



Giant Epignathus (Teratoma of Palatine Tonsil): A Case Report

Case Report

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Abstract

Teratomas are benign tumours containing tissues derived from ectoderm, endoderm and mesoderm. Epignathus is a rare congenital teratoma and originates from oropharyngeal region. We present a case of giant epignathus arising from tonsillar region in a neonate. A male neonate that was born with a 38-week cesarean section presented with a pedunculated mass from left tonsilla palatina and protruding outside the mouth. The patient did not have any airway problem. Magnetic resonance imaging and computed tomography scan showed no intracranial extension. The patient was operated on the postpartum 3rd day and the mass was excised successfully. After histopathological examination, mature teratoma was diagnosed. During post-operative 6 months control visit, there was no recurrence. Epignathus is a rare congenital oropharyngeal teratoma, it should be diagnosed in the fetus as early as possible. Teratomas of the tonsilla palatina are extremely rare. In such cases, the mass may cause airway obstruction and feeding difficulties so complete resection is curative in most cases during the early neonatal period.

Keywords: Epignathus, teratoma, fetal anomaly, palatine tonsil, pediatric otorhinolaryngology, case report

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Introduction

Teratoma is composed by multiple tissues derived from the three germ cell layers (ectoderm, endoderm and mesoderm). The incidence of teratomas is 1:4,000 live births. (1). Teratoma can be seen almost anywhere in the body. The clinical presentation varies depending on the size and location of the lesion. Epignathus is a term commonly used for teratomas originating from the oropharyngeal region such as jaw, palate or pharynx (2).

The incidence of epignathus is 1:35,000 to 1:200,000 in live births. Epignathus that arise from tonsillar region is very rare (1).

In this case report, we present a rare case of giant epignathus with different localization in a male neonate. The clinical characteristics, diagnosis and treatment of this rare disease are also reviewed.

Case Presentation

A male neonate with a birth weight of 3.8 kg was born by elective caesarean section

delivery at the 38 weeks of gestation. He was presented with a giant mass protruding from the oral cavity. Apgar score of the newborn at the first and fifth minutes of birth was eight. The history of the mother did not reveal polyhydroamniosis although she had no regular prenatal follow-up. Prenatal ultrasonography showed that the fetus had a large mass protruding from its mouth. A caesarean section delivery was preferred because of fetal anomaly. After delivery, the patient did not have any airway problem and did not require oxygen support because the mass did not totally obstruct the baby's oral cavity. The patient was evaluated in the neonatal intensive care unit. Otolaryngological examination revealed a mass covered by skin and hair, attached to left palatine tonsil with a peduncle, extending the buccal mucosa and protruding from oral cavity. In radiological evaluation with magnetic resonance imaging (MRI) and computed tomography, it was observed that the mass did not compress the trachea and had no intracranial extension (Figure 1). The patient was operated under general anesthesia on postpartum 3rd day. Double excisions were performed for complete resection. First, the extraoral part of the mass was excised with double ligation using cautery and suture. Thus, access to the pedicle was provided. The mass removed from floor of the mouth and buccal mucosa, then excised totally with left palatine tonsil (Figure 2). Size of the mass was approximately 18x12x7 cm and it weighed 950 g. The histopathological examination

revealed mature teratoma. In the sections, adnexal structures, glandular formations, common neuroglial elements, choroid plexus formations, adipose tissue, mature structures consisting of muscular tissues and cystic structures lined with multilayered squamous epithelium containing keratin were observed. The routine blood tests and chromosomal analysis were normal.

The patient was followed up for approximately one week in the intensive care unit. He was fed through a nasogastric tube for a while. He was discharged on the seventh postoperative day and oral feeding began on the postoperative second month. On a follow-up period of 6-months, there was no recurrence of the epignathus. The patient was examined by pediatricians and his growth and development was normal (Figure 3).



Figure 2a. The giant mass protruding from oral cavity b. Surgical specimen (measuring 18x12x7 cm)

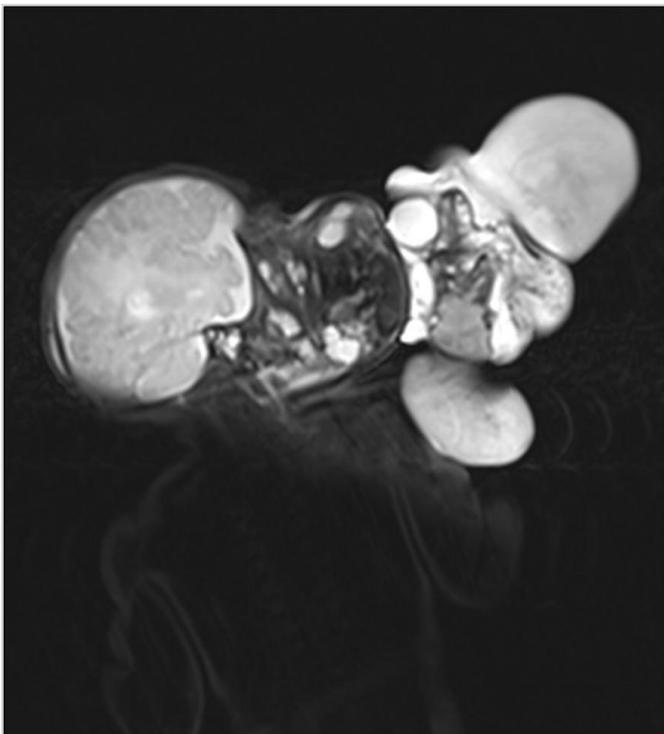


Figure 1. The preoperative T2 weighted MRI coronal section of the mass. Heterogeneous mass with cystic and solid components that associated with tonsillar and buccal region, protruding from oral cavity

MRI: Magnetic resonance imaging

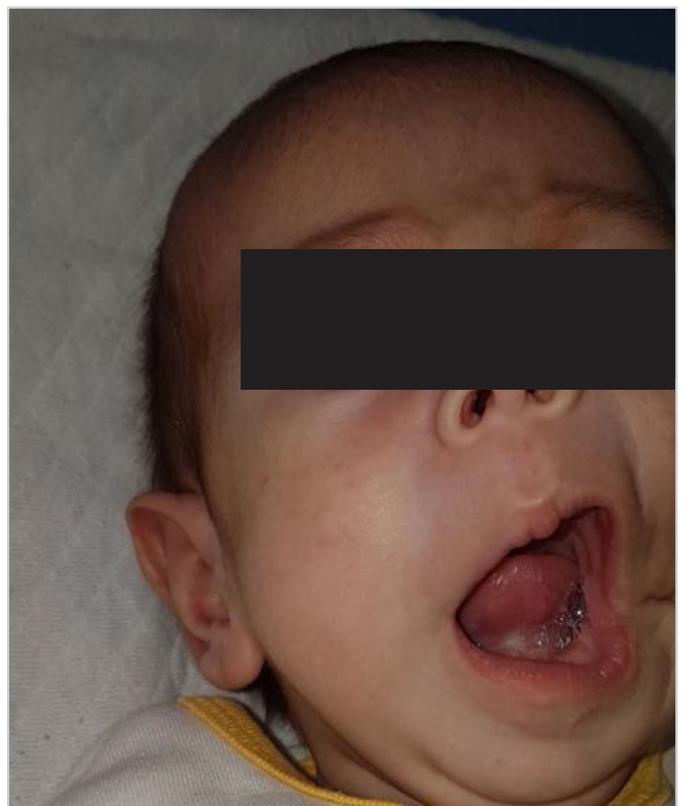


Figure 3. Patient at six months of age, with minimal facial deformity

Discussion

Teratomas are among the most common extragonadal germ cell tumors presenting in childhood and contain tissue from all three embryonic germ layers; ectoderm, endoderm and mesoderm. They are often located in the midline and paraaxial regions. They are most commonly seen in the sacrococcygeal region, anterior mediastinum, testis, ovary and retroperitoneum (2, 3).

Teratomas are classified into four types: Dermoid (hairy polyps), teratoid, true teratoma and epignathus. In terms of differential diagnosis, the hairy polyp presents at birth as pedunculated polypoid mass. It arises from naso-oropharynx mostly, and histologically composed of derivatives of ectoderm and mesoderm (4). However epignathus is a teratoma that arises from oropharyngeal region and includes tissues from ectoderm, mesoderm and endoderm.

The etiology of epignathus is still unclear. The most popular theory attributes its origin to disorganized growth of pluripotential cells in the region of Rathke's pouch (5). There is no evidence suggesting that epignathus is caused by environmental agents, Mendelian or polygenic inheritance (6).

Teratomas contain ectoderm, endoderm and mesoderm layers. In addition, teratomas can be mature, immature or mixed tumors. In many cases, the presence of immature tissue indicates a diagnosis of malignancy (7). In our case, a mature teratoma was diagnosed. Mature teratomas are thought to have a benign character.

In recent years, the use of ultrasonography and MRI in prenatal follow-up for congenital anomalies has increased. MRI is a complementary diagnostic tool for epignathus to detect the relation of the tumor to the fetal airway and intracranial structures (8). There was no intracranial extension in our patient.

Epignathus may be a condition that can require tracheotomy in the neonate, leading to airway obstruction. In our case, the mass of the patient was derived from the tonsillar fossa and extending out of the oral cavity and did not cause respiratory problems, therefore no tracheostomy was needed. Postnatally, early surgical intervention is necessary. Early diagnosis, the establishment of a secure airway, complete excision of the tumor and timely follow-up should increase the survival of newborns with oral teratomas (9, 10).

Conclusion

Epignathus is a rare congenital oropharyngeal teratoma, it should be diagnosed in the fetus as early as possible. Teratomas of the tonsilla palatina are extremely rare. In this case report, we present a rare case of giant epignathus with different localization. Complete excision of the tumor is imperative to prevent relapse, maintain a secure airway and provide normal oral feeding of the patient.

Informed Consent: Consent was obtained from the patient's family.

Peer-review: Externally peer-reviewed

Authorship Contributions

Conception: F.A., Design: F.A., Analysis and/or Interpretation: M.M., M.A.E., Literature Review: M.M., Writing: F.A., Critical Review: M.A.E.

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Main Points

- Epignathus is a term commonly used for teratomas originating from the oropharyngeal region.
- Epignathus that arise from tonsillar region is very rare.
- Teratomas contain ectoderm, endoderm and mesoderm layers. In addition, teratomas can be mature, immature or mixed tumors.
- Epignathus may be a condition that can require tracheotomy in the neonate. MRI is an important imaging test in detecting airway obstruction.
- Epignathus should be diagnosed in the fetus as early as possible.

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