Rare disease

Oropharyngeal teratoma, a rare cause of airway obstruction in neonates

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Summary

Oropharyngeal teratomas are extremely rare congenital tumours, and because of their location in the upper airway they represent one of the most unusual causes of airway obstruction during the neonatal period. We report a 2-month-old girl who presented with repeated vomiting, failure to thrive and recurrent bouts of stridor and cyanosis since birth. On examination there was a mass originating from the oropharynx. The mass was excised under general anaesthesia by CO₂ laser. Histopathological examination revealed a teratoma of the oropharynx. We discuss the clinical presentation, diagnosis and treatment of this case as well as a review of the literature.

BACKGROUND

Respiratory failure in newborn babies may be because of a wide variety of causes. Rare causes are obstruction of the upper airways by congenital tumours such as lymphangiomas, gliomas, encephaloceles, haemangioma and teratomas.¹ Teratomas are benign tumours consisting of several parenchymal cell types originating from the germ layers ecto-, meso- and endoderm. Teratomas in the head and neck region are rare (2–9% of all reported teratomas) and are even rarer in the oropharynx.² Although teratomas are of benign histopathological nature, they may be life threatening because of their location in the upper airway representing one of the most unusual causes of respiratory distress during the neonatal period.²

CASE PRESENTATION

A 2-month-old girl born to non-consanguineous parents was admitted to our department with episodic stridor and cyanosis since birth. Also she had feeding difficulty, repeated vomiting and failure to thrive.

The baby was born to a healthy 27-year-old mother. She was born full term via normal spontaneous vaginal delivery in an institution, and cried immediately after birth. The pregnancy and delivery were uncomplicated. There were neither polyhydramnios nor masses on routine ultrasound examination during gestation. There was no similar type of illness in the family and she is the first-born baby. On detailed clinical examination, the baby was conscious and active. Her respiratory rate was 65 per minute and heart rate 140 per minute. Anaemia and jaundice were absent. Her facial features were normal and anthropometric measurements were as follows: body weight, 2.5 kg; head circumference, 35 cm, chest circumference, 32 cm, length, 52 cm. Cervical lymph nodes were not enlarged. Examination of oral cavity revealed a well-circumscribed, whitish, non-pulsatile rounded mass occupying the left side of the oropharynx protruding inside the child's mouth. The mass was prominent during

crying. The nose and nasal cavity were normal. No other congenital anomalies and no cleft palate were detected.

INVESTIGATIONS

MRI of the baby's head and neck revealed a well-defined soft tissue lesion, arising from the oropharynx, epicentral upon the free edge of the epiglottis and the uvula in the midline. It measured 2×1 cm in diameter with hyperintense signal in T1 and T2 with loss of signal intensity in fat suppression (figure 1). It did not show any relation with the intracranium or the large blood vessels in the neck.

DIFFERENTIAL DIAGNOSIS

It should be differentiated from other causes of airway obstruction and respiratory failure in the neonates. Rare causes include obstruction of the upper airways by congenital tumours as lymphangiomas, gliomas, encephaloceles, haemangiomas and teratomas.¹

Lymphangiomas are rare congenital malformations of the lymphatic system that occur throughout the body with greater frequency in the cervicofacial area. Cystic lymphangiomas have been more reported in the literature. They present with respiratory obstruction and problems with feeding in neonates.³

TREATMENT

After fiber-optically controlled intubation with a tube 3.0 passed over the flexible endoscope, the swelling could be inspected under general anaesthesia. The mass appeared to originate from the left posterior pillar measuring 2×1 cm in diameter. Excision was performed transorally using the CO₂ laser in order to minimise blood loss and to keep a good overview (figure 2). The postoperative period was uneventful.

The excised mass was sent for histopathological examination which revealed a polypoid mass covered by keratinised stratified squamous epithelium appended with sebaceous glands and hair follicles, separated by bundles

Figure 1 (A) MRI of the head and neck axial views T1 and T2 showing a soft tissue mass epicentral upon the free edge of the epiglottis and the uvula in the midline. (B) MRI of the head and neck sagittal view showing a soft tissue mass (arrow) epicentral upon the free edge of the epiglottis and the uvula in the midline.

of smooth and skeletal muscles fibres. Mucinous acini were detected (figure 3). No immature elements were detected. There were no signs of malignancy. The pathological diagnosis of mature solid teratoma of the oropharynx was made.

OUTCOME AND FOLLOW-UP

Stridor and cyanosis attacks were no longer observed and the child was able to tolerate oral feedings. Six months later, she is doing well without complaints concerning respiration or feeding and has started to gain weight.

DISCUSSION

The term teratoma, derived from the Greek word 'teraton' (meaning monster), was initially used by Virchow in 1863.^{1 4} Teratomas are malformations composed of an assemblage of tissues often foreign to the site in which they normally arise.² There is variability in the

classification of these tumours. The most widely accepted terminology describes four types of teratomas:^{1 4 5}

1. Dermoids (hairy polyps) are the most common form and are composed of ectoderm and mesoderm, like in our patient.
2. Teratoid tumours are rarely seen and are composed of all three germ layers, but are poorly differentiated.
3. True teratomas, all germ layers are differentiated to the level of specific tissues. There is one case of true teratoma in the oropharynx reported in the literature.⁶
4. Epignathi are limb-like structures protruding from the mouth and present the highest form of teratoma differentiation in which fetal organs have developed.

Teratomas occur in 1 in 4000 births and display a female predominance.^{2 5 6} They may originate anywhere along the midline and clinical behaviour varies significantly according to site and size. The sacrococcygeal region is the most common site, followed by the gonads,

Figure 2 The oropharyngeal mass appeared to originate from the left posterior pillar, 2×1 cm in diameter, was excised transorally using the CO₂ laser.

the retroperitoneal region and the anterior mediastinum. Teratomas of the head and neck region account for about 2–9% of all reported teratomas. Most of them are located in the cervical region. The second most common location in the head and neck region is the nasopharynx. Teratomas have also been reported in the parapharyngeal space, the orbit and the larynx. The oropharynx is an uncommon site of involvement.^{1 2 4–6}

The pathogenesis and origin of teratomas in the head and neck are controversial and multiple theories are suggested. It is proposed that teratomas in the head and neck region developed by pinching off of remnants of the three germinal areas, which become marginated in the normal migratory pathway as the different structures of the head and neck develop. The remnants developed into predetermined organ systems, giving rise to heterotopic tissues encountered.^{6 7}

There is no evidence to suggest environmental risk factors in the pathogenesis of pharyngeal teratoma. Furthermore, there are no karyotypic abnormalities involved, and these lesions are not thought to be inherited in a Mendelian or polygenic manner.⁷

Therefore, parents should be reassured that they are not at increased risk of having another child with this lesion.

Beside the mass, oropharyngeal teratomas usually present with obstruction of the upper respiratory tract. Oropharyngeal teratomas may be life threatening because of airway obstruction. Other symptoms include snoring, rhinorrhoea, recurrent cough, dysphagia, vomiting and failure to gain weight. It was reported that small teratoma can be diagnosed later on in life presenting with speech problems.^{1 6–8}

Some cases of congenital oropharyngeal teratomas with intracranial extension were reported. In the majority of

Figure 3 (A) Keratinised stratified squamous epithelium with hair follicles (arrows). (B) Pink smooth muscle fibres dissecting between lobules of mature fat (F).

these reports the tumours are associated with stillbirth, perinatal death or significant morbidity following attempted resection.⁹

Prenatal ultrasound may show the tumour and polyhydramnios due to obstruction to swallowing caused by the tumour.^{1 2 4-6} It was reported that oropharyngeal teratoma can be identified by an elevated level of maternal serum α -fetoprotein at 16 weeks with sonographic diagnosis at that time.

MRI is helpful in revealing the extent of oropharyngeal involvement and defining tracheal anatomy in preparation for tracheostomy. It is also helpful in defining vascularity and intracranial extension and making a diagnosis by the pattern of imaging density caused by the mixed tissue types.^{1 2 4 5}

Treatment of head and neck teratomas consists of surgical excision. If respiratory distress accompanies the lesion, establishment of an adequate airway has priority.^{4 5} Intubation may be difficult due to the mass. Optimal optic control during intubation can be obtained by using a flexible endoscope with the tube passed over the endoscope. If necessary, a tracheostomy has to be performed. Surgical excision should be undertaken as an urgent, but not emergent, procedure if the airway can be managed successfully. Before treating any congenital pharyngeal mass the surgeon must ensure that the mass has no relationship to vascular structures in the neck and does not have an intracranial extension or communication.^{4 7 8} If the diagnosis is made antepartum, the born child may be maintained on fetal circulation until successful intubation or tracheostomy.^{7 9} There are some reported cases that necessitated deliveries with ex utero intrapartum treatment procedure using a classical incision. The neonate's small size and short umbilical cord required complete exteriorisation to secure the airway.⁵

Teratoma is a histopathological diagnosis and contains histological evidence of two or three germ layers. The ectodermal derivatives of teratoma include keratinising squamous epithelium, skin, hair follicles, sebaceous glands and sweat glands. Mesodermal components can include bone, cartilage, smooth muscle, fat and blood vessels. Gastric and intestinal mucosa and respiratory epithelium are common manifestations of the endodermal remnants. Although the vast majority of teratomas are histologically benign, reports of malignant teratoma do exist. In addition, immature cellular components are associated with a greater likelihood of malignant degeneration.⁹

The prognosis is excellent after complete surgical excision, respiratory obstruction and oral feeding improve post-operatively and the baby starts to gain weight.⁴ Recurrence is rare and is due to incomplete surgical excision.^{4 6}

Learning points

- ▶ Teratomas are tumours consisting of several parenchymal cell types originating from the germ layers ecto-, meso- and endoderm. Teratomas in the head and neck region are rare (2–9% of all reported teratomas) and are even rarer in oropharynx.
- ▶ Although teratomas are of benign histopathological nature, they may be life threatening because of their location in the upper airway representing one of the most unusual causes of respiratory distress during the neonatal period. Other symptoms include snoring, rhinorrhoea, recurrent cough, dysphagia, vomiting and failure to gain weight.
- ▶ Antenatal diagnosis can be made by prenatal ultrasound and fetal MRI.
- ▶ Treatment of head and neck teratomas consists of surgical excision. If respiratory distress accompanies the lesion, establishment of an adequate airway has priority by intubation or tracheostomy.
- ▶ Definite diagnosis of teratoma is by histopathological examination of the excised mass.
- ▶ The prognosis is excellent, recurrence is rare and is due to incomplete surgical excision.

Competing interests None.

Patient consent Obtained.

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