

Oral Teratoma (Epignathus) in a Newborn: A Case Report

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Abstract Congenital nasopharyngeal teratoma or epignathus is a very rare congenital neoplasms, it may be mature, immature or malignant. We report the case of a 04-day old female neonate, with a teratoma of the nasopharyngeal area. Examination showed a polypoid mass protruding from the nasopharynx measuring 3 x 2 x 2 cm, covered on one side with numerous fine and coarse hair. Surgical excision of the mass was performed under general anesthesia on day 4 after birth. Pathology revealed a well differentiated mature teratoma. The patient had no complication in the postoperative period.

Keywords: Oral teratoma, a newborn, respiratory distress

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1. Introduction

Congenital nasopharyngeal teratoma or epignathus is a very rare congenital neoplasms that distort facial anatomy and often cause acute respiratory embarrassment at birth. histologically, it contains elements derived from all three embryonic germ layers (ectoderm, mesoderm and endoderm), it may be mature, immature or malignant [1,2,3,6]. Teratomas may arise from different sites of the body; the most common site in the newborn is the sacrococcygeal region. Epignathus is a extremely rare lesions and comprise less than 2% of all teratomas. The treatment of choice is resection which depends on the site and size of the tumor [1,2,3].

2. Case Report

Our patient was a 04-day old female neonate, the product of a full term pregnancy from a 28 year-old mother G1P1, via a normal spontaneous vaginal delivery with the birth weight of 3000 grams, the length was 48 cm and the head circumference was 35 cm. Apgar score of the newborn was normal. Feeding was not possible associated to a mild respiratory distress.

Examination showed a polypoid mass protruding from the nasopharynx measuring 3 x 2 x 2 cm attached to the right nostril with smooth surface covered on one side with numerous fine and coarse hair. There was no associated anomaly.

Head and neck CT-scan demonstrated a soft tissue tumor in nasopharynx consisted of cystic, fat, cartilage and bony elements; brain CT scan was noted normal.

Surgical excision of the mass was performed under general anesthesia on day 4 after birth.

The cut section showed bone, soft tissue. Histopathological examination of multiples sections from both specimens of the mass revealed features of a mature teratoma. Tissues from all three germ layers are seen, ectodermal tissue in the form of stratified keratinising squamous epithelium with dermal appendages, mesodermal layer in the form of adipose tissue and cartilage; endodermal layer was demonstrated in the respiratory epithelium, which is pseudostratified and ciliated.

The patient had no complication in the postoperative period. At present, she is well and asymptomatic at 2 month follow-up.

3. Discussion

Teratomas are neoplasms composed of tissue elements foreign to the anatomic site of origin [1,2,3]. The term teratoma is derived from the Greek word "teraton", which means 'monster,' and initially was used by Virchow in his first edition of his books on tumors, published in 1863 [2,8]. They are true neoplasms originating from pluripotent cells and are composed of tissues from all three germinal layers usually benign in nature. : ectoderm, mesoderm, and endoderm [2,3,6].

They are more common in female [7,9,10]. Carney et al. found malignant teratoma to be more common in men in a ratio of 5:4 [2,3]. When present during early childhood, they are usually benign. Small teratomas may not be diagnosed at birth, although routine prenatal ultrasonography may diagnose obstructive tumors before birth by as early as 21 weeks gestation [2,8].

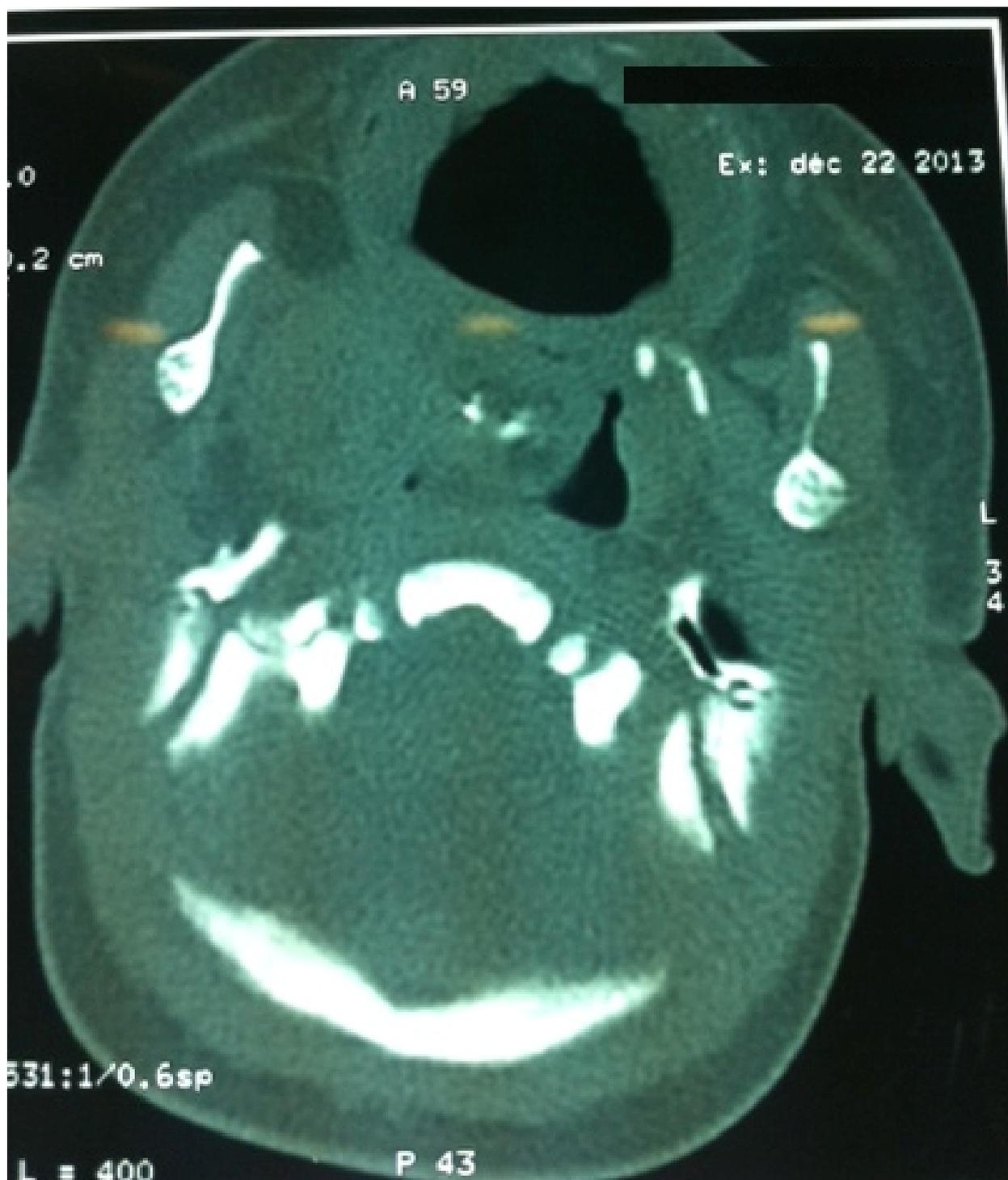


Figure 1. axial view of ct showed a mass in the nasopharynx

They occur with an incidence of 1:4000 live births [1,2,3]. Sacrococcygeal teratomas are the most common type (45-65%) followed by teratomas of the gonads (10-35%), anterior mediastinum (10- 12%), retroperitoneum (3-5%), cervical (3-6%), presacral (3-5%), central nervous system (2-4%) and other sites (<1%) [3,4]. Nasopharyngeal teratomas are extremely rare lesions and comprise less than 2% of all teratomas [1,3,4].

Epignathus is commonly used to describe a congenital teratoma in the nasopharyngeal region; it is rare in the newborn; only 13 cases have been reported in the literature. And it is a potentially lethal fetal anomaly and 80 to 100% of these cases succumb to acute respiratory

distress secondary to airway obstruction at the time of birth [2,3,8].

The etiology of epignathus is unknown and may arise from pleuripotential cells from the soft or hard palate in the region of Rathke's pouch that grow in a disorganized manner, or in the nasopharynx in the region of the basisphenoid, tongue, sinuses, mandible or tonsil. [1,3,8]. Teratomas are associated with concomitant malformations, with cleft palate being the most commonly associated anomaly. This is thought to be because of mechanical obstruction caused by the neoplasm, preventing closure of the palatal shelves. Other malformations associated with epignathus teratomas are bifid tongues and noses [2,8].

Preoperatively, the main Clinical differential diagnosis of neonatal oral mass included but was not limited: embryonic congenital rhabdomyosarcoma, retinoblastoma, nasal glioma, a congenital granular cell tumor of the gingival and sphenoid meningoencephalocele. CT scan and MRI play a key role in differentiating neonatal nasopharyngeal teratomas from other causes of neonatal oral mass to a teratoma or a congenital granular cell tumor of the gingival; rhabdomyosarcoma is an appropriate

consideration, with the oral and maxillofacial area being the preferred site and 2% occurring congenitally [2,5,7].

The main therapy of teratoma is complete surgical excision which depends on the site and the size of the tumor. The Exclusion of intracranial extension is an important part of preoperative management [3,8]. Prognosis is excellent; recurrences are rare, it occurs due to incomplete surgical resection [2,3,7].

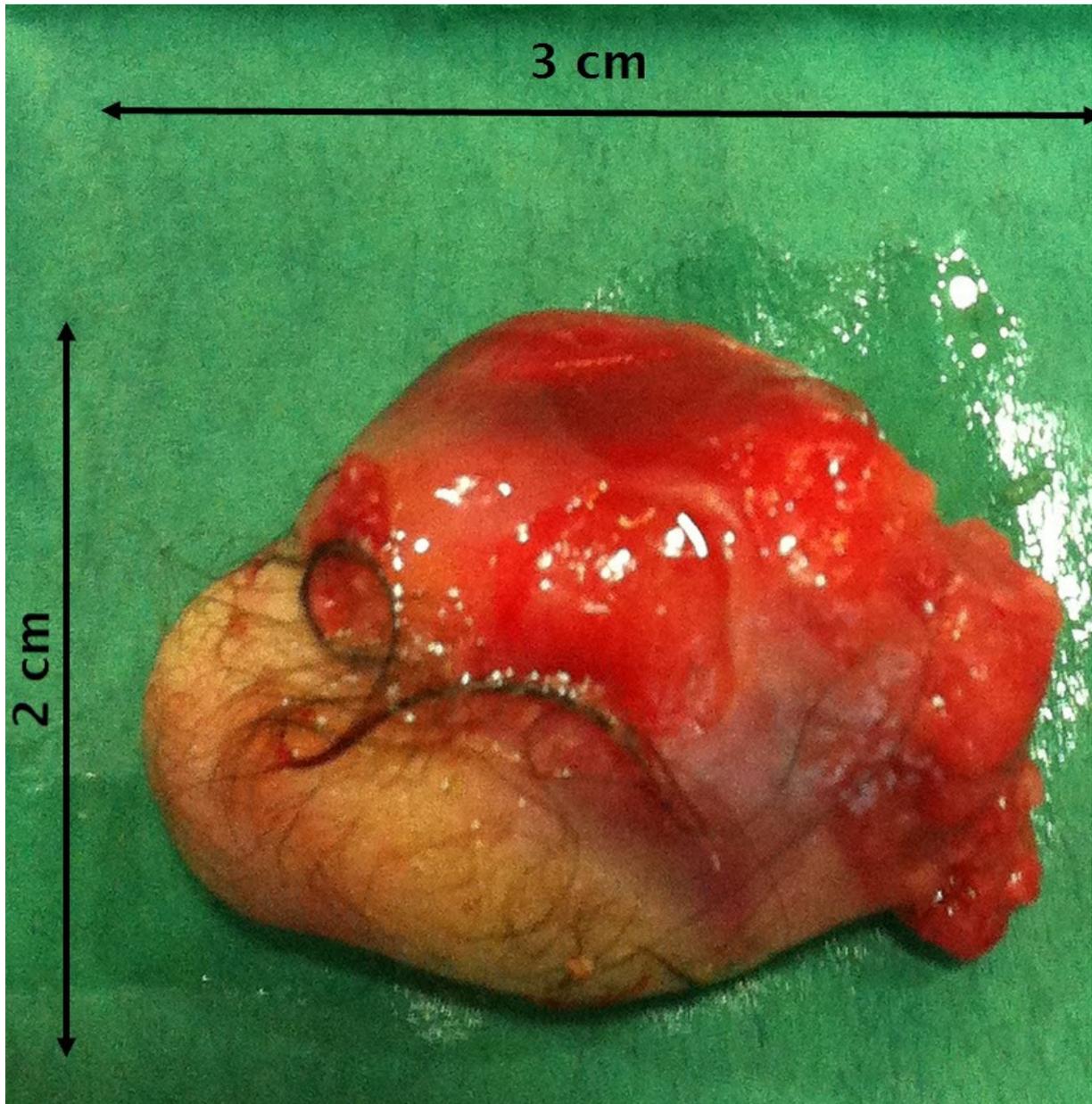


Figure 2. the resected epignathus covered on one side with numerous fine and coarse hair

4. Conclusion

Teratomas of head and neck are exceedingly rare and only about 10% of teratomas are found in this area. Nasopharynx and cervical region are the most common sites [3,7].

These lesions can be detected early by prenatal ultrasound and a strategic plan developed early in the prenatal period. The airway management should be tailored to each individual patient which is crucial to the prevention of neonate mortality and morbidity [1,3,5].

Conflict of Interest

None.

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