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Nasopharyngeal epignathus causing nasal airway blocking in a 4-year old child*

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SUMMARY

Congenital oral teratoma or epignathus is a very rare lesion, especially in older children. We report on a 4-year old female infant presenting with nasal blocking, little watery discharge and recurrent otitis. First adenoids were suspected. An endoscopic examination of the nose and nasopharynx demonstrated an irregular mass in the nasopharynx, covered partly by mucosa. A MRI-scan (T1-weighted) showed fat-intensity. Histological examination revealed a teratomatous malformation.
Although rare, teratoma should be included in the differential diagnosis of a nasopharyngeal mass in the paediatric age group.

Key words: epignathus, teratoma, nasopharynx, adenoids

INTRODUCTION

Congenital oral teratoma, also referred to as epignathus, is an unusual lesion estimated to affect 1 in 200.000 live births (Holt et al., 1979). Neonatal asphyxia is the usual presentation of this uncommon entity with a high mortality. In most cases it is detected prenatally by ultrasound or immediately after birth. We present a case of nasopharyngeal epignathus and its diagnostic work-up in a 4-year old child.

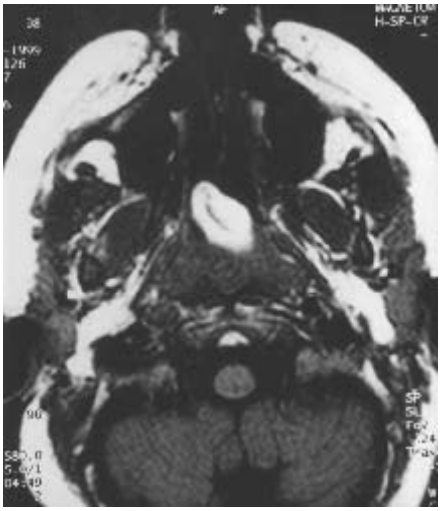

Figure 1a. MRI-scan, axial, T1-weighted. The lesion in the nasopharyngeal space could be seen.

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CASE REPORT

A 4-year old female child was referred to us to perform an adenotomy. It had a history of nasal breathing difficulties, snoring and recurrent otitis media. The child was otherwise healthy and without evidence of disease. It was born after an uneventful pregnancy to a 28-year old mother and her 33-year old husband. Both were Caucasian and had no known hereditary disease.
Nasal endoscopy showed a solid mass, which filled nearly the whole nasopharyngeal space. Both tympanic membranes showed a slight retraction. No other abnormality was recorded.

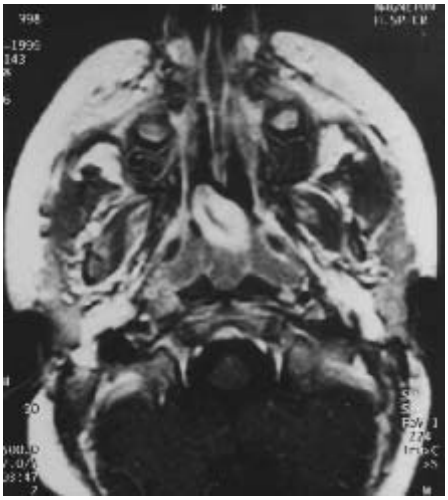

Figure 1b. MRI-scan, axial, T1-weighted with contrast media.

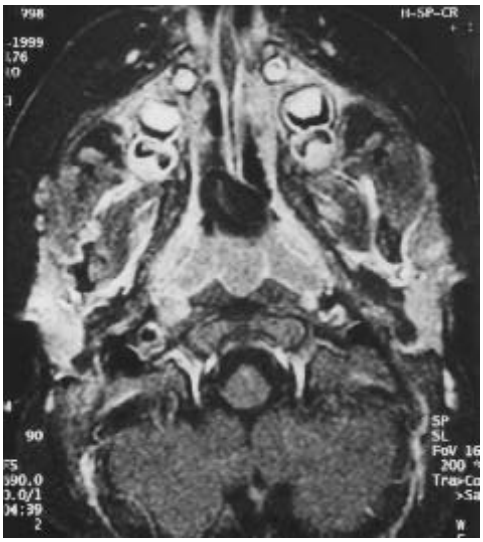

Figure 2. MRI-scan, axial, T1-weighted with fat saturation.

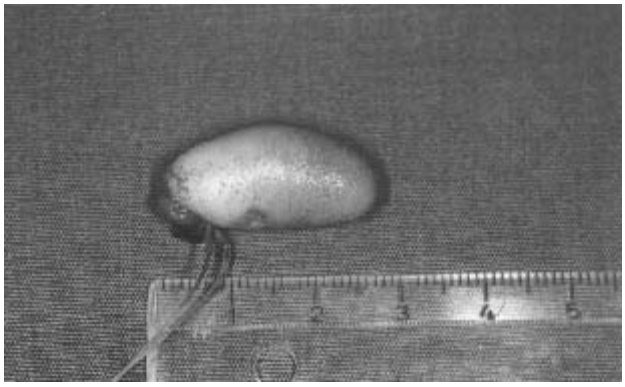

Figure 3. Macroscopic specimen.

To receive further data about the mass, we performed a MRI scan of the skull. In an axial T1-weighted scan, a hyper intensive mass, located in the nasopharyngeal space was seen (Figure 1a). Application of contrast media showed no further enhancement (Figure 1b). The mass showed central hypo intensity, with the presence of a different tissue, e.g. cartilage. A T1-weighted scan with fat saturation demonstrated a fat specific behaviour (Figure 2).

So it was suspected that the mass was highly constituted of fat and no increased vessel density could be seen. Resection of the tumour was performed by exposure of the nasopharynx via a paramedian incision of the soft palate.

The macroscopic specimen was a 2.5 cm long solid tumour, coated with mucosa (Figure 3).

Histological examination revealed different tissues, including fat, hairy structures and cartilage (Figure 4). A squamous epithelial layer coats the tissues. The histological diagnosis was an epignathus.

The child is well and without evidence of local recurrence one year after initial presentation and surgery.

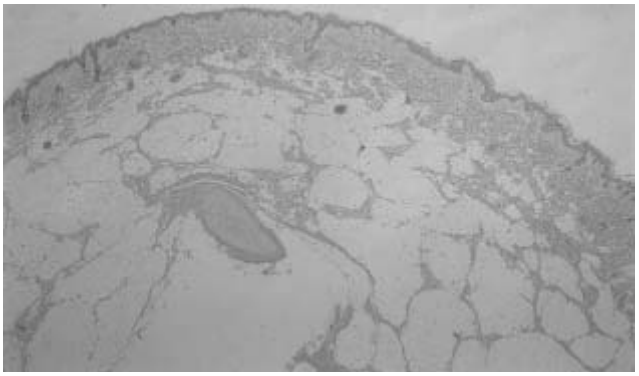

Figure 4. Histological picture of the specimen (hematoxylin-eosin, × 300).

DISCUSSION

Although teratomas are the most common congenital neoplasms with an incidence of 1:20.000 to 1:40.000 live births (Tapper and Lack, 1983), oropharyngeal teratoma, also referred to as epignathus, is a much rarer type. The term was advocated by Ehrich for "teratoid parasites of the mouth" (Ehrich, 1945). It carries a grave prognosis (Chervenak et al., 1985; Teal et al., 1988).

These lesions arise from the sphenoid bone and hard palate, grow into the oral and nasal cavities and may extend intracranially.

As the tumour usually is congenital, it could be detected by ultrasound scan, performed for estimation of gestational age and maturation (Smith et al., 1993; Brühwiler et al., 1995). Polyhydramnios is often an associated abnormality probably reflecting impaired fatal swallowing (Teal et al., 1988; Levine et al., 1990). In smaller cases, like this one, clinical symptoms might mimic adenoids. Frequently these lesions are unresectable and infants born with these lesions usually die secondary to respiratory compromise (Teal et al., 1988).

There are no associated karyotype abnormalities and they are not thought to be inherited in a Mendelian or polygenic fashion. Therefore obstetricians may reassure parents that they have no increased risk of bearing another child with this lesion (Ekici et al., 1996).

Histologically, the majority are benign teratomas composed of poorly organized tissues derived from each of the three layers of the embryonic disc (Potter and Craig, 1976). Cytogenetic studies of epignathi and other extragonadal teratomas have shown karyotypes identical to that of the host, suggesting an origin from a diploid totipotential cell dividing by mitosis (Levine et al., 1990). The suggested site of origin is the craniopharyngeal canal, a defect in the sphenoid bone at which in embryonic life the buccopharyngeal membrane, Rathke's pouch, and the rostral notochord are in close approximation (Wilson and Gehweiler, 1970).

The lesion is able to continue through the skull base and can reach an intracranial extension (Smith et al., 1993).

The present material is remarkable in respect of the detection of the tumour at 4 years of age, mimicking adenoids. MRI can visualize the precise anatomical plane and the tissue composi-

tion. It allows the detection of different tissues like fat, cartilage and bone within the mass. It can be used for prenatal and postnatal diagnosis of epignathus (Williamson et al., 1989). As usually the patients are infants, MRI is preferred to prevent exposure to radiation.

Successful surgical treatment of epignathus was first reported in 1951 (Ochsner and Ayres, 1951). To gain access to the whole nasopharynx and some blood vessels, we performed an exposure of the lesion via a transpalatinal approach. A transnasal endoscopic approach would not allow a removal of the mass in one piece.

To prevent invasion of the skull base or facial deformities, early surgery is advocated.

Our case shows, that epignathus should be included in the differential diagnosis of nasopharyngeal masses of children.

REFERENCES

1. Brühwiler H, Mueller MD, Rabner M (1995) Ultraschalldiagnose eines Epignathus in der 17. SSW. Ultraschall in Med 16: 238-140.
2. Chervenak FA, Isaacson G, Touloukian R (1985) Diagnosis and management of fatal teratomas. Obstet Gynecol 66: 666-669.
3. Ehrich, WE (1945) Teratoid parasites of the mouth (epishenoids, epipalati, epignathi). Am J Orthodont Oral Surg 31: 650-659.
4. Ekici E, Soysal M, Kara S, Dogan M, Gokmen O (1996) Prenatal diagnosis of epignathus causing acute polyhydramnios. Acta Obstet Gynecol Scand 75: 498-501.
5. Holt, GR, Holt JE, Weaver RG (1979) Dermoids and teratomas of the head and neck. Ear Nose Throat J 58: 520-531.
6. Levine B, Alvares M, Wedgwood J (1990) Contemporary management of a potentially lethal fatal anomaly: A successful approach to epignathus. Obstet Gynecol 76: 962-966.
7. Ochsner A, Ayers WB (1951) Case of epignathus: survival of host after its excision. Surgery 30: 560-564.
8. Potter El, Craig JM (1976) Pathology of the foetus and the infant (ed 3). Chicago, IL, Year Book pp 188-236.
9. 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: 945-952.
10. Tapper D, Lack EE (1983) Teratomas in infancy and childhood. A 54-year experience at the Children's Hospital Medical Centre. Ann Surg 198: 398-400.
11. Teal LN, Angtuaco TL, Jimenez JF, Quirk JG (1988) Fatal teratomas: Antenatal diagnosis and clinical management. J Clin Ultrasound 16: 329-336.
12. Williamson RA, Weiner CP, Yuh WTC (1989) Magnetic resonance imaging of the anomalous foetuses. Obstet Gynecol 73: 952-956.
13. Wilson RA, Gehweiler JA (1970) Teratoma of the face associated with a patent canal extending into the cranial cavity (Rathke's pouch) in a three-week-old child. J Pediatr Surg 5: 349-359.

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