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jbpm1@sbpc.org.br, adagmar.andriolo@gmail.com

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Menezes Filho, Marcelo P.; Simão, Nábia Maria M. S.

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# Giant epignathus of the palate: a case report

## *Epignathus gigante do palato: relato de caso*

Marcelo P. Menezes Filho; Nábia Maria M. S. Simão

Faculdade de Medicina de Campos.

### ABSTRACT

Epignathi are congenital malformations, with a high level of mortality due to their obstructive mechanism and respiratory involvement. Most of them are benign; however, cases of malignant epignathus have been described. The possibilities of malignancy increase in the presence of neuroepithelium, an important confounding factor. Epignathus is described in a newborn, besides its clinical and histopathological study. Surgery with complete resection is the definitive treatment. Residual tissue is linked to recurrence, curiously without increasing the risk of malignancy.

**Key words:** teratoma; teratoid severe abnormalities.

### INTRODUCTION

Teratomas are rare malformations originated in the three embryonic germ layers (ectoderm, mesoderm, and endoderm) from pluripotent cells that did not complete migration from allantois to genital ridge during the fourth and fifth weeks of gestation<sup>(1)</sup>. The sacrococcygeal region is their most common site of occurrence, followed by ovaries and testes, retroperitoneum and mediastinum. Teratomas of the head and neck are uncommon, representing 1%-9% of all teratomas, and generally originate in the cervical region. They occur almost exclusively in neonates. Teratomas arising from the hard palate, oral mucosa, tongue, and chin, protruding from the oral cavity are called epignathi<sup>(1)</sup>. Their most important complication is the respiratory compromise, presenting high mortality risk associated with upper airway obstruction. Another commonly present clinical manifestation is the difficult deglutition<sup>(2)</sup>. Although most epignathi are benign, the probability of recurrence is high, with surgical resection being the treatment of choice<sup>(3-5)</sup>. We describe a case of surgically treated giant epignathus, with clinical and anatomopathological study.

### CASE REPORT

A 35-year-old pregnant woman presented with weight loss accompanied by accentuated polyhydramnios. In the

ultrasonography performed at 35 weeks and two days of gestation, a single fetus was identified, in longitudinal lie, cephalic presentation and anterior dorsal position. Cardiac and motor activities were normal. Placenta exhibited anterior implantation, heterogeneous texture and maturity grade I/II in the Grannum classification. Severe polyhydramnios and estimated fetal weight of 2,868 g. Presence of an echogenic mass protruding from the oral cavity, measuring  $7.64 \times 6.99$  cm (**Figure 1**).

At 36 weeks and four days, the patient was scheduled for a cesarean section. The newborn presented respiratory distress, requiring mechanical ventilation. Six hours after delivery, the voluminous mass measuring  $10.7 \times 10.2 \times 8.7$  cm was excised; the palate was ascertained as its site of origin during the surgical procedure (**Figure 2**).

The anatomopathological study revealed a bright oval-shaped elastic-firm tumor mass, slightly bosselated, dull grey with violaceous regions, with a 2.7-cm raw area (**Figure 3**). The sections exhibited surface with multivariate aspect, represented by regions sometimes solid, white and firm, frequently stony hard, sometimes greenish pink and multinodular; coexistence of cavities filled with brownish liquid material or grayish mucoid material (Figure 3).

Histological sections exhibited mature tissues derived from the three germ layers, composed of keratinizing squamous epithelium with skin annexes, brain tissue, gastrointestinal epithelium, pulmonary tissue and mesenchymal elements, including fatty

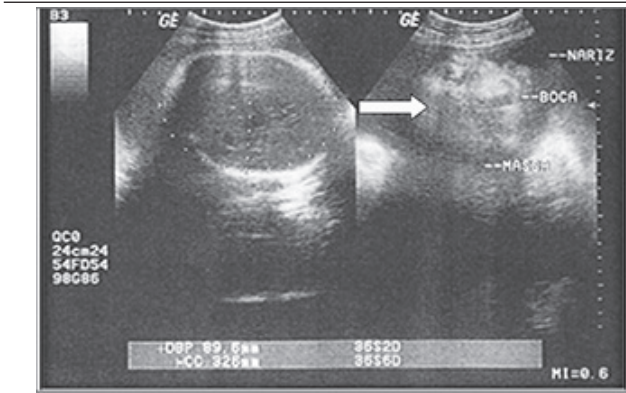


FIGURE 1 – Ultrasonographic aspect

Presence of an echogenic mass protruding from the oral cavity.



FIGURE 2 – Aspect at birth

Presence of a voluminous mass arising from the oral cavity.

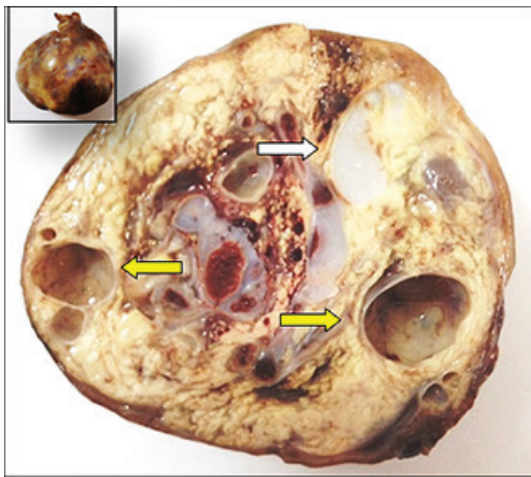


FIGURE 3 – External surface and cut surface of the tumor

Bosselated external surface (in the insert) and cut surface of the mass with solid cartilaginous (white arrow) and cystic (yellow arrows) areas.

tissue, smooth muscle, cartilage and bone (**Figure 4**). The picture is consistent with teratoma, predominantly mature, with (microscopic) focus of immature neuroepithelium (occupying up to one power field, 4× magnification) (**Figure 5**). Elements of embryonal carcinoma or endodermal sinus tumor were not observed.

Tumor relapse occurred on day 30, detected by cranial magnetic resonance, which showed an expanding extracranial formation of  $5.6 \times 1.8$  cm, at the right infratemporal fossa, extending to the midline, obstructing the nasopharyngeal air column almost completely.

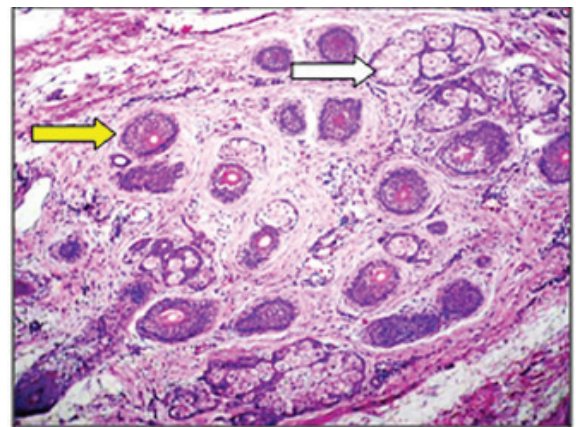


FIGURE 4 – Photomicrograph (HE staining, 10× objective lens)

Presence of skin annexes: hair follicles (yellow arrow) and sebaceous glands (white arrow).

HE: hematoxylin and eosin.

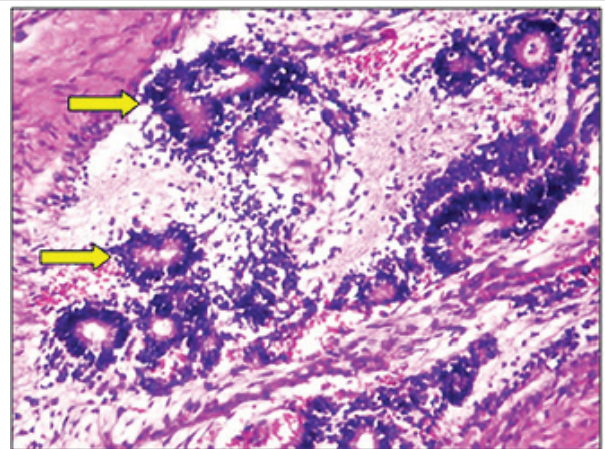


FIGURE 5 – Photomicrograph (HE staining, 40× objective lens)

Presence of mature epithelium (yellow arrows).

HE: hematoxylin and eosin.

A new operation was necessary to completely remove the lesion. The new histopathologic diagnosis was compatible with mature teratoma.

The newborn recovered well, and is currently healthy, with preserved quality of life.

## DISCUSSION

Teratomas occur in approximately 1:4,000 births, with female predilection, more frequently in the sacrococcygeal region. Oral teratomas are extremely rare, and almost only seen in neonates. They occur in approximately 1:200,000 liveborns, encompassing 2% of all teratomas. Most are benign, although malignancies have already been described<sup>(6-8)</sup>. Nasopharyngeal teratomas present a female predominance of 6:1; however, those of the oral cavity demonstrate no clear sex predilection.

Teratomas are generally 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, but poorly differentiated; 3) true teratoma, also containing the three embryonic germ layers, but differentiated in recognizable structures (cartilage, teeth, etc.); and 4) epignathus, highly differentiated in organs or limbs, rare, with high mortality rate<sup>(2)</sup>. The present case was classified as epignathus.

Yoshimura *et al.*<sup>(9)</sup> distribute teratomas in just three types: type 1 (skin and fatty tissue derived from two germ layers); type 2 (teratoma with tissues representing the three germ layers, with bone, teeth, neural tissue and tissues of the gastrointestinal tract); and type 3 (parasitic twins with well-differentiated organs and limbs). The current report is about type 2.

We describe a case of epignathus, a rare congenital nasopharyngeal tumor, classified as mature teratoma, in which the neonate is born with a mass protruding from the mouth.

Its etiology remains uncertain. Several theories have been formulated to explain its origin, including some involving mythology and mysticism<sup>(10)</sup>. Today there are still some theories, among them: traumatic implantation of skin or mucosa together with deeper tissues; congenital inclusion of germ layer cells that failed to complete their development in the embryonic phase; incomplete formation of conjoined twins at the third week of pregnancy, what would affect palate fusion, which happens between the seventh and the ninth week of pregnancy<sup>(1)</sup>; uncontrolled proliferation of pluripotent cells in Rathke's pouch<sup>(5,11,12)</sup>. The most

accepted theory is that epignathus is a parasitic fetus attached to the palate. When pedunculated, it is attached to the nasopharynx, in the basisphenoid region, or to the dorsal surface of the palate. The term *fetus in fetu* is reserved for highly differentiated masses, with the presence of limbs and organs. It is difficult to distinguish between epignathi and structures resulting from abortive attempts at twinning, due to their highly differentiated organic structure<sup>(13)</sup>.

As in the cases reported by Too *et al.*<sup>(8)</sup>, Chauhan, Gurusprasad and Inderchand<sup>(14)</sup>, and Clement *et al.*<sup>(15)</sup>, the studied case presented polyhydramnios at the ultrasound assessment performed during pregnancy. This manifestation has been attributed to the impaired fetal deglutition as a consequence of a large intraoral mass obstructing the oropharynx<sup>(12, 14, 15)</sup>. Prenatal diagnostic ultrasound allows careful assessment and delivery planning, improving prognosis in selected cases<sup>(11, 15)</sup>. Kumar *et al.*<sup>(11)</sup> state that large teratomas of the head and neck, in the absence of adequate resuscitation and a meticulous planning of airway management, are associated with higher mortality rates.

The giant epignathus may lead to death during the neonate period due to airway obstruction, as in fact happened in the case reported by Kumar *et al.*<sup>(11)</sup>. The newborn must undergo surgery in the first days of life; complete surgical excision is the definitive treatment. Incomplete removal may result in recurrence<sup>(6, 16)</sup>. Kontopoulos, Gualtieri and Quintero<sup>(17)</sup> revealed a successful intrauterine correction of teratoma, what seems to be a promising therapeutic strategy, exposing the newborn to fewer risks, although further studies on these procedures are necessary. Maeda *et al.*<sup>(18)</sup> described a case of epignathus with coexisting malformations that did not evolve to the need for ventilatory support, unlike the present case. The authors, though, mention another complication, which was the necessity of feeding through a nasogastric tube until complete resection of the tumor<sup>(18)</sup>.

Neonatal teratomas are mostly benign (composed of mature tissue). Just 5% of the cases present histopathological criteria to be considered malignant (foci of immature tissue). Incomplete resection and the presence of primitive neural tissue imply risk of malignancy. According to Benson, Fabbri and Russell<sup>(16)</sup>, recurrence of a congenital epignathus after resection does not necessarily indicate its malignancy, however, the patient must be kept under continuous clinical follow-up<sup>(3)</sup>.

Al-Madhi *et al.*<sup>(2)</sup> reported a case of epignathus with foci of immature adipose tissue at microscopy, but these foci normally undergo spontaneous differentiation, and the expected biological behavior of the tumor is benign. In the literature there are reports of giant epignathi<sup>(2, 4)</sup>. Rayudu *et al.*<sup>(4)</sup>, Too *et al.*<sup>(8)</sup> and Kumar *et al.*<sup>(11)</sup> described malignant epignathi. Histologically, the tumors



were composed of a mixture of embryonic cells and mature tissues, derived from all the three germ layers, nevertheless, the main component was primitive neuroepithelium<sup>(4, 8, 11)</sup>. The presence of calcifications at the microscopic exam is around four fold more common in benign teratomas than in malignant ones, thus, it may be an important diagnostic indicator. Inversely, the presence of primitive neural tissue suggests its malignancy<sup>(16)</sup>. According to Too *et al.*<sup>(8)</sup>, the presence of immature neuroepithelium is the only histopathological finding in teratomas that may indicate they are not benign. Therefore, even if most teratomas have benign biological features and are diagnosed as benign, the presence of immature neural tissue is an important factor; once malignant teratomas have already been described in the literature<sup>(2, 4, 8, 19)</sup>.

Histologically, teratomas may present diverse characteristics. In oral teratomas, the most frequently observed tissues are nerves and cartilage. Other commonly seen tissues are smooth muscle, bone and the respiratory epithelium<sup>(3)</sup>. In the present case, the histological sections exhibited mature tissues derived from the three germ layers, composed of keratinizing squamous epithelium with skin annexes, brain tissue, gastrointestinal epithelium, pulmonary tissue and mesenchymal elements, including adipose tissue, smooth muscle, cartilage and bone. Similarly the case published by Ram *et al.*<sup>(7)</sup> also presented smooth muscle, cartilage and brain tissue.

Although a microscopic focus of immature neural tissue was observed, epignathi, in their majority, are mature. This microscopic focus in a predominantly mature teratoma generally

undergoes spontaneous differentiation, and its expected biological behavior is benign. Notwithstanding, there are reports of immature voluminous epignathi<sup>(2, 9)</sup>.

Patients with oral teratomas often present coexisting anomalies, cleft palate being the most common. This happens due to the tumor obstructive mechanism, which prevents adequate closure of the palate<sup>(12)</sup>. Other common malformations are bifid tongues and noses<sup>(5, 19)</sup>. Contrarily to Teixeira *et al.*<sup>(5)</sup> and Jiang *et al.*<sup>(19)</sup>, who reported cleft palate-associated cases, in the present case there was no coexisting palate or oropharynx malformation.

Clinical differential diagnoses of epignathus teratoma in the neonate include: congenital embryonal rhabdomyosarcoma of the tongue, nasal glioma, retinoblastoma, heterotopic thyroid, cystic lymphangioma, neuroectodermal tumors and others<sup>(2, 7)</sup>.

## CONCLUSION

Epignathi are rare tumors originated in the hard palate, oral mucosa, tongue, and chin, protruding from the oral cavity. In the past, they were surrounded by myths and theories without the enlightening precepts of science<sup>(10)</sup> and were considered incompatible with life. Complete surgical resection is considered the definitive treatment, avoiding recurrence, ensuring an open airway and improving oral functions.

## RESUMO

Epignathus é um tipo de malformações raras, com elevado índice de mortalidade devido ao comprometimento respiratório por mecanismos obstrutivos. Na maioria dos casos, é benigno; entretanto, casos de epignathus maligno já foram descritos. A presença de neuroepitélio é um fator que aumenta a possibilidade de malignidade e pode confundir o diagnóstico. Descreve-se um epignathus em neonato, além de seu estudo clínico e histopatológico. O tratamento definitivo é a ressecção cirúrgica completa; focos residuais associam-se à recidiva, sem necessariamente aumentar o risco de malignidade.

**Unitermos:** teratoma; anormalidades teratoides graves.

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#### MAILING ADDRESS

Marcelo P. Menezes Filho

Av. XV de novembro, 309, apto 1.002; Centro; CEP: 28035-100; Campos dos Goytacazes-RJ, Brazil; e-mail: marcelopmfilho@hotmail.com.