Head and neck teratomas

Received: 16 September 2008 / Accepted: 23 January 2009
© Association of Oral and Maxillofacial Surgeons of India 2009

Ajaz Shah¹ · Suhail Latoo² ·
Irshad Ahmed¹ · Altaf.H.Malik¹

¹ Dept. of Oral and Maxillofacial Surgery
² Dept. of Oral Pathology and Microbiology

Govt. Dental College, Srinagar,
Jammu and Kashmir, India

Abstract Teratomas are complex lesions composed of diverse tissues from all 3 germinal cell layers and may exhibit variable levels of maturity. Head and neck teratomas are most commonly cervical with the oropharynx (epignathus) being the second commonest location. In this article, clinical presentation, behaviour and associated significance of head and neck teratomas have been highlighted. Because of their obscure origin, bizarre microscopic appearance, unpredictable behaviour and often dramatic clinical presentation, a dental surgeon or a dental specialist should be aware of their clinical presentation and pathology.

Keywords Teratomas · Epignathus

Address for correspondence:

Ajaz Shah
Dept. of Oral and Maxillofacial Surgery
Govt. Dental College, Srinagar
Jammu and Kashmir
India

Introduction

Teratomas are complex lesions derived from totipotential cells. They are composed of diverse tissues from all 3 germinal cell layers and may exhibit variable levels of maturity. The word itself is derived from the Greek word teraton, meaning monster, and was used initially by Virchow in the first edition of his book on tumors, which was published in 1863 [1].

Teratomas occur in 1 of 4000 live births, demonstrating a female predominance. Teratomas are most commonly located in the sacrococcygeal region, followed by the ovaries, testis, anterior mediastinum, retroperitoneum, and finally the head and neck, which account for less than 5% [1,2].

Head and neck teratomas are most commonly cervical with the oropharynx (epignathus) being the second commonest location. Etymologically, the term 'epignathus tumor' should be reserved for tumors arising from the jaw. Ehrich (1945) classified oropharyngeal tumors according to the site of origin as episphenoid, epipalatine or epuranus, and epignathus tumors. However, the term epignathus tumor is generally applied to teratomas of the oropharyngeal cavity in neonates without specifying the site of origin [3,4,5].

Teratomas of the head and neck are interesting because of their obscure origin, bizarre microscopic appearance, unpredictable behaviour and often dramatic clinical presentation. Since teratomas of the

head and neck frequently present with respiratory distress, a dental surgeon should be aware of their clinical presentation and pathology.

Etiopathogenesis

There are several theories regarding the histogenesis of teratoma formation. Ahfeld first proposed the twin theory in the late 1800's [6–8]. Schwalbe later suggested that teratomas arise, during embryogenesis, from displaced germ cells during migration to the genital ridge. The most popular theory speculates that teratomas arise from totipotential embryonic tissues that are somehow displaced during ontogeny [6].

For head and neck teratomas, the theory postulates that embryonic tissue in proximity to the primitive streak and notochord somehow loses its governing influence over the totipotential cells, allowing unorganized differentiation. This is presumably due to alterations in cellular membrane chemistry [3].

Classification

A significant degree of confusion has arisen regarding nomenclature for the various subtypes of teratomas. In the late 19th century, Dr. Arnold derived the classification system for teratoid malformations. The system consists of four

groups: dermoid cysts, teratoid cysts, true teratomas and epignathi [9].

Teratomas range from benign, well-differentiated (mature) cystic lesions to those that are solid and malignant (immature). Additionally, teratomas may be monodermal and highly specialized and, rarely within some mature teratomas, may contain elements (most commonly squamous components) undergo malignant transformation [1].

Epidemiology

Cervical teratomas

The reported incidence of cervical teratomas ranges from 2.3% to 9.3% of the total. Only 217 cases of cervical teratomas have been reported in the world literature, of which 86 were in females and 70 in males. All races have been represented and there is no obvious genetic predisposition. Ninety percent of cases are diagnosed in the pediatric population; however, an adult form does exist [10–12].

Oropharyngeal teratomas

The second most common head and neck site for teratoma formation is the nasopharynx. It was first described in medical literature by Dr. Ahfeld in 1874 and only 30 cases have been reported in medical literature [13]. According to Levine et al.

(1990), the incidence of epignathus tumor is between 1 in 35,000 and 1 in 200,000 live births [14]. Nogales et al. (1977) reported a single case of epignathus tumor in 800,000 deliveries in east Andalusia [7]. These lesions are not familial in nature. A marked female predominance of six to one has been cited [15].

Epignathus teratoma occurs more frequently in young mothers and is associated with polyhydramnios secondary to swallowing difficulties during pregnancy. Placental edema may be associated with an epignathus tumor because of fetal cardiac decompensation secondary to the extensive vascularization of the tumor. Cases of pre-eclampsia, and an increase in amniotic α -fetoprotein have been described [16].

The site of origin appears to be the craniopharyngeal canal, a defect of the sphenoid bone, where in embryonic life; the buccopharyngeal membrane, Rathke's pouch, and the notochord are closely related. The implantation base of the tumor may be single or multiple [17]. The majority of epignathus tumors have their point of attachment at the base of the skull in the posterior region of the nasopharynx, near Rathke's pouch and the craniopharyngeal canal, on the midline. They usually involve the hard palate or sphenoid bone [14]. These tumors are often accompanied by maxillary deformity, even if the implantation base is not located in the bones of the face [18]. Few cases have an intracranial extension along the craniopharyngeal canal. These intracranial extensions are generally fatal [15,19,20].

Clinical features

Cervical teratomas

They often present as midline mass with lateral extension and can measure more than 10 cm in diameter. Palpation usually reveals a firm, mobile, multilobular, cystic mass with well-defined margins. Respiratory and obstetrical complications are frequent. The respiratory symptoms range from mild positional dyspnea to complete apnea. Obstetrical complications include preterm labor, polyhydramnios and birth dystocia. The polyhydramnios is thought to arise from interference of the mass with normal fetal swallowing and breathing. The larger masses are more likely to cause complications, but small teratomas can grow explosively, causing precipitous decompensation [10–12,21,22].

Associated malformations are rare but have been described. Only one case of metastatic disease has been reported [23]. Seventy-five percent of cervical teratomas present during the neonatal period [10–12].

Oropharyngeal teratomas

The clinical presentation of oropharyngeal teratomas depends on the size and location of the mass. Clinical manifestations can range from failure to thrive and mild positional dyspnea to severe airway compromise. Nasopharyngeal teratomas may be sessile or pedunculated, and have been reported to protrude from the mouth and nares. Diagnosis is usually made early in infancy and rarely after 2 years of age [4,24].

Teratomas of the nasopharynx are often associated with cranial structural anomalies. Palatal fissures are common and hemicrania and anencephaly have been reported. It is theorized that the teratoma physically interferes with the normal fusion of embryonic tissues in the early developmental period [3,25,26]. The cleft palate of mechanical origin is the most common associated anomaly [14]. The tumor prevents closure of the palate, [15,27] whereas normally the palatine processes join in the midline and merge to form the secondary palate during the eighth week [28]. Among the malformations associated with epignathus tumor are bifid tongues and even noses [29,30]. The median bud between the two halves of the tongue represents nonfusion of the three embryological buds of the tongue. Bifidity and glossoptosis indicate very early development of the tumor, which prevents fusion of the primitive tongue buds and anterior positioning of the tongue. The tongue normally appears at about the fourth week of development and is positioned in the floor of the mouth during the sixth to eighth week. The mandible appears subsequently. These two observations of micrognathia in circumstances where glossoptosis and cleft palate are apparently secondary to the presence of a congenital tumor support the opinion that in many of cases of Pierre Robin sequence, functional disorders secondary to glossoptosis are responsible for the microretrognathia. This is in keeping with the opinion of Enlow (1975) and others about the role of the tongue in the growth of the horizontal ramus and symphysis of the lower jaw. Functional disorders secondary to glossoptosis apparently can prevent growth

of the jaw, resulting in mandibular microretrognathia. Cleft palate, glossoptosis, and microretrognathia are pathognomonic for Pierre Robin sequence. Duplication of the pituitary gland is rare. Seven cases are described in the literature; five of these were associated with an epignathus teratoma with or without agenesis of the corpus callosum. Despite the persistent malformations, the child's weight, height, and neurobehavioral development are satisfactory [2,20,29].

Epignathus teratomas are rare congenital malformations giving rise to oropharyngeal tumors that result in life-threatening early neonatal asphyxia [4].

Radiographic findings

Cervical teratomas

Timely and accurate diagnosis is critical so therapy is not delayed. Evaluation should precede after the airway is secured. This may require endotracheal intubation, bronchoscopy, or tracheostomy. Plain radiographs demonstrate calcifications in 16% of cases and is diagnostic when present. Tracheal deviation is seen in more than 50% of cases. Ultrasonography can be performed at the bedside without sedation or radiation, and can reveal a complex neck mass with cystic and solid components. CT reveals differing densities and scattered calcifications. In addition to defining the location and extension of the tumor, delineation of distorted normal tissues and evaluation of cervical lymph nodes and the mediastinum is also possible. The role for MRI has not been described in the evaluation of cervical teratoma [31].

Nasopharyngeal teratomas

Prior to evaluation the airway must be secured. This can be accomplished with an oral airway, oral intubation, or, if necessary, a tracheostomy. Plain radiographs and sonography are sometimes helpful. Encephalocele is probably the most important preoperative diagnosis to make. Computerized tomography helps define the extent of the lesion as well as intracranial extension. The teratomas show cystic and solid areas and areas of fat density on CT and MRI. Areas of bone formation are also seen. A teratoma can be suspected when a multiloculated lesion with focal areas of low attenuation and high intensity is seen on CT and T1 W MR images respectively. Areas of

bone and tooth formation are characteristic and these are in the form of mature tissue unlike other bone forming tumors. The primary differential diagnostic considerations for a suspected cervical mass are cystic hygroma, branchial cleft cyst, thyroglossal duct cyst (all noncalcified and mainly cystic) and hemangioma (calcified and mainly solid). When the nasopharyngeal origin becomes clear the differential consideration is an encephalocele or meningoencephalocele. Developmental branchial and thyroglossal cyst are often multilocular with only a thin rim of enhancement and do not contain fat. Occasionally these cysts may be septated and therefore mimic a multilocular teratoma. Thyroglossal duct cysts tend to be oblong or roughly tubular, oriented vertically along the path of migration of the primitive thyroid gland. Hemangiomas are masses of muscle density on CT which may or may not enhance. Phleboliths if present are diagnostic. A meningoencephalocele shows intracranial continuation through a bony defect in the skull base and has CSF density areas within. Further there is distortion of intracranial structures and evident continuity with intracranial structures. If doubt persists the diagnosis can be confirmed by aspiration and chemical analysis of fluid which resembles CSF in composition [3,32].

Pathology

Teratomas have heterogeneous histological characteristics. Mature teratomas may contain all manners of adult tissues with varying degrees of organoid development, whereas immature lesions may produce embryonic or extra embryonic fetal tissue. Upto 90% of childhood teratomas contain derivatives from all three embryonic germ layers. Of childhood teratomas 20–40% contains some immature tissues. In childhood, 75–85% of teratomas of the head and neck region usually contain neuroectodermal elements (both mature and immature). Approximately 80% are benign and 20% are malignant [1].

Treatment and prognosis

Surgical resection is the treatment of choice for all head and neck teratomas. Because of the potential for rapid growth of the cystic component and airway compromise, the treatment of cervical and nasopharyngeal teratomas is immediate excision [33].

Malignant degeneration of teratomas is estimated to be between 5% and 30% [34]. It has never been described in association with epignathus teratomas [7,14].

The significance of immature elements in the surgical specimen is unclear since malignancy and recurrence have not been reported after local excision of head and neck teratomas. The most important complication of oral teratomas is respiratory compromise, which is the main cause of death in neonates. In addition, oral teratomas may interfere with swallowing [1].

Conclusion

Teratomas of the head and neck are rare neoplasms, usually presenting in the neonatal period. Though most tumors are benign, they are often malignant with regard to their location. They can enlarge rapidly, so prompt airway intervention is important. The diagnosis needs to be made promptly, usually with the aid of computerized tomography, followed by excisional biopsy.

References

- Gnepp DR (1985) Teratoid neoplasms of the head and neck In: Barnes L, Surgical pathology of the head and neck. New York: Marcel Dekker 1411–1433
- Shah FA, Raghuram K, Suriyakumar G, Dave AN, Patel VB (2002) Congenital Teratoma of Nasopharynx. *Ind J Radiol Imag* 12(2): 201–202
- Vandenhoute B, Leteurtre E, Lecomte-Houcke M, Pellerin P, Nuyts JP, Cuisset JM, Soto-Ares G (2000) Epignathus Teratoma: Report of three cases with a review of the literature. *Cleft Palate-Craniofac J* 37(1): 83–91
- Keswani RK, Chugh TD, Dhall JC, Mehrotra GC (1968) Epignathus: A case report. *Br J Plast Surg* 21(4): 355–359
- Chervenak FA, Tortora M, Moya FR, Hobbins JC (1984) Antenatal sonographic diagnosis of epignathus. *J Ultrasound Med* 3(5): 235–237
- Ashley DJ (1973) Origin of teratoma. *Cancer* 32(2): 390–394
- Nogales FF Jr, Gasca L, Lamas R, Blanco A, Sanz JF (1977) Case report: Epignathus, clinical, radiologic, and pathologic considerations. *Int J Gynecol Obstet* 15(1): 41–43
- Senyuz OF, Rizalar R, Celayir S, Oz F (1992) Fetus in fetu or giant epignathus protruding from the mouth. *J Pediatr Surg* 27(12): 1493–1495
- Taylor BW, Erich JB, Dockerty MB (1966) Dermoids of the head and neck. *Minn Med* 49(10): 1535–1540
- Jordan RB, Gauderer MW (1988) Cervical teratomas: An analysis, literature review and proposed classification. *J Pediatr Surg* 23(6): 583–591
- Abemayor E, Newman A, Bergstrom L, Dudley J, Magidson JG, Ljung BM (1984) Teratomas of the head and neck in childhood. *Laryngoscope* 94(11pt1): 1489–1492
- Hawkins DB, Park R (1972) Teratoma of the pharynx and neck. *Ann Otol Rhinol Laryngol* 81(6): 848–853
- Lopes MA, Pereira CM, da Cruz Perez DE, Vargas PA, de Almeida OP (2005) Benign teratoma of the buccal mucosa in a 9-year-old girl: Report of case and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 100(5): 598–602
- Levine AB, Alvarez M, Wedgwood J, Berkowitz RL, Holzman I (1990) Contemporary management of a potentially lethal fetal anomaly: a successful perinatal approach to epignathus. *Obstet Gynecol* 76 (5 Pt 2): 962–966
- Ng HN, Ong CL (1993) Two case reports of intracranial teratoma diagnosed antenatally. *Ann Acad Med Singapore* 22(5): 823–825
- Kaplan C, Perlmutter S, Molinoff S (1980) Epignathus with placental hydrops. *Arch Pathol Lab Med* 104(7): 374–375
- Ekici E, Soysal M, Kara S, Dogan M, Gokmen O (1996) Prenatal diagnosis of epignathus causing acute polyhydramnios. *Acta Obstet Gynecol Scand* 75(5): 498–501
- Zakaria MA (1986) Epignathus (congenital teratoma of the hard palate): A case report. *Br J Oral Maxillofac Surg* 24(4): 272–276
- Catalano PJ, Urken ML, Alvarez M, Norton K, Wedgwood J, Holzman I, Biller H (1992) New approach to the management of airway obstruction in 'high risk' neonates. *Arch Otolaryngol Head Neck Surg* 118(3): 306–309
- Smith NM, Chambers SE, Billson VR, Laing I, West CP, Bell JE (1993) Oral teratoma (epignathus) with intracranial extension: a report of two cases. *Prenat Diagn* 13(10): 945–952

21. Carney JA, Thompson DP, Johnson CL, Lynn HB (1972) Teratomas in children: clinical and pathologic aspects. *J Pediatr Surg* 7(3): 271–282
22. Holt GR, Holt JE, Weaver RG (1979) Dermoids and teratomas of the head and neck. *Ear Nose Throat J* 58(12): 37–60
23. Touran T, Applebaum H, Frost DB, Richardson R, Taber P, Rowland J (1989) Congenital metastatic cervical teratoma: Diagnostic and Management considerations. *J Pediatr Surg* 24(1): 21–23
24. Sengupta P, Mukhopadhyay SD, Palit T, Khasnobis PK (1982) Epignathus. *J Indian Med Assoc* 78(12): 198–200
25. Papageorgiou C, Papathanasiou K, Panidis D, Vlassis G (2000) Prenatal diagnosis of epignathus in the first half of pregnancy : A case report and review of the literature. *Clin Exp Obstet Gynecol* 27(1): 67–68
26. Ramirez Robles LJ, Gomez Partida G, Trujillo Gomez JJ (2001) Prenatal diagnosis of an epignathus: A case report and review of the literature. *Ultrasound Obstet Gynecol.* 18(2): 178–181
27. Bennett JP (1970) A case of epignathus with long term survival. *Br J Plast Surg* 23(4): 360–364
28. Langman J (1984) Tête et cou. In: *Embryologie Médicale*. 4th ed. Paris: Masson 301–330
29. Bale MP, Reye RD (1976) Epignathus, doublepituitary and agenesis of corpus callosum. *J Pathol* 120(3): 161–164
30. Hirshowitz B, Malher D, Heifetz M (1973) Epignathus and bifid nose: a report of a rare congenital anomaly. *Br J Plast Surg* 26(3): 271–273
31. Lusk RP, Lee PC (1986) Magnetic resonance imaging of congenital midline nasal masses. *Otolaryngol Head Neck Surg* 95(3 pt 1): 303–306
32. Jund R, Leunig A, Rohrbach H, Rasp G (2001) Nasopharyngeal epignathus causing nasal airway blocking in a 4-year-old child. *Rhinology* 39(2): 112–114
33. Mahour GH, Wooley MM, Trivedi SN, Landing BH (1974) Teratomas in infancy and childhood: experience with 81 cases. *Surg* 76(2): 309–318
34. Pavlin JE, O’Gorman A, Williams HB, Crepeau RJ, Shapiro RS (1984) Epignathus: A report of two cases. *Ann Plast Surg* 13(5): 452–456

Source of Support: Nil, **Conflict of interest:** None declared.