

CASE REPORT

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Congenital epignathus associated with a cleft palate: a case report

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Abstract

Background: Epignathus is a rare, benign, congenital teratoma of the hard palate with an estimated incidence of 1 in 200,000 live births. Epignathus frequently leads to a high mortality (80–100%) due to airway obstruction in the neonatal period.

Case presentation: We report a case of successful management of a rare oropharyngeal teratoma in a African newborn girl who was referred to our institute with a large protruding intraoral mass, combined with cleft palate, causing some respiratory compromise. The palatal mass was removed on the fifth day after birth, and a palatoplasty performed on day 30.

Conclusion: Epignathus is a life-threatening condition at the time of delivery. Appropriate management begins with securing the airway, followed by complete resection of the tumor.

Keywords: Epignathus, Teratoma, Cleft palate, Airway obstruction

Introduction

Epignathus is a rare teratoma that arises from the oral cavity, most commonly from the palate. Congenital tumors of the oral cavity frequently affect normal development of adjacent structures, and tumors developing during morphogenesis of the palate are commonly associated with a cleft palate [1].

In general, teratomas are uncommon malformations that arise in the three embryonic germ layers (ectoderm, mesoderm, and endoderm). They are felt to be the result of pluripotent cells that did not complete migration from allantois to the genital ridge during the fourth and fifth weeks of gestation [2].

The physiologic impact of a teratoma is a result of its location and size. If located in the upper aerodigestive tract, they are high risk, frequently causing

life-threatening complications from compromise of the airway. Because of this, they have a high mortality rate, and prenatal diagnosis may allow for better preparation to help optimize care at the time of birth [3–5].

Some teratomas extend beyond the mouth, and these lesions can be associated with mid-face defects including cleft lip and palate. There are reports of palatal teratomas extending beyond the oral and nasal cavity and even extending into the cranial cavity with resultant destruction of brain tissue [3, 6].

We have reported a case of congenital teratoma arising from the hard palate in a newborn girl. This report aims to show that diagnosis and successful surgical treatment of such a rare case are possible in a low-resource environment. It therefore contributes to the existing literature on management of giant epignathus.

Case

A 1-day-old female African newborn weighing 3500 g was referred for management of a large oral mass that protruded from the mouth and caused maxillary

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protrusion and flattening of the nose. The patient was having difficulty breathing (Fig. 1).

She was born by spontaneous vaginal delivery to a mother who was unaware of the date of her last menstrual period. She had no prenatal medical care, so there was no opportunity for prenatal diagnosis.

Initial examination revealed a mass that occupied the majority of the oral cavity and protruded from the mouth. On palpation, the mass appeared to arise from one side of the palate, and there was an adjacent palpable cleft. No other congenital anomalies were evident on physical examination.

The patient had some breathing difficulty, but the airway was patent. She was administered oxygen and intravenous fluids, and a nasogastric feeding tube was successfully placed. Feeding was commenced at 35 ml, eight times per day, and she stabilized.

On day 5, she was taken to surgery. Laryngoscopy was performed using a standard Macintosh blade size 1, and an easy intubation with a 2.5 endotracheal tube. The mass was excised under general anesthesia. Good hemostasis was achieved, and complete resection of

the tumor was accomplished. There were no immediate postoperative complications.

Gross examination revealed a large fibrocystic tumor involving the oral and nasal cavities with its base originating from the hard palate. It measured 6 × 5 × 4.5 cm and was well circumscribed (Fig. 2).

Histopathologic examination revealed a congenital primarily epidermal teratoma.

She had an uneventful postoperative course, although a palatal defect remained (Fig. 3). She had no difficulty opening or closing her mouth. Bottle feeding with expressed breast milk was initiated shortly after surgery and was well tolerated. She was able to breastfeed by postoperative day 5.

On postoperative day 30, the patient was taken back to surgery, where she underwent successful palatoplasty (Fig. 4). She was discharged home with her mother on postoperative day 9 after palatoplasty (Fig. 5).



Fig. 1 One-day-old female baby with an oropharyngeal mass

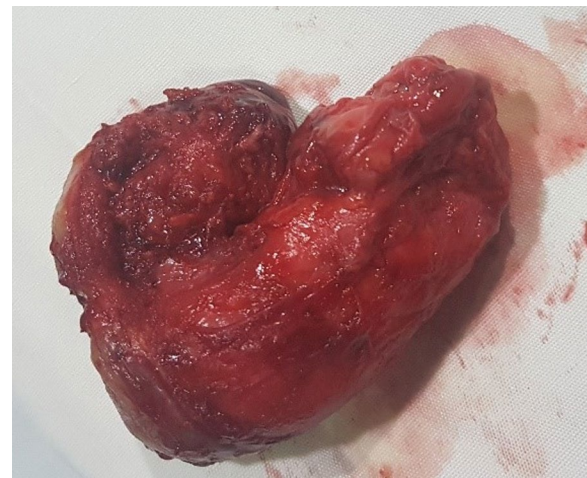


Fig. 2 Tumor after total excision



Fig. 3 Five-day-old female baby, with a palatine defect post removal of palatine tumor

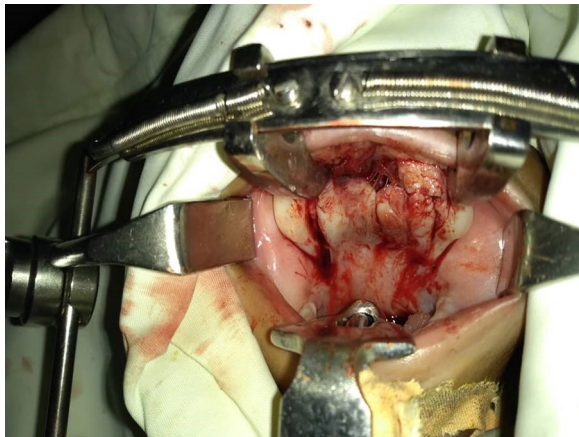


Fig. 4 Reparation of palatine defect (intraoperative view)

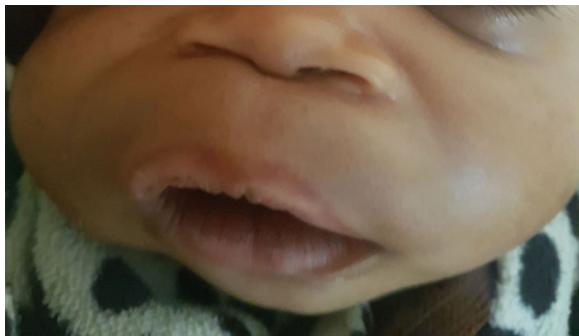


Fig. 5 One-month-old female baby, with a palatine defect repaired

Discussion

Congenital oral tumors are commonly recognized at birth, but they can also be diagnosed antenatally by ultrasound or Magnetic resonance imaging (MRI). They can be solid or cystic and can be benign or malignant, but the majority are benign. *In utero*, obstruction of the oral cavity can result in polyhydramnios [7–9].

Teratomas occur in approximately 1 in 4000 births, and there is a female predilection. They are most commonly located in the sacrococcygeal area but can present in many areas of the body. Epignathus is very rare, occurring in only 1/200,000 live births. They account for only 2% of teratomas and almost always present in neonates. Most are benign, although malignancies have been reported [4–6]. Although other teratomas have a 6:1 female predilection, those of the oral cavity do not seem to have a clear sex predilection. In our case, the baby was female.

As reported by Kumar *et al.*, giant epignathus can lead to death during the neonatal period due to airway obstruction or can lead to malnutrition due to the

inability of oral feeding. The newborn must therefore benefit from oxygenation and feeding by nasogastric tube while awaiting surgery. Complete surgical excision is the definitive treatment [5, 10].

Teratomas are often classified into four types:

- (1) *Dermoid*, also called “hairy polyps”, containing epidermal and mesodermal elements are the most frequent;
- (2) *Teratoid*, composed of ectodermal, mesodermal and endodermal elements, are very poorly differentiated;
- (3) *True teratoma*, also contain the three embryonic germ layers, but differentiated in recognizable structures such as cartilage, hair, teeth, etc.
- (4) *Epignathus*, are highly differentiated, rare, and have a high mortality rate due to their location [4].

Yoshimura *et al.* [11] proposed an alternative classification of teratomas with three types:

1. *Type 1* (skin and fatty tissue derived from two germ layers);
2. *Type 2* (teratoma with tissues representing the three germ layers, with bone, teeth, neural tissue, and tissues of the gastrointestinal tract); and
3. *Type 3* (parasitic twins with well-differentiated organs and limbs).

Histopathologic examination in this case revealed an epignathic teratoma containing mostly epidermal tissue. It was a type I teratoma by the Yoshimura classification system.

Oral teratomas can be associated with mechanical craniofacial abnormalities, the most common being cleft palate [12].

When a tumor develops before the eighth week, it can interfere with the normal closure of the secondary palate in the midline. It can also hinder the confluence of the two halves of the nose or tongue, resulting in a bifid nose or tongue. It can also contribute to abnormal maxillofacial development with micrognathia and ogival palate [13]. In our case, the baby had a nonfusion of the two palatal bones with resultant cleft palate.

Conclusion

Epignathus is a rare oral cavity teratoma commonly presenting with symptoms requiring urgent management of respiratory distress.

As in this case, large epignathus can cause some respiratory compromise, and it is commonly associated with cleft palate, but other anomalies may occur.

Urgent treatment consists of airway management and nasogastric feeding tube. The definitive treatment requires complete resection of the mass and correction of associated anomalies.

Abbreviations

IV: Intravenous; MRI: Magnetic resonance imaging; ENT: Ear, nose, and throat.

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Authors' contributions

MKK: principal surgeon of the case. FLM: report, data collection, and surgical assistant. JNK: ENT surgeon. Other authors: reviewers. All authors read and approved the final manuscript.

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Availability of supporting data

Not applicable.

Declarations

Ethics approval

The medical ethics committee of HEAL Africa Tertiary Hospital approved this study (approval number HA/CEM/MMA/03-2020).

Consent for publication

Written informed consent was obtained from the patient's parent for publication of this case report and accompanying images. A copy of the written consent is available for the review of the Editor-in-Chief of this journal.

Competing interests

The author(s) declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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