CASE REPORT

Parotid epithelial–myoepithelial carcinoma: an unexpected intraoperative finding

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SUMMARY

We present a case of a 72-year-old man with a parotid mass which was initially diagnosed as a pleomorphic adenoma. Intraoperatively, the tumour was found to be invading the facial nerve. It was later found to be an epithelial–myoepithelial carcinoma, a rare salivary gland tumour. Despite invasion of the facial nerve, function of the nerve remained intact.

BACKGROUND

Epithelial–myoepithelial carcinoma (EMC) is a rare malignant salivary gland tumour. We present a case of a 72-year-old man with a parotid mass which was initially diagnosed as a pleomorphic adenoma. Intraoperatively, there was macroscopic perineural involvement of the facial nerve by the tumour. Histopathological evaluation of the specimen revealed it to be EMC, a rare salivary gland tumour. Despite invasion of the facial nerve, function of the nerve remained intact, preoperatively as well as postoperatively.

CASE PRESENTATION

A 72-year-old man presented with a long-standing left parotid mass (3×4 cm), with intact facial nerve function (House-Brackmann Grade I). A preoperative fine needle aspirate (FNA) performed on this mass favoured the diagnosis of a benign pleomorphic adenoma.

INVESTIGATIONS

Contrast-enhanced CT showed a heterogenous mass (25×16×30 mm) in the superficial left parotid gland with multiple small cystic areas (figure 1).

DIFFERENTIAL DIAGNOSIS

Preoperative FNA favoured the diagnosis of a benign pleomorphic adenoma, but a malignant lesion such as an adenoid cystic carcinoma could not be ruled out.

Other differentials include benign lesions such as a Warthin's tumour, monomorphic adenoma, benign intraparotid lymph node or malignant lesions such as an adenocarcinoma or squamous cell carcinoma.

TREATMENT

The patient underwent a left superficial parotidectomy. The facial nerve appeared thickened and splayed from the pes anserinus to the stylomastoid foramen. There was a good plane between the parotid tissue and the facial nerve. In view of the possible benign nature of the tumour from FNA

and the intact preoperative facial nerve function, the facial nerve was left intact and not biopsied (figure 2).

Final pathology confirmed an EMC of parotid gland with high-grade transformation (figure 3). There were also features of lymphovascular and perineural invasion. Postoperative MRI suggested perineural extension of the tumour at the level of the distal mastoid segment (figure 4). A subsequent positron emission tomography scan showed no evidence of distant metastatic disease.

OUTCOME AND FOLLOW-UP

His case was discussed in the head and neck multidisciplinary team meeting, and adjuvant radiotherapy was offered. He subsequently developed a complete left-sided facial nerve palsy toward the end of his radiotherapy treatment. At 12 months, surveillance imaging confirmed no evidence of local or systemic disease, and he was offered facial reanimation surgery. He remains disease free at 18 months from surgery.

Cytological re-evaluation was conducted on the specimen obtained from the preoperative FNA. As there were only epithelial cells isolated in the specimen, the cytologist stated that the favoured differentials were still that of either a pleomorphic adenoma or adenoid cystic carcinoma.

DISCUSSION

Parotid gland masses are generally classified as either benign or malignant. They account for

Figure 1 Preoperative CT of the temporal bone. Axial view demonstrating cystic nature of left parotid gland.

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Figure 2 Intraoperative photographs. Abnormal facial nerve tracked towards stylomastoid foramen.

approximately 80% of all salivary gland neoplasms, of which 75–80% are benign.¹ The most common benign parotid masses are pleomorphic adenomas or Warthin's tumours.² Other less common benign parotid masses include monomorphic adenomas or benign intraparotid lymph nodes.² The most common malignant tumours of the parotid gland are adenocarcinoma, squamous cell carcinoma, mucoepidermoid carcinoma, lymphoma and adenoid cystic carcinoma.² Less commonly, malignant tumours such as acinic cell carcinoma, synovial sarcoma and EMCs are isolated.²

The assessment of a parotid mass involves careful history taking, physical examination (including the assessment of facial nerve function), and other investigations such as radiological imaging or FNA of the parotid mass. The role of cytological evaluation of specimens obtained via FNA in patients suspected of having malignant parotid masses is however, debateable. This is due to the relatively high false-negative rates (40%) and low sensitivity (64%) of this test.³ The test is however, still largely viewed as a safe and cost-effective adjunct in the preoperative evaluation and may assist the clinician in forming a treatment plan for parotid masses.² It has relatively high positive

Figure 3 Microscopic observation of H&E-stained specimen displaying low-grade and high-grade epithelial–myoepithelial carcinoma of the parotid with perineural invasion.

Figure 4 Postoperative MRI. Coronal view demonstrating enhancement of the distal mastoid segment of the facial nerve.

predictive value (83%), negative predictive value (87%), accuracy (86%) and a very high specificity (95%) in confirming suspicion of malignancy.⁴ This is significant as it allows a surgical team to not only plan the surgical resection, but it may also completely alter the course of management of the patient should the FNA reveal alternative diagnoses such as lymphoma.² It may also assist the surgeon in terms of offering the patient the option of avoiding surgery-specific situations such as where the FNA has revealed a benign diagnosis (and as such is a more reliable test in this setting), or if a patient is a high-risk surgical candidate due to their poor general health, or in the setting of disseminated malignancy.²

Cystic parotid gland lesions such as that which was present in this patient are often difficult to accurately diagnose prior to surgery.⁵ The differentials may include benign or malignant tumours of the parotid (discussed above) which have undergone cystic degeneration (most commonly Warthin's tumour), infected/necrotic lymph nodes, sialoceles or lymphoepithelial cysts.⁵ Careful evaluation of the cystic components through the use of MRI in conjunction with cytological information from FNA may be useful in differentiating benign and malignant cystic parotid masses.⁵

EMC is a malignant salivary gland tumour which accounts for approximately 1% of all salivary gland tumours⁶ and has been presumed to be of intercalated duct origin.⁷ It is usually a low-grade tumour which predominantly affects the parotid gland but can often affect other seromucous gland sites such as the respiratory tract, lacrimal gland and breasts.⁸ The cellular architecture can vary between tubular, papillary, cystic or solid patterns.⁸ EMCs often display a high rate of recurrence (35–40%) but a low risk of metastasis and mortality, unless it displays high-grade transformation or dedifferentiation (DEMC), which in both instances is rare.^{8,9}

Patients presenting with high-grade EMC are often old with an average age of 75.9 years.⁹ The tumours are invariably more aggressive than typical EMC with an average size of 6.3 cm.⁹ Fifty-five per cent display extraglandular extension and patients are also more likely to have metastases to lymph nodes (50%) and distant sites (30%).⁹

Treatment often comprises of primary surgical resection of the tumour.¹⁰ High-grade EMC usually displays a good response to radiotherapy and chemotherapy. Adjuvant

radiotherapy is often required in cases of large primary tumours (>4 cm) and positive surgical margins.¹⁰ The role of chemotherapy in the treatment of malignant tumours of the salivary gland has been largely reserved for palliation in advanced disease or in cases of DEMC.¹⁰ There has however, been recent evidence emerging on the role of chemotherapy in the treatment of base of tongue EMC.¹¹

EMC usually takes a chronic course. High-grade transformation and DEMC usually confers the patient a poorer prognosis.¹⁰ Long-term follow-up for these patients is warranted as metastases can even occur much later in life.¹⁰

Learning points

- ▶ Epithelial–myoepithelial carcinoma (EMC) is a rare malignant salivary gland tumour.
- ▶ It is usually a low-grade tumour which predominantly affects the parotid gland.
- ▶ EMC usually displays a good response to radiotherapy and chemotherapy.
- ▶ High-grade transformation and dedifferentiation usually confers the patient a poorer prognosis.
- ▶ In the setting of EMC with perineural invasion, but intact facial nerve function, our case demonstrated good local control with facial nerve preserving parotidectomy and adjuvant radiotherapy.

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Competing interests None.

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REFERENCES

- 1 Speight PM, Barrett AW. Salivary gland tumours. *Oral Dis* 2002;8:229–40.
- 2 Bussu F, Parrilla C, Rizzo D, *et al.* Clinical approach and treatment of benign and malignant parotid masses, personal experience. *Acta Otorhinolaryngol Ital* 2011;31:135–43.
- 3 Sergi B, Contucci A, Corina L, *et al.* Value of fine-needle aspiration cytology of parotid gland masses. *Laryngoscope* 2004;114:789.
- 4 Zbären P, Schär C, Hotz MA, *et al.* Value of fine-needle aspiration cytology of parotid gland masses. *Laryngoscope* 2001;111:1989–92.
- 5 Kikuchi M, Shinohara S, Fujiwara K, *et al.* [Cystic parotid gland lesion evaluation]. *Nihon Jibiinkoka Gakkai Kaiho* 2010;113:441–9.
- 6 Palmer RM. Epithelial-myoeplithelial carcinoma: an immunocytochemical study. *Oral Surg Oral Med Oral Pathol* 1985;59:511–15.
- 7 Corio RL, Sciubba JJ, Brannon RB, *et al.* Epithelial-myoeplithelial carcinoma of intercalated duct origin: a clinicopathologic and ultrastructural assessment of sixteen cases. *Oral Surg Oral Med Oral Pathol* 1982;53:280–7.
- 8 Seethala R, Barnes EL, Hunt J. Epithelial-myoeplithelial carcinoma: a review of the clinicopathologic spectrum and immunophenotypic characteristics in 61 tumors of the salivary glands and upper aerodigestive tract. *Am J Surg Pathol* 2007;31:44–57.
- 9 Roy P, Bullock M, Perez-Ordoñez B, *et al.* Epithelial-myoeplithelial carcinoma with high grade transformation. *Am J Surg Pathol* 2010;34:1258–65.
- 10 Yamazaki H, Ota Y, Aoki T, *et al.* Lung metastases of epithelial-myoeplithelial carcinoma of the parotid gland successfully treated with chemotherapy: a case report. *J Oral Maxillofac Surg* 2013;71:220–6.
- 11 Peters P, Repanos C, Earnshaw J, *et al.* Epithelial-myoeplithelial carcinoma of the tongue base: a case for the case-report and review of the literature. *Head Neck Oncol* 2010;2:4.

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