

Massive lingual teratoma in a neonate

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ABSTRACT

Teratoma of the tongue is a rare entity. We present a male newborn with massive lingual teratoma and cleft palate, which surprisingly did not cause immediate airway obstruction. This case illustrates a huge mass in the oral cavity, which was missed on antenatal ultrasonography because it did not present with polyhydramnios. The mass was excised under general anaesthesia. Histopathologically, it consisted of all three layers of embryonic elements with predominantly glial tissue. Postoperatively, the patient developed hypoglossal nerve palsy, and no recurrence was detected after four years.

Keywords: airway obstruction, lingual teratoma, tongue teratoma, tongue tumour

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INTRODUCTION

Teratoma is a neoplasm that arises from pluripotent cells and is composed of a wide diversity of parenchymal cell types, usually of all three germ layers. They usually contain tissues that are foreign to the organ or anatomical site from which they arise. The cause is unknown but it is postulated that a nest of pluripotential cells get sequestered during embryogenesis. The incidence is 1 in 4,000 live births with a female predominance.⁽¹⁾ The most common sites of teratomatous growth are the sacrococcygeal region, followed by ovaries and testes, retroperitoneum and mediastinum.⁽²⁾ Teratoma of the head and neck constitutes 1%–9% of all teratomas and usually arises in the cervical region.⁽³⁾ Oral teratomas specifically arising from the tongue are exceedingly rare.

Teratomas have been classified into four subtypes. These are: (1) Dermoids, which contain tissues of mesoderm and epiderm origin; (2) Teratoids, which contain tissue from three primary germ layers but is poorly-differentiated; (3) True teratomas, which are similar to teratoids but differentiated into recognisable tissues histologically; and (4) Epignathi, which are also tridermal in origin, but differentiated into recognisable organs, sometimes with limbs or even a visible second



Fig. 1 Clinical photograph taken at birth shows a multilobulated, pedunculated reddish mass protruding from the oral cavity.

foetus. Histopathological examination of previously-reported cases of teratomas of the tongue showed that the tumours were all benign in character, with no evidence of malignancy despite histological immaturity.⁽³⁾ This is in striking contrast to sacrococcygeal, gonadal, mediastinal and retroperitoneal teratomas, where a malignant component may be present in 10%–50% of patients. Moreover, teratomas of the head and neck that occur in adults are usually malignant.

Teratomas can be detected prenatally. Teratomas of the oral cavity may be associated with polyhydramnios in utero, secondary to foetal pharyngeal obstruction. Prenatal ultrasonography (US) may detect an incidental tongue mass suggestive of teratoma. Mixed echogenic signals suggestive of semisolid and semicystic components are typical findings on US. Tongue teratoma may also present with elevated maternal serum alpha-foetal protein.⁽⁴⁾ This elevation may be due to the presence of abundant extramedullary haematopoietic liver tissues.⁽⁵⁾ Definitive treatment of teratoma of the tongue is complete surgical excision. Surgery is curative and the prognosis is excellent. Recurrence has not been reported.

CASE REPORT

A baby boy was born on January 10, 2002 as a full-term spontaneous delivery, with birth weight of 3.2 kg. Antenatal US at 21 weeks and 32 weeks did not detect any abnormalities. The baby was born with

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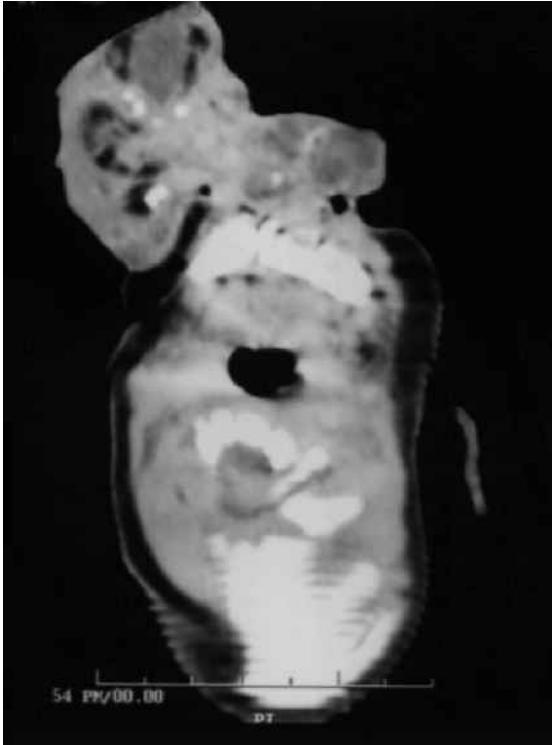


Fig. 2 Axial CT image shows a multilobulated, heterogeneous mass with calcification and fatty tissue arising from the tongue.



Fig. 3 Reconstructed sagittal CT image shows the posterior extent of the mass and patent upper airway facilitating intubation.

a big, dark red, multilobulated mass arising from the tongue, associated with a cleft palate (Fig. 1). No other dysmorphic features were noted. The Apgar score at one and five minutes were 7 and 8, respectively. The baby did not develop any respiratory distress in the first three days of life; however, he was observed in the neonatal intensive unit while preparations were made for surgery. On day three of life, the mass was noted to become bigger and at the same time, his haemoglobin level had decreased to 12 g/dL, with no

evidence of bleeding. Later on, the baby developed a couple of episodes of cyanosis and hypercapnia which necessitated intubation. Oral intubation was successful on the first attempt. Computed tomography (CT) was performed, and showed a large lobulated calcified heterogeneous mass arising from the tongue (Figs. 2 & 3).

As the patient developed signs of sepsis, operation was deferred until the seventh day when his condition was optimised. Intraoperatively the mass arose from the left lateral border of the tongue and measured 10 cm × 7 cm × 5 cm. The rest of the tongue, especially the base, was normal. Access to oral cavity was maintained with Desmoris retractors applied over the upper and lower jaws. The tongue was pulled with temporary anchor sutures, and the mass excised using a cold instrument with a few millimetres of healthy margin. Bleeding was minimal and haemostasis was achieved with the use of bipolar diathermy. Postoperatively, the patient maintained an open mouth posture, which was gradually resolved in one week. Histopathological examination showed a tumour containing embryonic elements of all three primary germ layers, i.e. ectoderm, mesoderm and endoderm. It had predominantly glial tissue. An ipsilateral hypoglossal nerve palsy was observed at the three-month follow-up (Fig. 4). Cleft palate repair was subsequently performed at the age of one year. The patient had regular follow-ups and responded well to speech therapy. No recurrence was detected after four years.

DISCUSSION

Teratomas are monstrous lesions, which contain an assemblage of tissues foreign to the part in which they arise. Teratomas in childhood are commonly found in the sacrococcygeal region, the gonads and the mediastinum, whereas in the head and neck region, these lesions are distinctly uncommon. Teratoma arising from tongue is very rare and a literature search showed only 14 other reported cases since it was first reported in 1966. In our case, glial tissue was present in the teratoma. The presence of glial tissue in lesions of the oral cavity was reported as early as 1922 and this was named Heterotrophic glioma.⁽⁶⁾ Initially, the origin of the glial tissue occurring in the oral cavity was thought to have developed from tissue following an extra-cranial separation of embryoneuroglia. It has been suggested that the glial tissue in the tongue might arise in an analogous manner to that of the tongue muscles by the dislocation of neuroectodermal cells, which accompany migrating muscle-forming cells into the tongue.⁽⁷⁾ Others suggest it come from parthenogenetic transformation of totipotential germ cells.⁽⁸⁾ The true aetiology remains unknown.



Fig. 4 Clinical photograph of the patient taken at three years of age shows no recurrence disease, but there is a left hypoglossal nerve palsy.

Teratomas are commonly diagnosed antenatally on US, with the larger ones usually associated with maternal polyhydramnios because it interfere with foetal swallowing.⁽⁹⁾ In our case, since there was no polyhydramnios, facial region abnormalities are usually not looked for on routine US. Furthermore, antenatal US appearance of a large mass may not trigger the suspicion of a non-specialised medical officer. In our part of the country, antenatal US is performed by medical officers or general practitioners, who may not readily suspect such a rare entity. The interesting discovery of a cleft palate together with an oropharyngeal teratoma has been observed in our case and previous midline teratoma cases. Any mass that develops between palatal shelves can cause secondary cleft palate. The mass interferes with normal palatal closure and this suggests that the teratomatous mass developed before the ninth week of gestational age.⁽¹⁰⁾ Other midline lesions, such as tongue hamartoma, choristoma, and encephalocele extending to the oral cavity, has also been reported to be associated with a cleft palate.

With regard to the management in this case, preoperative CT was useful in formulating a differential diagnosis, defining the tumour's origin and determining the anatomic extent of the disease. Calcifications and cysts within a mass are typical of teratomas of the head and neck than other sites. This lesion should be differentiated from encephalocele, glioma, haemangioma, congenital rhabdomyosarcoma and neurofibromatosis. Airway obstruction commonly occurs in nasopharyngeal teratomas as neonates are obligate nasal breathers. Tongue teratoma usually gives rise to feeding difficulties rather than airway problems.⁽¹¹⁾ However, respiratory distress can occur and may result in post-

natal morbidity. Immediate tracheostomy or intubation can be a lifesaving procedure if planned carefully antenatally.⁽¹²⁾ In certain centres, the ex utero intrapartum procedure is performed in an airway-compromised newborn. In our case, however, antenatal follow-up did not detect any abnormalities, and the delivery was uneventful with no respiratory embarrassment, thus such procedures were not necessary.

The aim of surgical management was to remove the diseased mass as well as to provide good airways in the long term. Retrospectively, the tumour would have been better removed with CO₂ laser. Unfortunately, at the time of this patient's presentation, our centre had not yet obtained the equipment. CO₂ laser aids in haemostasis and minimises blood loss, which is of paramount importance to the neonate. Additionally, it is also helpful in obtaining exact surgical margins while maintaining normal tissue. Excision under local anaesthesia is also possible if the mass attachment is only thin stalk.⁽¹³⁾ An important learning point is that a big oropharyngeal mass in neonate can present with normal liquor and could be missed with routine US. Furthermore, the extirpation of mass should be done as soon as possible, even though there is no initial upper airway obstruction because the problem will develop eventually.

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