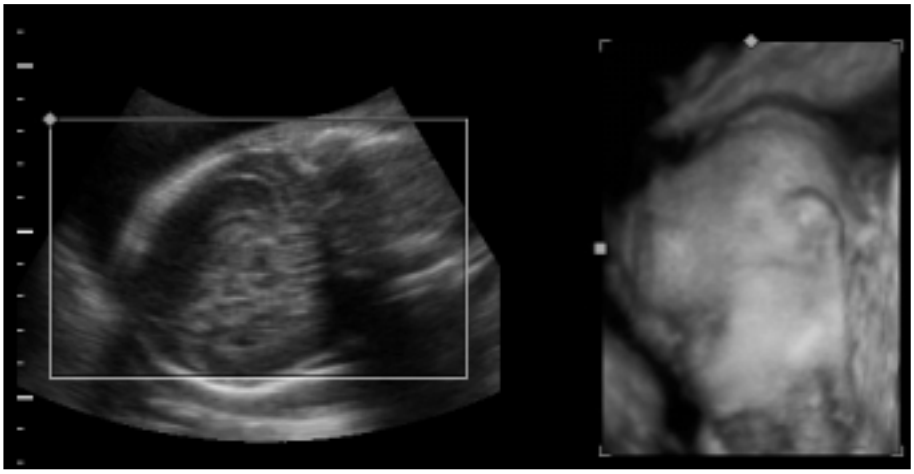

Figure 1. Ultrasound view of the foetus.

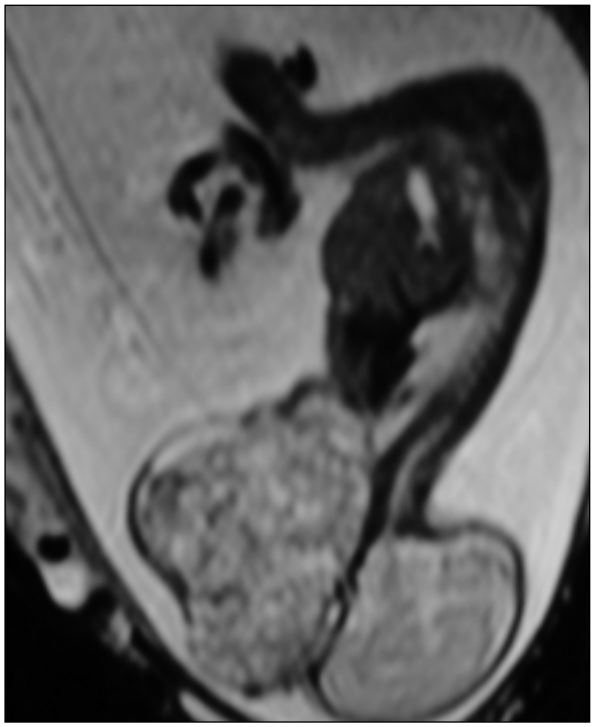

Figure 2. MRI view of the 7x8x11 cm mass occupying the maxillofacial and cervical region.

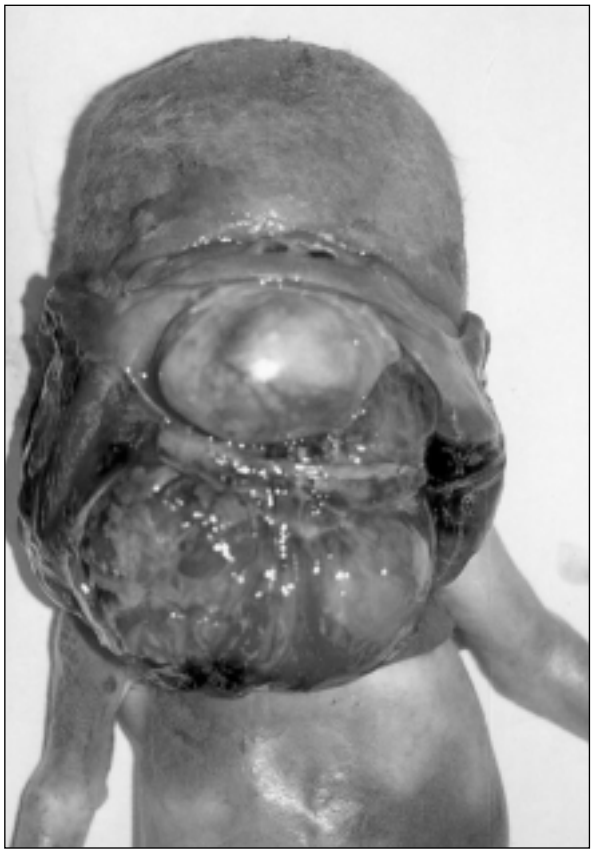

Figure 3. View of the dead born foetus.

These tissues exhibit various degrees of maturation.[3] They are classified in four groups as dermoid cyst, teratoid cyst, teratoma and epignathus. Dermoid cyst derives from endodermal and mosedermal germ layers. Tumors composed of all three germ layers that are poorly differentiated are called teratoid cyst and those which are well differentiated are called teratoma. Epignathi are oral tumors containing foetal organ and structures.[4]

Epignathus teratomas are rare congenital tumors associated with midline abnormalities. They have an incidence of 1/35.000-1/200.000.[5] Epignathus teratomas are normally nonfamilial. It is 3 times more prevelant among women.[5] The reason of this predominance is yet unclear. Epignathus teratomas are more common among the babies of young mothers as in our case. Mean age of the mother is 24.6.[5] There are no findings suggestive of an environmental factor or a karyotypic abnormality.[3]

(Vandenhaute) The aetiopathogenesis of epignathus is unclear. According to a theory, it developes due to insufficient fusion of midline tissues during the embriogenesis at the 7-9th gestational weeks.[4-5] Another theory suggests that uncontrolled growth of primordial germ cells in improper places with the lack of regulation effects of adjacent cells and factors.[4] In the recent studies epignathus teratomas were reported to have a neuroectodermal tissue dominance although cells of three germ layers are seen, as seen in our case.[3]

Histopathologically 51% are mature teratoma, 49% are immature teratoma, 5.8% are yolk sac tumors.[3,6] Our case was reported as immature teratoma histopathologically.

In a review of Isaac Jr[6] about the perinatal germ cell tumors, it was mentioned that, in the literature 16 cases with hard palate origin, 14 cases with nasopharyngeal origin, 6 cases with sphenoid origin and 6 cases with oropharyngeal origin were reported.

In our case intraoral teratoma of oropahryngeal origin was present. Although most of the cases

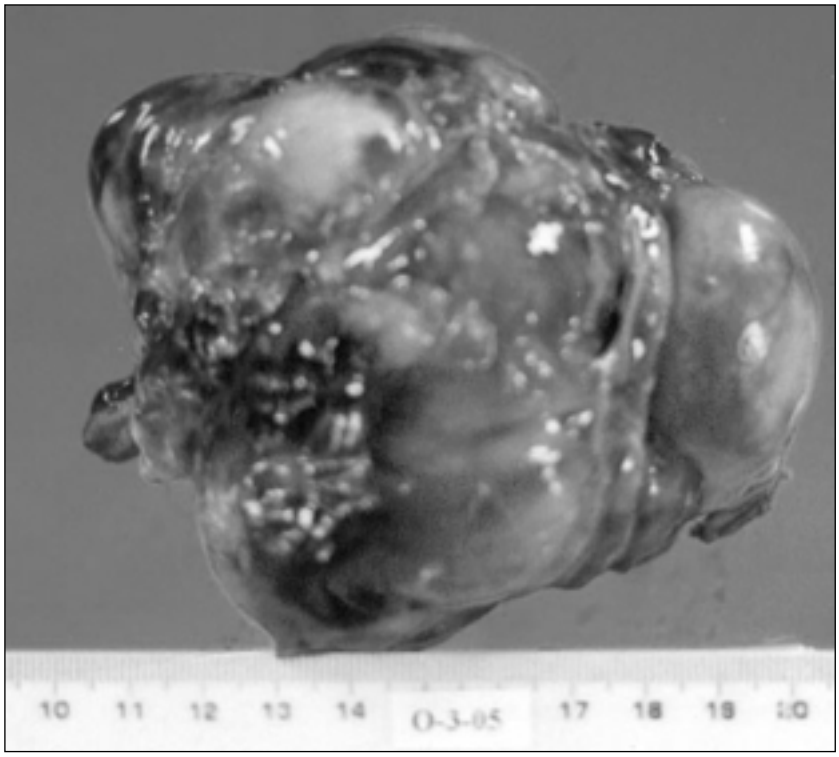

Figure 4. Macroscopic view of the mass.

were reported to originate intraorally, the classical epignathus tumor is of jaw or alveolar bone origin.[7] According to the settling place, head-neck teratomas with palatal origin are called epipalatus, those with sphenoidal origin are called episphenoid, those with cranial origin are called epicranium.[4] Most of the epignathus tumors have a binding place to the cranial base in the posterior nasopharyngeal region. Although most of them are related with hard palate and sphenoid bone, there are some developing from midline or laterally.[5] In our case the mass was not related with any oropharyngeal region.

The clinical presentation is associated with the settlment place and the size. Usually a great epignathus tumor fills the mouth and protrudes from the oral cavity. A small one is usually pediculated and may localize anywhere in the oropharyngel region. A huge oropharyngeal mass compresses and replaces the normal tissues in the lower part of the face and jaw and frequently causes maxillary deformity.[5] The teratoma of our case was localized intraorally. The mass was not protruding from the mouth, however, it was extending to the cervical region and additionally, maxillary and mandibular bony structure and the tongue was not present. To our knowledge, there are no cases of oropharyngeal teratoma impeding the development of maxillary and mandibular bone and the tongue.

Cranial base involvement of teratoma may extend both intracranially and extracranially like a sand watch.[4,5] There exists a craniopharyngeal extension in some cases. Intracranial extension is often fatal.[5] Intracranial extension shall be suspected in case of sphenoid base involvement and be confirmed by CT or MRI.[4,5] In our case there were no sphenoid bone involvement and MRI scans revealed no intracranial extension, that should be a sign in our favor about the prognosis of the foetus.

As in our case, maternal polyhydramniosis can be seen due to oropharyngeal obstruction by the teratoma in the intrauterin period and difficulty in swallowing or due to eosophageal compression.[3,4]

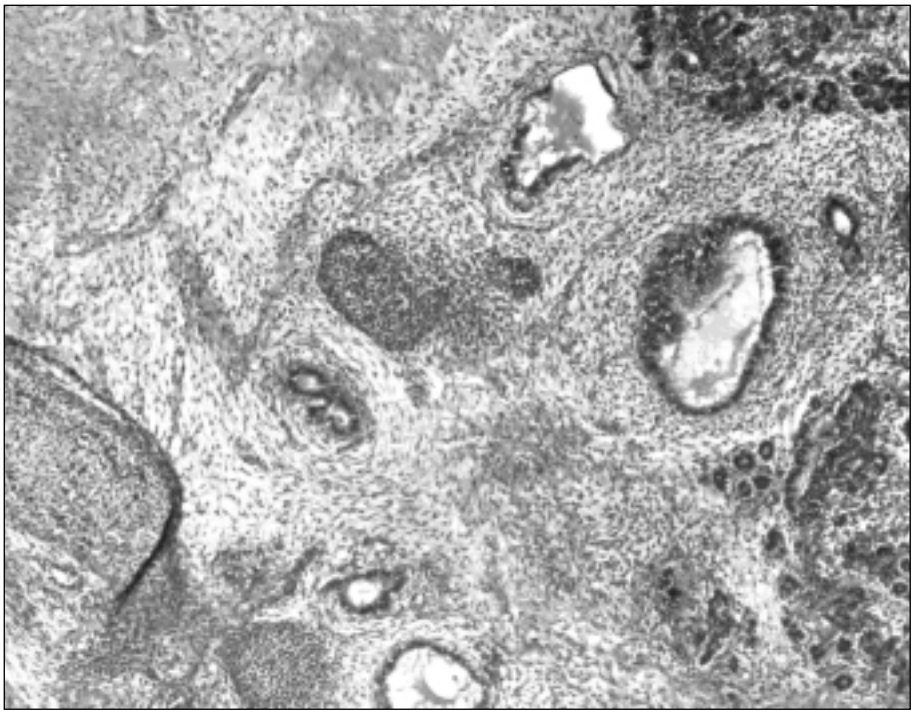

Figure 5. Histopathologic view of the mass compatible with immature teratoma.

Epignathus can be diagnosed antenatally by ultrasound and in appropriate cases multidiciplinary treatment can be applied.[5] Antenatal diagnosis was achieved in our case, however, because of nondevelopment of maxillary and mandibular bones and the tongue, impossibility to reconstruct these organs surgically the pregnancy was terminated with the decision of the family.

Embrional rabdomyosarcoma of the tongue, retinoblastoma, nasal glioma, heterotopic thyroid, cystic lymphangioma, nasoethmoidal meningoencephalocele, sphenoid meningoencephalocele and giant epulis shall be considered in the differential diagnosis.[5]

Although rarely seen, head and neck teratomas are emergent cases during the neonatal period due to airway obstruction and high mortality rates in infants.[3] Usually, those diagnosed antenatally have 3 folds higher mortality than those diagnosed neonatally.[6]

Multiple surgical procedures can be needed to obtain an optimal result in large tumors. Intracranial extension must be excluded before the surgery.[6] In case of hydrocephaly or intracranial mass in a neonate with epignathus, surgery is avoided because the prognosis is poor.[8] No recurrance was reported after complete resection.[5]

In the exutero intrapartum treatment procedure, during the ceserian sectio of the term foetus, airway patency is achieved with intraoperative intubation or tracheostomy just after the foetus leaves the uterus and before the umblical cord is cut, then the maternofoetal circulation is ceased and the birth is completed.[7]

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