原文題目(出處)：	A large teratoma of the hard palate: a case report
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《Abstract》

Congenital teratoma is a rare malformation, and few papers have been published about it. This journal presents a case of a large teratoma that arose from the hard palate in a neonate, and offers a brief discussion about it.

《Introduction》

- "Teras" in Greek = *Monster*

Origin

- Teratomas are true neoplasms, which are presumed to be derivates from primordial germ-cell (原始胚胎細胞), and typically consist of tissues from all three embryonic germ layers: ectoderm, mesoderm, and endoderm. Often they show uncoordinated growth.
- Teratoma was defined by *Weaver et al.* as a tumour consisting of multiple tissues that are not indigenous to their site of origin. Some authors may incorrectly call the lesions that do not have derivatives of layers of all three germ cells "teratoma"

Sites

- Teratomas may develop in almost any area of the body, but usually in median sites. A reported incidence of 1:4000 live births with around 2%–9% of these in the head and neck, and the other common sites are the sacrococcyx, anterior mediastinum, testicle, ovary, or retroperitoneum.
- "Epignathus" is commonly used to describe a congenital teratoma in the oropharyngeal region, but should be reserved for tumours that arise from the jaw, specifically the alveolus of the mandible. Oropharyngeal teratomas should be subdivided according to their site. However, regardless of their anatomical site, "epignathus" is the most used term for an oropharyngeal teratoma.

《Case Report》

- A white female infant was born prematurely after 35 weeks' gestation and the mother was seen prenatally at 19 weeks. Incidental ultrasonography showed that the fetus had a univentricular heart and a large mass protruding from its mouth that arose from the hard palate.
- The mother went into labour at home and gave birth by spontaneous vaginal delivery. The child was in respiratory distress and was initially supported by bag and mask ventilation. She was immediately transferred to hospital where a nasopharyngeal tube was inserted. From there she was transferred to the tertiary referral centre and a tracheostomy carried out when she was 4 hours old.
- Examination showed an obstructive polypoid mass that included areas covered in skin and hairs, and cystic, ulcerated lobes.
- Computed tomography showed a complex mass protruding from the oral cavity. It was viewed three-dimensionally and seen to arise from the palate with a thin element extending into the nasal septum. It consisted of cystic, fat, bony, and neural elements, with defined teeth within the bone. There was a tiny defect in the floor of the base of the skull, though there was no evidence of intracranial

involvement.

- A magnetic resonance scan confirmed that there was no intracranial extension.
- Genetic testing failed to show a syndrome, and amniotic fluid was of normal karyotype. On auscultation there was a 4/6 ejection systolic murmur.
- A univentricular heart and persistent ductus arteriosus had been diagnosed antenatally and was confirmed postnatally as a double-outlet left ventricle with transposition of the great arteries and tricuspid atresia. Evidence of cardiac failure was seen by day 9, but cardiac surgery was delayed until after treatment of the craniofacial anomaly because of risk of infection and poor weight gain.
- The tumour was excised at 4 weeks. Cystic areas were decompressed and the pedicle (roughly 2.5 cm in diameter) was sectioned to deliver the mass. Nasal integrity was confirmed. The temporomandibular joints moved well although the micrognathic mandible was depressed posteriorly.
- After the tumour had been removed, a cleft soft palate became apparent. Postoperatively the palatal wound epithelialised well. She had a gastrostomy at the age of 8 weeks.
- By 7–8 weeks the neonate required continuous positive airway pressure, and later, ventilation. At 12 weeks her pulmonary arterywas banded and the persistent ductus arteriosus ligated with pronounced improvements in her symptoms of heart failure.

《Histopathological examination》

- The mass was not encapsulated, and there was a mixture of well-defined tissues derived from the ectoderm, mesoderm, and endoderm. Skin, fat, bone, cartilage, squamous and respiratory epithelium, lymphoid tissue, salivary gland type tissue, and neuroglial tissue with choroid plexus and ependyma, were all present within the mass. There were elements of focal necrosis, but only one small focus of undifferentiated neuroblasts.
- It was diagnosed as teratoma epipalatus. We were uncertain about whether this should be regarded as an immature teratoma, or as a mature teratoma with immature elements “appropriate to age”, because the tissues overall were immature but consistent with the degree of maturity expected in a neonate.

《Discussion》

Benign? Or Malignant?

- Neonatal teratomas are usually benign, there being a higher incidence of malignancy in teratomas in adults.
- Calcification is four times more common in benign than malignant teratomas, so may be an important diagnostic indicator.
- The presence of primitive neural tissue also suggests malignancy.
- Clinician should also be aware to the possibility of other germ cell tumours; and the most worrying of which in neonates is the yolk sac tumour (named by Huntington and Bullock). Half of malignant teratomas contain this lesion, which is an embryonal carcinoma previously named endodermal sinus tumour. (There was no yolk sac tumour within our teratoma.)
- Benign teratomas may recur after excision, but this does not necessarily imply malignancy; the clinician should maintain follow-up.

Why Teratomas?

- It has been suggested that the tissues of a teratoma derive from totipotential cells sequestered during embryogenesis.
- Another theory is that germ cells may give rise to teratomas by parthenogenetic

development.

- Or, a teratoma may originate from incomplete formation of Siamese twins.
- In our case, incomplete division of blastula may provide enough cells for the production of a teratoma but insufficient for a complete twin. This accounts for the formation of the most highly differentiated teratoma known as 'fetus in fetu', which has a reported incidence of 1 per 500,000 births.

Predisposing Factors

- There is little knowledge about predisposing factors because of the rarity of the tumour. (An incidence of between 1: 35, 000 and 1: 200, 000 live births has been suggested)
- It is generally thought that there is no ethnic or geographical pattern to incidence. Most reports do not suggest sex as a risk factor, but some described a slight female predominance, and Carney et al. found malignant teratoma to be more common in men in a ratio of 5:4.
- Some authors, including Andze et al., have reported increased α - fetoprotein concentrations prenatally, which are suspicious of a teratoma.

Diagnosis

- The teratoma may be diagnosed antenatally on ultrasound or magnetic resonance, which permits early multidisciplinary management.
- In our case, the tumour presents as a cystic, and solid lesion that originates from the palate. This diagnosis is confirmed by polyhydramnios, which is thought to be a consequence of impaired fetal swallowing and it has been proposed that cardiac decompensation caused by circulation within a large vascular teratoma may also contribute.
- Antenatal diagnosis allows a controlled environment to be planned for the birth with multidisciplinary care. In our case labour began at home, and respiratory obstruction could have been life-threatening had the nasal airway not been patent, and ventilation with bag and mask not begun.
- The differential diagnosis is limited, but includes hamartoma, dermoid cyst, and heterotopic gastrointestinal cyst. Teeth within the mass confirm teratoma.

Managements

- Resection is the treatment of choice, as there may be a small chance of malignant transformation the longer they are left.
- If the tumor presents as an obstructive mass causing respiratory embarrassment and might cause death, it demands immediate establishment of an airway, often with tracheostomy.
- The neonate's prognosis worsens as the size of the tumour increases. The ultimate prognosis of lesions with intracranial involvement is poor and operation is inappropriate.

Conclusion

Teratoma may not always be life-threatening like a small pedunculated tumour with no functional obstruction, but it may also present as an obstructive mass causing respiratory embarrassment or with intracranial involvement affecting the brain development. Clinicians should still be aware of this kind of disease in infants, though the occurrences are not very high.

題號	題目
1	Which of the following statements about the Teratoma is false? (A) Compare to the ones in adults, the neonatal teratomas have a higher benignancy. (B) If the neonates have yolk sac tumor, the clinicians should be more aware of malignancy. (C) Reoccurrence of teratomas means malignancy. (D) Calcification may be an indicator between benignancy and malignancy of the teratomas.
答 案 (C)	出處：Oral & Maxillofacial Pathology, 2nd edition p.32
題號	題目
2	Teratomas usually derivate from? (A) Ectoderm (B) Mesoderm (C) Endoderm (D) All of above
答 案 (D)	出處：Oral & Maxillofacial Pathology, 2nd edition p.32