

# True Teratoma of the Nasopharynx

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**Abstract:** Teratomas are true neoplasm composed of tissues from all the three germinal layers. Those arising from the nasopharynx are a rare entity. We describe a female neonate with a teratoma arising from the nasopharynx based on a wide stalk and presenting as a protruding mass from the oral cavity which was missed on routine antenatal ultrasonography but, which fortunately did not cause any immediate airway obstruction at birth. She was managed by surgical excision using harmonic scalpel.

## INTRODUCTION

Congenital germ cell tumors are rare with an incidence of 1 out of 4000 live births, the most common site being the sacrococcygeal region. Teratomas arising from the head and neck region constitute less than 10% of the total presenting teratomas<sup>1</sup>. Teratoma is a neoplasm composed of tissue elements which are foreign to the anatomic site from which it arises and consists of tissues derived from all 3 embryonic germ layers namely ectoderm, mesoderm and endoderm.

Routine prenatal ultrasonography may diagnose obstructive tumors before birth as early as 21 weeks of gestation, but small teratomas may be missed antenatally<sup>2</sup>. There is a danger of perinatal airway obstruction leading to neonatal mortality if these large oral tumors are not diagnosed before birth. These embryological neoplasms may also interfere with normal development and elevation of the palatal shelves resulting in a cleft palate<sup>3</sup>.

## CASE REPORT

A 3 day old female neonate presented in the otorhinolaryngological emergency services of our hospital with a large cystic mass (5cm × 4cm) protruding from the oral cavity resulting in a persistently open mouth (Fig 1) and hampering breast feeding. There was no history of respiratory distress, and child was being spoon fed with expressed breast milk. General physical examination did not reveal any other gross congenital anomaly.



**Figure 1:** Clinical photograph of the child showing the mass protruding from the oral cavity.

The child was a full term, normal spontaneous vaginal delivery born to a young primigravida with a birth weight of 2.3kgs. The antenatal period was uneventful and the routine antenatal ultrasound of the mother did not reveal any polyhydramnios or any other congenital abnormality in the fetus. There was no history of consanguinity or congenital malformations in the family.

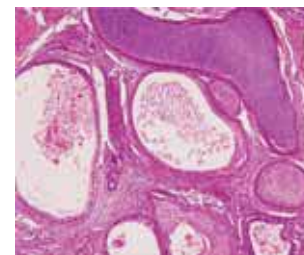
Examination of the oral cavity was difficult as the mass was completely filling it. However, a defect in the palate was noted and the mass could be seen arising with a wide stalk from the region of nasopharynx. An urgent non contrast computed tomographic scan was done, which revealed a predominantly fluid attenuating lesion filling the entire oral cavity and projecting outside with no obvious intracranial communication (Fig 2). There was an associated cleft palate. An ultrasound of the lesion showed well vascularised mass with solid and cystic areas. Serum Alfa-fetoprotein level was 50.76 ng/ml on day 7 of life.

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**Figure 2:** Computed tomographic scan of the child showing the mass arising as a stalk from the nasopharynx with a piece of bone in the stalk.

Excision of the mass from the nasopharynx was done using a harmonic scalpel, which cut and coagulated the tissue at the same time, hence causing minimal blood loss. Histopathology examination revealed a mature teratoma (Fig 3).



**Figure 3:** Histopathology picture showing mature hyaline cartilage with haphazardly placed glands and squamous epithelium.

Post operative period was uneventful. The child was kept on nasogastric tube feeds for a week, following which oral feeds were started and was discharged from the hospital. Subsequent outpatient department visits showed good healing with no residual / recurrent lesion. The child is now being planned for cleft palate repair.

## DISCUSSION

In 1863, Virchow coined the term “teratoma”, from the Greek word *teraton*, meaning “monster” and “onkoma” meaning “swelling” to describe mammoth sacrococcygeal growths<sup>4</sup>. It is a rare tumor, but occurs most commonly in the sacrococcygeal region. Teratomas of the head and neck represent approximately 6 – 10% of all teratomas and are most often found in the cervical region, followed by the nasopharynx and oropharynx. They may also arise from the sphenoid bone, hard or soft palate and the jaw. They grossly appear as heterogeneous masses with solid and cystic components and are believed to be result of, migration and entrapment of mesoderm and endoderm within ectoderm during embryogenesis of the oral cavity. These congenital tumors are composed of tissues derived from all three embryonic germ cell layers. Four basic histological types are recognized<sup>1-5</sup>: 1) Dermoid tumors (most common form, which consist of epithelial lining with skin elements composed of ectodermal and mesodermal cells) 2) Teratoid tumors (consist of all 3 germ layers but are poorly differentiated) 3) True teratoma (consist of histologically identifiable tissue from all three germ layers having solid or cystic components) and 4) Epignathus (Also known as “fetus in fetu”

or “parasitic fetus” containing developmental fetal organs and limbs. Epignathus is a misnomer and its etymological meaning is “upon the jaw.” It is however very rare and has a high mortality rate). These embryological neoplasms may interfere with normal development and elevation of the palatal shelves, resulting in a cleft palate deformity. They usually protrude through the mouth, leading to an appreciable risk of obstruction of the upper airway and death soon after birth.

Management begins prenatally as these lesions can be diagnosed on ultrasonography<sup>6</sup>, depending on the size and location of the tumor. Therefore antenatal ultrasound should be done with a high degree of suspicion, as associated polyhydramnios may not be present in all cases. Polyhydramnios occurs because of the infant’s inability to adequately swallow amniotic fluid. When a congenital oral teratoma is suspected, the child should be electively delivered via cesarean section. Airway is of prime concern at birth, which may be obstructed by a large tumor or by a smaller pedunculated tumor going posteriorly that is noticed only upon intraoral examination, and managed via nasal intubation or tracheostomy. The tumor may protrude out from the oral cavity and present with no immediate problems, as in the case here. In addition, the lesion and the palatal defect may cause subsequent feeding or speech problems, which can be dealt with later. Alternatively, ex-utero intrapartum treatment<sup>6,7</sup> procedure has been described for fetuses with giant neck masses and tracheal obstruction for safe nonemergent airway management while the fetus is still on placental support. Avoiding airway emergencies is the goal of this treatment strategy.

Computed tomographic scan and magnetic resonance imaging are the imaging methods of choice for determining the size, contents, site of

origin and extent of the tumor, in particular for assessing intracranial extension. Definitive treatment involves total excision of the tumor and appropriate reconstruction of the palatal defect at a later stage. If this is achieved, then future growth of the child will not be hampered.

## CONCLUSION

Congenital intraoral tumors are rare, but teratomas of the nasopharynx are rarer still. Diagnosis depends on antenatal clinical suspicion and pathological examination. Management in these cases is challenging, and airway management often assumes the priority. The wide range of presenting features in these cases requires an individualized approach to excision and subsequent reconstruction. Although all reported pure oral teratomas have been benign, the risk of malignant change does exist, and long-term follow-up is advisable, even if the tumor has been totally excised with free margins.

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