CASE REPORT

Vascular hamartoma of the parotid gland

Yasser Al Omran,¹ Sara Mathew George,² Shahrzad Zonoozi,¹
Mohammed-kamal Al-Omran²

¹Barts and The London School of Medicine and Dentistry, London, UK
²Salmaniya Medical Center, Kingdom of Bahrain

Correspondence to

Yasser Al Omran,
y.alomran@smd10.qmul.ac.uk

Accepted 9 April 2014

SUMMARY

Hamartomas of the parotid gland are highly unusual. We present a case of a vascular hamartoma of the parotid gland arising as a 1.5 cm mass in a 5-year-old girl. As far as we are aware, no other case with the same features has been reported. We present this case to generate greater consideration of hamartomas as part of the differential diagnosis of parotid lesions, and to reflect over the criteria used to identify a hamartoma from other parotid lesions, as there appears to be some confusion in the literature.

BACKGROUND

Albrecht introduced the term 'hamartoma' in 1904.¹ He used it to describe an inborn error of tissue development that is characterised by an atypical mixture of tissues indigenous to a particular region and tissue(s) foreign to that region. Willis further refined the term in 1967.² He described a hamartoma as a non-neoplastic developmental malformation composed of an admixture of mature cells and tissues indigenous to the anatomic area of occurrence, often with one element predominating.² Unlike neoplasms, which are autonomous new growths, hamartomas are self-limiting; they develop until maturity of the tissue is attained and do not exhibit constant unhindered growth.³

Hamartomas may appear anywhere in the body, but they have a predilection to the kidney, liver and lungs. Their localisation to head and neck regions are seldom, and few reports of such lesions can be seen in the literature. Parotid gland hamartomas are particularly rare, as only three definitive cases have been previously reported in the English-language literature.^{2 4 5} However, none of these cases present with the same features as that in this report. We present an unusual case of a vascular hamartoma of the parotid gland, which, to the best of our knowledge, is the first report to describe a case of parotid hamartoma with these specific histological features.

CASE PRESENTATION

A 5-year-old girl presented with a 2-month history of an asymptomatic mass in the area of the right parotid gland. Physical examination of the neck unveiled a fullness of the tail of the right parotid gland, anterior to the mastoid process. The patient was otherwise healthy. Hearing and facial nerve function were normal and there was no evidence of cervical lymphadenopathy. Contrast-enhanced CT of the neck revealed a well-defined ovoid mass within the superficial portion of the parotid gland; the rest of the parotid gland appeared to be normal

(figure 1). Results of laboratory tests, including bleeding and clotting time, complete blood count, urinalysis and serological tests were normal.

Under general anaesthesia, an oblique incision was made in the upper neck, behind the angle of the mandible. The skin flap was elevated anteriorly and posteriorly to expose the tumour and the parotid gland. The tumour was adherent to the surface of the superficial lobe of the parotid gland; the facial nerve was not exposed. The tumour was enucleated and excised completely.

On macroscopic examination, the excised mass from the parotid gland was well-circumscribed, red in colour and 1.5 cm in diameter. Microscopic examination revealed an unencapsulated mass of nodules of connective tissue, composed predominantly of numerous blood vessels of varying calibre ranging from capillaries to medium-sized blood vessels (figure 2A). Normal salivary gland lobules composed of salivary gland acini and ducts were seen in the vicinity of the blood vessels (figure 2B). A single layer of endothelial cells surrounded the blood vessels (figure 2C), and these stained positive with CD31, an immunohistochemical marker for blood vessels (figure 2D). The overall histopathological findings were diagnostic of vascular hamartoma.

Figure 1 Axial CT with contrast that shows a well-defined lesion (arrow) in the area of the right parotid gland.

CrossMark

To cite: Al Omran Y, George SM, Zonoozi S, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-203615

Figure 2 (A) Low-magnification view showing blood vessels of different sizes (H&E $\times 100$ magnification). (B) Low-magnification view of the lesion in the parotid gland showing a thick-walled vessel and salivary gland tissue in the vicinity (H&E $\times 100$ magnification). (C) Higher magnification view showing endothelial cells lining the lumen of the blood vessels (H&E $\times 200$ magnification). (D) Low-power view showing CD31 positive staining of endothelial cells lining the lumen of the blood vessels (H&E $\times 100$ magnification).

OUTCOME AND FOLLOW-UP

The patient made a smooth recovery, and there was no evidence of recurrence at her 6-month follow-up appointment.

DISCUSSION

The term 'hamartoma' may be applied to tumour-like lesions that arise during development of an organ or tissue. They are composed of disorganised, differentiated and mature tissues indigenous to a particular bodily region and although they may resemble neoplasms, in most cases they show no tendency to transform into one.⁶ They can present in any part of the body, but their preponderance to the head and neck region is rare.⁷ Thus, if they do form in these areas, there is a great potential for misdiagnosis. For example, Ehrich appeared to incorrectly report a hamartoma as a teratoid of the mouth.⁸ Hamartomas of the parotid gland are more unusual; as, to the best of our knowledge, there are only three other definitive reported cases of parotid gland hamartomas in the English-language literature.^{2 4 5} In all of these cases, including ours, the lesion presented as an asymptomatic mass in the parotid gland. Additionally, parotid gland hamartomas do not seem to have a gender bias and seem to present at any age with patients presenting at 4, 5, 16 and 70 years of age.^{2 4 5} However, due to a lack of reports, a general background is difficult to establish. As a result, parotid hamartomas are subject to misdiagnosis and require greater attention so that this may be avoided.

The parotid gland is the most common location for salivary gland neoplasms (73%), of which 80% are benign.^{9 10} Therefore, consideration of the features of a parotid hamartoma and its difference to other neoplasms might help with their differentiation and help explain unusual pathological findings during frozen section analysis. This is important, especially since the differential diagnosis may include malignant neoplasms of the parotid gland. However, differentiation from other neoplasms may be problematic as the features of parotid hamartoma in the current and the previous three reported cases differ in nature. In two reports the parotid hamartoma was reported to be encapsulated,^{2 4} while in this case and in another,⁵ the parotid hamartoma was unencapsulated. In fact, the authors described hamartomas as 'an unencapsulated admixture', and

that this may separate them from other parotid neoplasms, such as oncocytomas (proliferation of oncocytic cells), which are encapsulated.⁵ However, as this is not a universal feature of parotid hamartomas, this may be inaccurate.

Additionally, the definition proposed by Willis may not be specific enough to differentiate a hamartoma from other parotid tumours and thus may also be problematic. Maiorano *et al*¹¹ reported on the clinicopathological features of a distinct parotid tumour in a newborn, showing normal glandular structures admixed with mature lipocytes and blood vessels in variable proportions, and identified the pathology as a sialoangiolipoma. However, as it was composed of normal-looking acinar and ductal structures, interposed with mature lipocytes and blood vessels, it has indigenous tissues admixed with mature tissues and thus it fits into the Willis definition of a hamartoma.² Nevertheless, it was identified as another tumour-like malformation. Moreover, although the authors of one of the previous definitive reported cases of hamartoma made the diagnosis of hamartoma of the parotid gland, they could not determine with certainty whether the parotid lesion was hyperplastic or hamartomatous; suggesting a possible misdiagnosis.⁴ Therefore, the current definition of a hamartoma may present with problems in diagnosis; and although the archetypical pattern of disorganised 'normal' parotid tissue may be suggestive of hamartoma in this case, it may not be in other situations.

Conversely, care should be undertaken when investigating for 'normal' tissues in the diagnosis of hamartoma, especially on fine-needle aspiration. For example, a non-diagnosis or a false-positive diagnosis may be concluded based on the discovery of 'normal' salivary gland characteristics within the parotid tissue, as had been reported in a previous case report.⁴ Clinicians and pathologists should consider this to prevent further morbidity of the patient for want of a better diagnosis.

Fortunately, the clinical management of parotid hamartomatous lesions generally parallels that of other benign parotid masses, that is, a simple enucleation or a conservative excision. However, although parotid hamartomas are rare, there is a need for greater awareness of hamartomas in order to prevent the likelihood of overly aggressive procedures in the fear of

neoplastic lesions or to prevent lack of action in the case of non-diagnosis. Therefore, parotid hamartomas should be encompassed within the umbrella of differential diagnoses for parotid masses.

Learning points

- ▶ Hamartomas of the parotid gland are incredibly rare.
- ▶ The current criteria used to diagnose hamartomas of the parotid may not be specific enough to form a diagnosis of a hamartoma.
- ▶ Clinical management of parotid hamartomatous lesions generally parallels that of other benign parotid masses.

Contributors YAO carried out the writing of the manuscript, literature search and approval of the images used in the case report. SZ edited and revised the manuscript. SMG reviewed and edited elements relating to pathology, and was involved in the selection of the images used in the case report. M-KA-O supervised, commented and helped in the revision of the manuscript, and was responsible for acquiring informed consent from the patient's parents. All authors have read and approved the final manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Albrecht P. Verhandlungen der Deutschen Pathologischen Gesellschaft. Willis RA. ed *Pathology of tumours*. 4th edn. London: Bunerworths, 1967:7.
- 2 Samuel J, Fernandes CC. Hamartomas of the head and neck. A report of 4 cases. *S Afr Med J* 1985;68:265–7.
- 3 Wenig BM, Heffner DK. Respiratory epithelial adenomatoid hamartomas of the sinonasal tract and nasopharynx: a clinicopathologic study of 31 cases. *Ann Otol Rhinol Laryngol* 1995;104:639–45.
- 4 Tsuda H, Morinaga S, Mukai K, *et al*. Hamartoma of the parotid gland: a case report with immunohistochemical and electron microscopic study. *Virchows Arch A Pathol Anat Histopathol* 1987;411:473–8.
- 5 Nicolau Y, Faquin WC, Deschler DG. Hamartoma of the parotid gland: report of a unique case. *Ear Nose Throat J* 2010;89:E8–10.
- 6 Gardner DG. The concept of hamartomas: its relevance to the pathogenesis of odontogenic lesions. *Oral Surg Oral Med Oral Pathol* 1978;45:884–6.
- 7 Zarbo RJ, McClatchey KD. Nasopharyngeal hamartoma: report of a case and review of the literature. *Laryngoscope* 1983;93:494–7.
- 8 Ehrlich WF. Teratoid of the mouth. *Am J Orthod* 1945;31:650–9.
- 9 Eveson JW, Cawson RA. Salivary gland tumours. A review of 2410 cases with particular reference to histological types, site, age and sex distribution. *J Pathol* 1985;146:51–8.
- 10 Eneroth CM. Histological and clinical aspects of parotid tumours. *Acta Otolaryngol Suppl* 1964;188(Suppl 191):1–99.
- 11 Maiorano E, Capodiferro S, Fanelli B, *et al*. Hamartomatous angiolipoma of the parotid gland (sialoangiolipoma). *Head Neck Pathol* 2008;2:36–40.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow