



An Unusual Lesion of Epignathus with Duplicate Tongue and Ranula in a Neonate

Sudhagar Eswaran¹ · Poornima Kumar¹ · Sunil Kumar¹

Received: 13 October 2020 / Accepted: 30 November 2020 / Published online: 7 January 2021
© Association of Otolaryngologists of India 2021

Abstract We report a rare case of epignathus (oropharyngeal teratoma) in a neonate, who presented with a midline mass covered with skin and multiple hairs protruding from the Palate and associated with bifid tongue and ranula. With the characteristic presentation, diagnosis of oro/oropharyngeal teratoma was made and a massive internet search revealed very few reported cases of “epignathus”. It is unfortunate that the survival of such neonates is only moderate. Prenatal scans and follow up in an institution can prepare the multidisciplinary team to save the child. EXIT procedure to excise the mass or secure the airway, with future repair of the palate is the treatment option available. This case report emphasizes the rare clinical presentation of the disease and the prenatal diagnosis of such a condition can help in prompt decision making and management.

Keywords Oro/oropharyngeal teratoma · Epignathus · Ranula · Duplicate tongue · Prenatal scans · Head and neck tumors · Airway emergency · EXIT procedure

Introduction

Teratomas are one of the oldest tumours known to mankind. The term teratoma is derived from the Greek word “teraton” meaning monster, owing to the giant size and associated disfigurements. The teratomas are commonly seen in sacrococcygeal region, ovary, testes, retro-peritoneum and mediastinum with incidence of 1 in 35,000 live birth [1]. This tumour is very rare in head and neck region accounting for only 2–9% of all teratomas [2]. These tumours show female preponderance [2]. Embryologically these tumours arise from pluripotent stem cells lost during the migration from yolk sac in 4th and 5th gestational week [3]. They have all three germ cell layers, hence they are capable of differentiating into bone, cartilage, teeth, and skin etc.

The term “epignathus” is used for teratomas of oropharyngeal origin arising from palate, base of sphenoid and are exceedingly rare with incidence of 1 in 35,000–200,000 live births [4]. They are also associated with multiple congenital anomalies leading to high incidence of mortality in peripartum period.

Case Report

1 week old male baby born at 38 weeks of gestation by normal vaginal delivery was referred to the ENT department from neonatal ICU in view of a mass protruding from mouth. Mother was 26 year old and this was her fourth pregnancy. She was not registered and no trimester scans were done. It was a home delivery. Other children were normal and did not have any similar issues. There was no respiratory distress and vitals were normal. Weight of the baby was 2.5 kg. Ryle’s tube feeding was started as the

✉ Sudhagar Eswaran
zwaggerz@gmail.com

Poornima Kumar
poornima204@gmail.com

Sunil Kumar
suku321@rediffmail.com

¹ Lady Hardinge Medical College, New Delhi, India

child was not capable of breast feeding. On examination, there was a 7 cm pedunculated mass seen protruding out of the oral cavity and there was polydactyly (Fig. 1). The mass was seen covered with skin and multiple hair could be noted over it. It was soft in consistency and pedicle was found to be attached to the palate (Fig. 2). There was duplication of tongue with cystic swelling in floor of mouth which was suspicious of ranula (Fig. 2). CT was done and it revealed hypo dense (most probably fat filled) mass arising from palate with a defect in the palate (Figs. 3, 4). Nasal cavity was found to be normal. Provisional diagnosis of oropharyngeal teratoma (epignathus) was made. Patient was planned for cardiology assessment and then Excision of the mass and the ranula was planned with repair of the palatal defect later but neonate succumbed to the disease. The purpose of the article is to emphasize on the rarity of this entity and its associated anomalies as seen in our case.

Discussion

Teratomas are germ cell tumours commonly seen in the sacrococcygeal region. Head and neck teratomas is a rare clinical entity. It is reported to be common in females than males [5]. In this case report, the neonate was a male patient. Oropharyngeal teratomas arising from the palate and base of sphenoid bone regions are termed as Epignathi [5].

The location of the tumour can jeopardize the neonate's airway and cause feeding difficulties in the post natal period. This tumour is associated with various other congenital anomalies like duplicate tongue, bifid uvula, ranula, congenital heart disease etc., and most often these neonates fail to survive because of these associated comorbidities and airway obstruction [5]. These tumours can be seen with syndromes like Pierre Robin syndrome, Down's syndrome

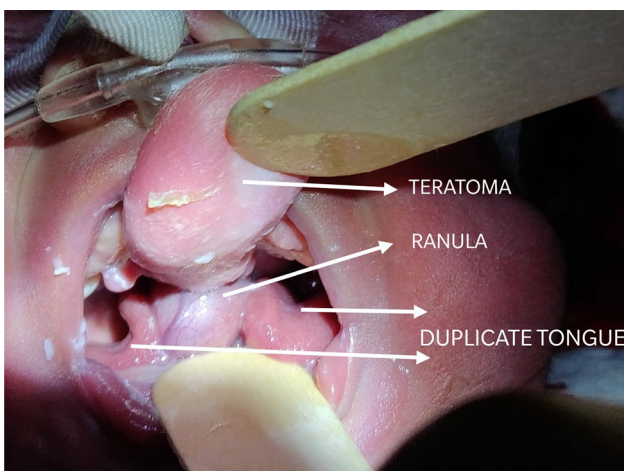


Fig. 1 Epignathus with ranula and duplicate tongue



Fig. 2 Skin lined midline protruding mass from the palate with multiple hairs over it characteristic of teratoma



Fig. 3 3D reconstructed image

and Trisomy 13 [6]. In this case, the neonate had a bifid tongue, ranula, cleft palate and polydactyly.

Pathogenesis is due to the faulty migration of primordial germ cells during fourth and fifth week of gestation [7]. These tumours can have various maturation level and it is possible to find tissues arising from all the germ layers at different stages of maturation [8]. They can be classified as Mature and Immature types on the basis of differentiation [9]. Mature type reveals skin and its appendages, fat, muscle, cartilage and bone whereas in immature type these

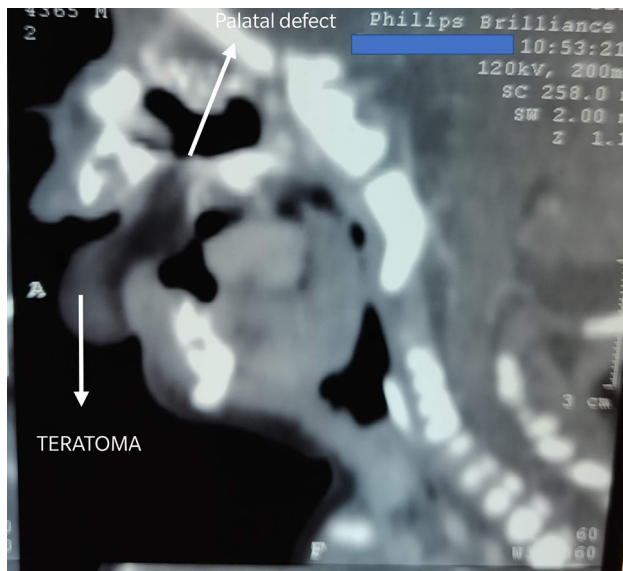


Fig. 4 Hypodense mass (fat filled) attached to the palate and with palatal defect

differentiation are not complete. The malignant transformation of this lesion is a very rare condition and commonly seen in the sacrococcygeal region [10].

Prenatal diagnosis plays a vital role in the management. Teratomas are often associated with polyhydramnios. Ultrasound and MRI are useful radiological investigations to confirm the diagnosis. In this patient, there was no history of registration of pregnancy and the trimester scans. Early diagnosis can help in the holistic planning of the peripartum management. Alpha fetoprotein can be used as a prognostic tumour marker in such lesions. Very high value of alpha fetoprotein is diagnostic of malignant transformation [11].

In case, large mass with propensity to cause airway obstruction is diagnosed in the trimester scans, then the planning of EXIT (Ex-utero Intrapartum Treatment) procedure or OOPS (Operation on Placental Support) is to be made [12]. Neonates airway is secured by tracheostomy or resection of the tumour while maintaining uteroplacental circulation. In cases where the diagnosis is made after birth, multidisciplinary approach is needed to resuscitate the neonate and establish the airway. Team of Neonatologist, NICU and ENT surgeons with Anaesthetist are needed in immediate management of the airway obstruction and then plan for teratoma excision. In some cases where the mass is not causing respiratory distress, excision of mass and the repair of defect should be planned. These neonates will need a nasogastric tube for feeding. But some neonates don't survive and succumb to the disease before or in the perioperative period.

Teratomas are rare tumours and in a massive internet search only few cases of Epignathus were found. Early diagnosis in the prenatal period plays a crucial part in the

peripartum management. The awareness of importance of trimester scans should be known by the common people. In conclusion, teratomas are rare growths in head and neck region with poor prognosis because of the associated anomalies, respiratory distress after birth and feeding difficulties. Holistic planning is needed to save the neonate and judicious counselling in the pre natal period is crucial for the parents as it will be a disheartening news to them about their baby. This case report emphasises the rarity of the tumour and the importance of prudent pre natal assessment with trimester scans, with which the diagnosis can be made early and the management can be planned in a multi disciplinary approach.

Compliance with ethical standards

Conflict of interest None.

References

- Weaver RG, Meyerhoff WL, Gates GA (1976) Teratomas of the head and neck. *Surg Forum* 27:539–544
- Kountakis SE, Minotti AM, Maillard A (1994) Teratomas of the head and neck. *Am J Otolaryngol* 15:292–296
- Rai M, Hegde P, Devaraju UM (2012) Congenital facial teratoma. *J Maxillofac Oral Surg* 11:243–246
- Prevedello MD, Kassam BA, Carrau LR, Snyderman HC, Thomas A, Garner P (2007) Transpalatal endoscopic endonasal resection of a giant epignathus skull base teratoma in a new born. *J Neurosurg* 107:266–271
- Vandehaute B, Leteurtre E, Leconte-Houche M, Pallerin P, Nayts JP, Luiset JM et al (2000) Epignathus teratoma: report of three cases with a review of the literature. *Cleft Palate-Craniofac J* 37:83–91
- Sanchez AV, Moreno HC, Salfado AS, Serena JC, Irra EJ, Sanchez PG (2000) Epignathus. *Cleft Palate Craniofac J* 37:83–91
- Beutel K, Partsch CJ, Janig U, Nikischin W, Suttrop M (2001) Oral mature teratoma containing epididymal tissue in female neonate. *Lancet* 357:283–284
- Rothschild MA, Catalano P, Urken M, Brandwein M, Som P, Norton K et al (1994) Evaluation and management of congenital cervical teratoma. Case report and review. *Arch Otolaryngol Head Neck Surg* 120(4):444–448
- Lionel J, Valvoda M, Al-Abdul Hadi KA (2004) Giant epignathus: a case report. *Kuwait Med J* 36(3):217–220
- Tonni G, De Felice C, Centini G, Ginanneschi C (2010) Cervical and oral teratoma in the fetus: a systematic review of etiology, pathology, diagnosis, treatment and prognosis. *Arch Gynecol Obstet* 282:355–361
- Dakpé S, Demeer B, Cordonnier C, Devauchelle B (2014) Emergency management of a congenital teratoma of the oral cavity at birth and three-year follow-up. *Int J Oral Maxillofac Surg* 43:433–436
- Santana EF, Helfer TM, Piassi Passos J, Araujo Júnior E (2014) Prenatal diagnosis of a giant epignathus teratoma in the third trimester of pregnancy using three-dimensional ultrasound and magnetic resonance imaging: case report. *Med Ultrason* 16:168–171

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.