

Epignathus are rare congenital teratomas of the oral, nasal and pharyngeal cavities

Abstract

We will describe the clinical case of a 3-month-old infant who showed non-severe obstructive airway symptomatology. She underwent a CT and was diagnosed as Epignathus, congenital teratoma of the mouth. Transoral surgical treatment was performed with excellent results. We emphasize the low frequency of this pathology and conducted a literature review on it, highlighting the importance of prenatal sonographic diagnosis in severe cases to make the best treatment plan for each patient.

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Introduction

The congenital teratoma of the mouth, also known as epignathus, is a rare disease. Like other teratomas, it presents tissues from the three germ layers and may contain differentiated organs. Its size, location and the amount of each of the three germ layers affect how it manifests.¹

Teratomas occur in 1 / 4,000 live births, of which the epignathus amount to 2-4% (1/100,000 - 1/200,000). These are tumors capable of threatening the life of the fetus and the newborn, they are especially dangerous at birth if they are located in narrow areas of the airway or if they cause complete blockage of the airway potentially leading to the death of the newborn when leaving the fetoplacental circulation.²

Clinical case

Infant 3-month-old female, without perinatal pathology. Birth at 39 weeks of gestational age. Birth weight 3200gr. Lower weight growth than expected in the last month.

The patient was referred to our Pediatric ENT Clinic because of repeated breathing pauses while sleeping and difficulty in feeding, with no other symptomatology. During physical examination we found a reactive infant with silent breathing, no hoarseness and no stridor. Examination of the oral cavity was normal; while examining the oropharynx a tumor was detected behind the uvula of which the inferior pole was observed Figure 1.



Figure 1 View of the oropharynx – The tumor can be seen behind the uvula.

A computed tomography revealed a tumor of 2.5 cm by 0.5 cm pedicled to the left lateral wall of the cavum that fell towards the supraglottis. (Figure 2 & 3). No other alterations were found.

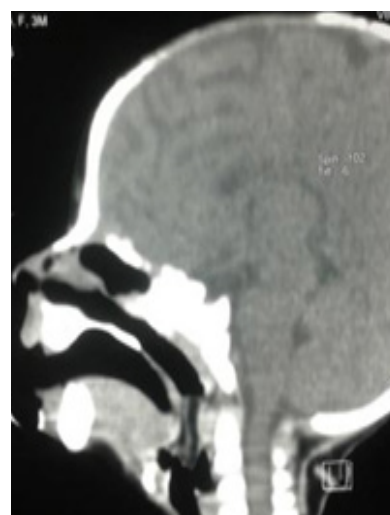


Figure 2 CT sagittal view.



Figure 3 CT coronal view.

Transoral resection of the tumor was performed. An orotracheal intubation was achieved without difficulties. The veil of the palate was retracted to achieve a good exposure of the tumor and it was completely resected. (Figure 4 & 5) The patient had an excellent postoperative evolution.



Figure 4 Transoral resection.



Figure 5 Resected tumor.

Discussion

Epignathus are congenital teratomas of the oral, nasal and pharyngeal cavities (craniopharyngeal canal) that can threaten the life of the neonate by obstruction of the respiratory tract and that cause alterations in skull, face and neck, mainly due to its mass effect (mordex apertus, cleft palate, bifid tongue, Pierre Robin sequence).^{3,4} The most accepted theory about its origin is that they are formed from the pluripotential cells of Rathke's pouch.⁵

Small tumors not located in narrow areas, which do not affect swallowing or breathing, tend to go unnoticed as in our patient, and are successfully operated on a deferred basis. Large tumors can be diagnosed prenatally by ultrasound, or may be suspected due to polyhydramnios caused. MRI is useful to obtain more information

about the lesion and to evaluate the possible intracranial invasion. Elevation of the alpha fetus protein is another indicator of this pathology.^{5,6}

Differential diagnosis must be made with neighboring tumor sites: Congenital tongue rhabdomyosarcoma, Retinoblastoma, Nasal glioma, Heterotopic thyroid, Cystic lymphangioma, Nasoethmoidal meningoencephalocele, Sphenoidal meningoencephalocele, Epulis.

Maintaining a secure airway is critical for patient survival. After the prenatal diagnosis the peripartum treatment must be planned, deciding whether to section the umbilical cord or not. In 1992 Catalano achieved the first survival of a patient using procedures initiated while maintaining fetoplacental circulation.⁷ Interventions under placental support are referred to as the EXIT technique and are based on a progressive increase in the aggressiveness of the maneuvers to maintain the airway, with tracheotomy being the safest option in some cases.

The anomalies most frequently associated with epignathus are correctable once the tumor has been removed. The prognosis of the giant epignathus seems to improve with a correct prenatal diagnosis and a planning of the airway control method. The EXIT protocol converts an emergency into a regulated and controlled intervention, and may be useful in the treatment of this pathology.

Acknowledgement

None.

Conflict of interest

None.

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