

Oropharyngeal Teratoma – A Congenital Malformation with Atypical Computed Tomography Findings: Unique and Dangerous

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Abstract

Newborn oropharyngeal teratomas are extremely rare disorders. Epignathus tumour is a congenital malformation characterised as a 'mature teratoma' located at the oropharyngeal region. Teratomas in the upper respiratory airway give rise to abnormal life-threatening airway obstruction during the neonatal period. It is associated with high morbidity and mortality, requiring proper and cautious planning and prompt surgery. We describe a rare case of oropharyngeal teratoma in a premature male neonate (35 weeks gestation) who presented with difficult intubation. On flexible laryngoscopy, it appeared as a large vallecular cyst, while the computed tomography scan demonstrated a posterior pharyngeal wall cystic lesion causing oropharynx narrowing. The patient was operated on 8th day postpartum. Histopathological examination was compatible with mature cystic teratoma.

Keywords: Oropharyngeal; Teratoma; Cyst; Epignathus; Computed Tomography scan

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INTRODUCTION

Teratoma is a type of three germ cell layers (endoderm, ectoderm and mesoderm) benign tumour comprised of different types of tissue such as bone, hair, muscle and teeth.¹⁻³ It is commonly found in the sacrococcyx (most

common - 45% cases)⁴, anterior mediastinum, ovaries, testes and retroperitoneum, but teratomas can occur anywhere in the body.^{1,5,6}

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Tumors originating from the oral cavity and nasopharynx in newborns are extremely rare.¹ Congenital teratomas arising from the palate or pharynx are commonly known as epignathus.^{1,7} It has an estimated incidence of 1:35,000 to 1:200,000 live births.^{1,5} This condition can be life-threatening during the neonatal period as it causes severe airway obstruction.^{4,7} Diagnosis of oropharyngeal teratoma depends significantly on both radiographic imaging and histopathology results. This case report aims to highlight the atypical findings of oropharyngeal teratoma on computed tomography (CT) scan.

CASE REPORT

A male baby was delivered prematurely at 35 weeks of gestation via an emergency lower segment caesarean section due to foetal distress. Antenatally, the mother had gestational diabetes mellitus, vaginal candidiasis complicated with polyhydramnios and a large for gestational age (LGA) foetus. Physical examination showed a syndromic baby with evidence of widened anterior fontanelle, long philtrum, low-set ears and rocker bottom feet. The baby was not vigorous at birth. He developed severe respiratory distress and a brief period of seizure. We proceeded with endotracheal intubation to provide airway protection. However, it was difficult because of the presence of a cyst blocking the vocal cord. A total of six attempts were made before successful endotracheal intubation by a consultant neonatologist. Based on chest radiograph findings, the patient was provisionally treated for congenital pneumonia with persistent pulmonary hypertension of the newborn (PPHN) complicated with right pneumothorax. A right chest tube was inserted.

A flexible laryngoscopy was done by a Otorhinolaryngologist, which showed a large vallecular cyst originating from the left posterolateral pharyngeal wall and obstructing the airway (**Figure 1**). Urgent contrast CT of the neck exhibited an ill-defined hypodense non-enhancing predominantly cystic lesion at the left posterior pharyngeal wall extending to the prevertebral spaces with narrowing of the oropharynx (**Figures 2a and 2b**). Blood culture and sensitivity showed the presence of *Burkholderia cepacia* (Complex), which was sensitive to ceftazidime. Other blood investigation findings were unremarkable.

The following day, the patient underwent an excision of the posterior oropharyngeal wall mass. Intraoperatively, a solid cystic mass noted at the posterior oropharyngeal wall that extends superiorly to the soft palate, posteriorly to the prevertebral muscle and

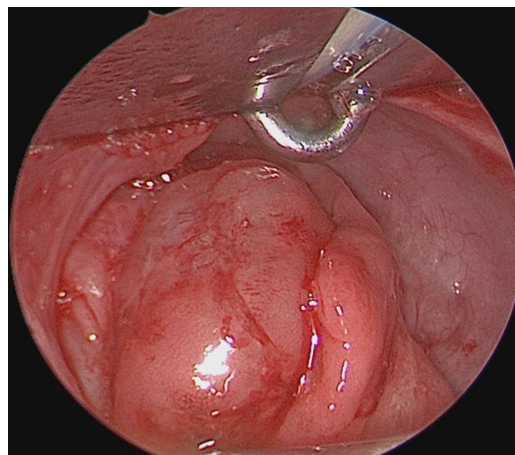


Figure 1: Flexible laryngoscopy showed a large vallecular cyst originating from the left posterolateral pharyngeal wall causing airway obstruction.

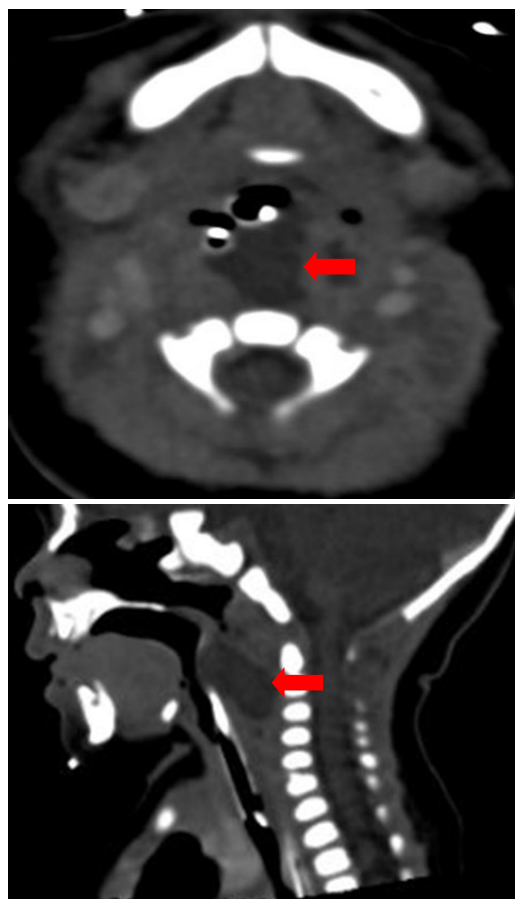


Figure 2: a) Axial contrast CT neck demonstrating ill-defined hypodense non-enhancing predominantly cystic lesion at left posterior pharyngeal wall extending to the prevertebral spaces with narrowing of the oropharynx (red arrow).

inferiorly above the post-cricoid region with clear mucoid content. Histology revealed that the sample was compatible with mature cystic teratoma with tissue

differentiation of ectodermal, mesodermal and endodermal origin seen (**Figures 3a and 3b**).

The patient was placed on long-term antibiotics and in the neonatal isolation ward for more than four months before discharge. He remains active and tolerates nasogastric feeding well. Repeated flexible laryngoscopy on day 43 post operation shows no recurrent oropharyngeal mass, mobile bilateral vocal cords with mild edematous of arytenoid mucosa. Another flexible laryngoscope done prior to discharge showed no recurrent oropharyngeal mass.

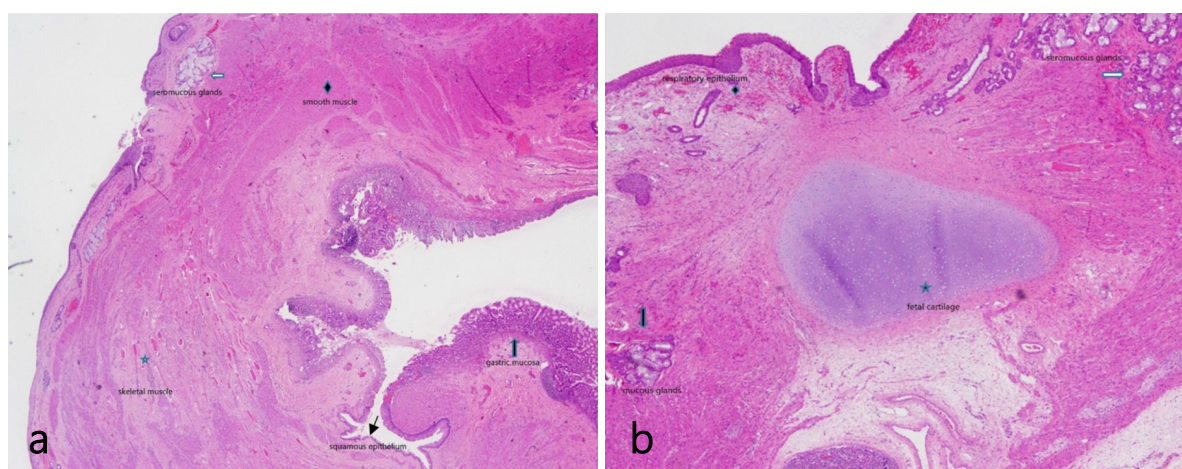
DISCUSSION

This case reports a premature male newborn with severe airway obstruction after birth, secondary to a posterior pharyngeal wall cystic lesion that was detected during difficult intubation and histopathologically confirmed as a teratoma. Teratomas ("monstrous tumours") are classified as common congenital tumours with an incidence rate of 1:4000 births,⁷⁻⁹ and approximately 2% originate from the oropharyngeal region.^{1,7,8} Oropharyngeal teratoma tends to display female predominance,^{2,9,10} contrary to this case. Teratoma can be mature, immature or mixed.¹¹ It is divided into dermoid, teratoid, true teratoma and Epignathus.^{2,4,12} The dermoid (hair polyps) typically consists of epidermal and mesodermal elements. It is the most common type. Teratoid comprises ectoderm, mesoderm, and ectodermal elements; however, they are poorly differentiated. True teratoma contains all three germ cell layers similar to the teratoid, but they are differentiated to the level of specific tissues (as in our patient). Meanwhile, epig-

nathus, resembling limb-like structures emerging from the mouth, is highly differentiated into recognisable organs or limbs.^{8,11} Even with the classification, the term epignathus tumour is widely applied to neonatal teratomas arising from the oropharyngeal cavity, especially those protruding from the mouth without specifying the site of origin.^{4,12}

Oropharyngeal teratoma gives rise to life-threatening neonatal asphyxia due to airway obstruction soon after birth.^{3,4,7} The aetiology remains unknown, but the most popular theory suggests it may emerge from the disorganised growth of pluripotential cells within Rathke's pouch region.^{7,10-12} It is notably associated with maternal polyhydramnios (40% cases) as in this case, where large pharyngeal or cervical mass leads to foetal swallowing difficulties, preventing amniotic fluid resorption.^{4,8} Most reported cases demonstrated elevated serum alpha-fetoprotein (AFP) level.^{7,8} Our patient's AFP assay taken postoperatively was normal for age (15,499.3 ng/ml).⁸ Regrettably, no AFP assay was taken before the surgery.

Radiological imaging is essential in identifying the location and the content of oropharyngeal teratomas. Typical teratoma CT scan findings will show a lobulated heterogeneous mass or soft tissue density mass originating from the oropharynx,^{6,12,13} associated with solid cystic components.¹⁴ Some cases exhibit areas of the fatty component, bones and intestinal tract.^{6,11} Calcifications are common CT findings, with a prevalence of almost 16% in postnatal teratoma cases.¹⁴ Presentation of a predominantly cystic lesion, as seen in our patient, is rare. Our patient's CT findings were atypical,



Figures: Sections show a well-circumscribed solid-cystic tumour comprising all three germ layers. The ectoderm consists of stratified squamous epithelium and skin adnexal structure (eccrine glands). Immature cartilage was seen. Mesoderm consists of smooth muscles, skeletal muscles, blood vessels and nerve bundles. Endoderm consists of salivary glands and foveolar type epithelium of gastric mucosa. No immature component or malignancy was seen. Figures 3a: Smooth muscle (diamond), seromucous gland (white arrow), gastric mucosa (black arrow), skeletal muscle (star). Figure 3b: mucous gland (black arrow), seromucous gland (white arrow), fetal cartilage (star).

exhibiting an ill-defined hypodense non-enhancing cystic lesion at the left posterior pharyngeal wall without any significant heterogenous soft tissue dense mass, solid component or fatty component. There was no evidence of intralesional calcification or bony density, unlike those reported in other cases.^{6,14} Due to these uncommon CT findings, histology examination played a significant role in confirming this patient's diagnosis of oropharyngeal mature cystic teratoma.

Common differentials for cystic oropharyngeal lesions are divided by location into midline and lateral. Midline lesions include ranula, thyroglossal cyst and dermoid cyst, whereas lateral lesions can be brachial cyst and lymphangioma.^{9,15} Ranula arises from the floor of the mouth as a sublingual or minor salivary gland retention cyst. It exhibits a typically thicker wall on CT.¹⁵ Thyroglossal cysts will demonstrate a smooth thin wall cystic lesion that can either originate from the anterior midline or paramedian location. Regardless, it should be within 2 cm of the midline.¹⁵ The dermoid cyst can be distinguished from teratoma histologically, where it only contains dermal and epidermal elements. Meanwhile, a brachial cyst is a cystic lesion arising laterally using the mandible, parotid gland or infrahyoid neck as landmarks for classification. It is a well-circumscribed, thin-walled fluid density mass that exhibits variable wall enhancement. This enhancement tends to increase with recurrent infections.¹⁵ Lymphangioma will appear homogeneous and cystic on CT. However, some may display inhomogeneity because of the presence of proteinaceous, fluid, blood, or fat components within the lesion.¹⁵

As our patient displayed several physical features such as widened anterior fontanelle, long philtrum, low-set ears and rocker bottom feet, the association of oropharyngeal teratoma to a specific syndromic disorder should be considered as a possibility and worth exploring. However, genetic testing was not proceeded for this patient due to financial constraint. There has yet to have any report that provides confirmed evidence of association between oropharyngeal teratoma with any specific syndromic disorders. Other articles described different physical features that were reported in teratomas of head and neck, which are divided into craniofacial and non-craniofacial anomalies. Craniofacial anomalies such as cleft palate (commonest feature), microcephaly, common carotid artery atresia and tonsillar malformations.^{1,8,13} Whereas non-craniofacial anomalies include imperforate anus, chondrodysplasia, left ventricular hypoplasia and pulmonary hypoplasia.⁸ Nonetheless, our patient did not exhibit any of these

commonly reported features mentioned above which made it unique.

The treatment for oropharyngeal teratoma is complete surgical excision, which in this case was successfully executed by the otorhinolaryngology team. This method of treatment is aligned with the pre-existing literature which concludes that a complete surgical excision brings greater and positive outcome to the patients.^{1,7,8,11}

Cases reported by Aubin *et al.*⁸ describes oropharyngeal masses which underwent surgical excision along with few aesthetic and functional sequelae, coupled with pre-operative and post-operative MRI, CT and ultrasound surveillance. Radiological imaging is vital to determine extension of the lesion, its anatomical relations with vital structures and determine the possibilities of resection. Only one out of the four patients required neoadjuvant chemotherapy prior to surgery due to its initial secreting nature and unresectability, which was then operable after the therapy. All patients showed good prognosis post-surgery with great recovery and no complication.

Aydemir *et al.* described a case of palatine tonsil teratoma which presented with severe airway obstruction, evaluated using both CT and MRI for extension of tumor, then proceeded with surgical excision of the teratoma.¹¹ Follow-up of the child shows no recurrence after surgery with good development and growth.

Our case agrees with previous articles that regular medical follow up is crucial to monitor and manage any postoperative complications.^{8,11-13} We suggest long-term medical follow-up to include clinical examinations, radiological imaging, CT or but MRI with serial AFP monitoring.^{7,8,11} Elevation of serum AFP may be associated with tumour recurrence, which warrants radiologic evaluation and surgery if confirmed.⁷ MRI and three-dimensional colour sonography are highly recommended in the 2nd trimester of pregnancy (14 weeks to 27 weeks) if there is any evidence of elevated maternal AFP.^{8,14} The combination of these imaging features will provide adequate in-utero imaging. When coupled with multidisciplinary cooperation, imaging will positively impact the survival and prognosis of newborns with oropharyngeal tumours.

CONCLUSION

Oropharyngeal teratoma is an extremely rare congenital malformation which relies on radiological imaging, such as a CT scan, for early detection and management.

However, teratoma may exhibit atypical CT findings, as described in this case. Therefore, histology is crucial to confirm the diagnosis when presented with atypical radiological or clinical features.

Take Home Message

- Oropharyngeal teratoma is extremely rare with estimated incidence of 1:35,000 to 1:200,000 live births
- It is associated with life threatening airway obstruction.
- CT findings may be atypical and non diagnostic.
- Histology confirmation is important.

Abbreviations

CT	Computed tomography
LGA	Large for gestational age
PPHN	persistent pulmonary hypertension of the newborn
AFP	Alpha-fetoprotein

Declarations

Patient Consent

Patient consent has been obtained. This case report submission is approved by the Jerudong Park Medical Centre (JPMC) and written consent was obtained from this patient following the JPMC Policy

Disclosure and Conflict of Interest

The authors declare that they have no conflicts of interest and no financial disclosures relevant to this case report.

Acknowledgments

None.

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