Osteocartilaginous choristoma: a case report

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Abstract

A choristoma is a benign tumour-like mass consisting of mature tissue derived from one or more germ cell layers that are foreign to the site at which they are located. Choristomas of the pharynx are rare with few cases being reported in the English literature. Management of these lesions is usually complete surgical excision. We report a case of osteocartilaginous choristoma arising from the pharynx.

Key words: Choristoma; Teratoma; Nasopharynx

Introduction

A choristoma is a tumour-like mass of normal tissue developed in an abnormal location. This is different from a hamartoma which is a tumour-like mass consisting of disorganized mature tissue which is indigenous to the site at which the lesion occurs. A choristoma is also different to a teratoma. The components of a teratoma are haphazard whereas the tissues forming a choristoma are precisely arranged.¹ A teratoma is composed of derivatives from all three germ cell layers² and can have malignant potential.

An osteocartilaginous choristoma is a benign tumour consisting of bone and cartilage where bone and cartilage do not normally occur. We wish to report a case of an osteocartilaginous choristoma in the pharynx of a male presenting with a sore throat and dysphagia.

After searching Pubmed, Medline and Embase we present this rare case of pharyngeal osteocartilaginous choristoma. Previously reported cases of osteocartilaginous choristomas have occurred in the mouth or oropharynx.^{3,4} Choristomas occurring in the nasopharynx have consisted of skin and associated structures^{5,6} and salivary gland tissue.⁷ We report a case of osteocartilaginous choristoma occurring on the posterior wall of the pharynx.

Case report

A 59-year-old man referred to ear, nose and throat outpatients in November 2000. He gave a two, month history of left sided throat pain, unresponsive to antibiotics. On questioning, he said that he had suffered from recurrent sore throats for the last 18 years. He had mild pain on swallowing, but no earache and found that food collected in his left tonsillar area. There were no abnormalities on examination. He was listed for an examination of his pharynx under anaesthesia. He underwent an examination under anaesthesia (EUA) of his pharynx. A firm chord like tumour was identified on the posterior pharyngeal wall from which a biopsy was taken. Three portions of grey-brown tissue were received by the pathology department. They measured $2.5 \times 1.0 \times 1.0$ cm in aggregate. Histological analysis of the sections taken from the tissue revealed fragments of tissue partly lined by stratified squamous epithelium. The underlying tissue contained small minor salivary gland type tissue in one area. In addition, there was a large area composed principally of fibrous tissue and mature adipose tissue. Within this area there were small nodules of hyaline cartilage. One of the nodules of cartilage was associated with a fragment of cancellous type bone (Figure 1). The histological features were those of an osteocartilaginous choristoma.

A magnetic resonance image (MRI) scan of his neck showed a fusiform, prevertebral mass and revealed the extent of the lesion (Figure 2). Recovery post-operatively was uneventful. He last visited the ENT clinic in May 2002. He had completely recovered with no difficulty swallowing or speaking. An MRI of his neck revealed the lesion to be completely excised.

Discussion

Osteocartilaginous choristomas form at any age. Reported cases occur from infancy to 73 years of age.^{3,5} Most reported cases have occurred in women aged between 20 and 40 years.³

Ear, nose and throat choristomas are rare but pharyngeal choristomas are even rarer. Pharyngeal choristomas run the risk of airway obstruction. The symptoms depend on their size and position. They may cause cyanosis, dyspnoea, choking, gagging and dysphagia,⁵ and pain.³ The first presentation may be severe dysphagia and respiratory distress in the newborn.^{5,6} They may be tied to normal tissue by a trunk or a stem. If the pedicle is long, it may be mobile enough to cause intermittent symptoms. Partial obstruction can cause excessive snoring. There are no reported cases of malignant transformation.

Choristomas can occur throughout the oropharynx, nasopharynx and hypopharynx. Of cases reported in the

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FIG. 1

Histological findings: an osteocartilaginous choristoma.

literature, osteocartilaginous choristomas have occurred in the mouth or the oropharynx.^{3,7} Choristomas in the nasopharynx have consisted of skin and associated structures⁵ and also fibrous tissue,⁶ or salivary gland tissue and hyaline cartilage in what were thought to be persistent adenoids in a 21-year-old man.⁷ There are three other cases reported. The first, was a choristoma occurring in a newborn in the oropharynx and the mandible which consisted of fat, skin tissue and associated structures. The second occurred in a three-month-old child extending from the soft palate to the epiglottis and consisted of skin and associated structures, fat and cartilage.8



FIG. 2 MRI scan of osteocartilaginous choristoma on the posterior wall of the pharynx.

The third case occurred in a newborn. A tumour originating from the left lateral pharyngeal wall was excised and the pathological diagnosis was 'choristoma of the nasopharynx'. The authors state that this was probably a hairy nasal polyp.⁹

Of cases in the literature, the long-term survival is only reported for two. A child born with a nasopharyngeal choristoma (skin tissue) was reported as being free of recurrence at five years.⁶ A 47-year-old man with a choristoma of labial gingiva (hyaline cartilage and metaplastic bone) was reported as having no recurrence at two-year follow up.²

Hawkins and Park report a case of a tumour in the nasopharynx, oropharynx and hypopharynx containing cartilage, bone and fat as well as squamous epithelium and ectodermal pancreatic tissue and respiratory epithelium (tissue from all three germ cell layers). Histologically, this was a teratoma not a choristoma.¹⁰

The origin of choristomas is unknown. One theory is that they arise from totipotential cells of one or more germ cell layers which have escaped the normal physiological controlling influences that are responsible for ordered growth and development. Other theories suggest that it may be a developmental abnormality as a result of notochordal influence on the mesoderm or an anomalous rest of parachordal mesoderm surrounding the notochord. Maternal viraemia is also postulated.¹ This is unlikely to be true in our case as the choristoma occurred in a 59-year-old man.

Clinical assessment of nasopharyngeal choristomas must involve the exclusion of encephaloceles. They need an MRI scan. It will also determine the extent of the lesion. Treatment is excision. Recurrence is unlikely if they are completely excised.5

References

- 1 Johnson JF. Oropharyngeal choristoma in a newborn. BrJRadiol 1980;53:1007-9
- Underwood JCE. Tumours benign and malignant. In Underwood JCE, ed. *General and Systemic Pathology*. Edinburgh: Churchill Livingstone, 1995;235
- Tohill MJ, Green JG, Cohen DM. Intraoral osseous and cartilaginous choristomas: Report of three cases and review of the literature. Oral Surg Oral Med. Oral Pathol. 1987;63:506-510
- 4 Seckin T, Unal T, Bir Y. Osteocartilagenous choristoma of the gingiva. Dent Update 1996;23:248-9
- Tahara T, Imate Y. Choristoma of the nasopharynx. ORL J Otorhinolaryngol Relat Spec 1994;56:299-301
- Sarin YK, Zaffar M, Sharma AK. Nasopharyngeal choris-6 toma. Indian Paediatrics 1993;30:827-8
- Malis DJ, Breisch EA, Billman GF. Cartilagenous choris-
- toma of the nasopharynx. *Clin Anat* 2000;**13**:263–6 Downs BW, Shores CG, Drake AF. Choristoma of the nasopharynx. *Otolaryngol Head Neck Surg* 2000;**123**:523 Block EG, Kirkh WM, Warma M, Clin Statistics 9
- Black FO, Kirsh WM. Weaver M. Choristoma of the pharynx in a newborn. Laryngoscope 1969;79:2068-71
- Hawkins DB, Park R. Teratoma of the pharynx and neck. Ann Otol Rhinol Laryngol 1972;81:848-53

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