CASE REPORT

Myoepithelial carcinoma of the parotid: a rare tumour that may provide diagnostic difficulty

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SUMMARY

This report presents the case of a 57-year-old man with a 6-week history of a slowly enlarging lump in the right parotid gland. Initial ultrasound investigation confirmed a 3 cm solid mass located within the superficial lobe of the right parotid gland. Sonographically, the mass demonstrated possible malignant features with internal heterogeneity of architecture and some loss of marginal clarity. Ultrasound-guided core biopsy (USCB) of the mass reported a mixed neoplasm with epithelial and myoepithelial appearances consistent with likely benign adenoma or myoepithelioma. A right superficial parotidectomy was subsequently performed. Histological and immunohistochemical analysis of the excised specimen showed a myoepithelial carcinoma. Postoperative CT and MR staging scans did not show evidence of metastases and no further treatment was given following discussion of the case at the regional speciality multidisciplinary meeting. This case illustrates the clinical and pathological features of this rare salivary gland tumour, but also discusses the diagnostic difficulties that may be encountered preoperatively.

BACKGROUND

Myoepithelial carcinoma, or malignant myoepithelioma, of the salivary glands is considered a rare entity. Only relatively recently described in the literature, many clinicians and pathologists are not familiar with the histopathological features, immunohistochemical profile, and clinical behaviour of this tumour. Diagnostic difficulty with fine-needle aspiration cytology (FNAC) is recognised, however ultrasound-guided core biopsy (USCB) of the parotid gland is a relatively recently reported technique, with few reports of false-negative diagnosis.¹

As a result of their diverse range of cytological features, myoepithelial carcinoma can be overlooked as a diagnosis, with many neoplasms broadly characterised as 'mixed malignant tumours', however, improving awareness of this lesion and the variations in its cytological and immunohistochemical profile, may lead to greater diagnostic recognition in the future. This report highlights the importance of triple assessment in the diagnosis of parotid masses, and the difficulties that may be faced in diagnosing this rare, but increasingly recognised tumour.

identified. He was otherwise fit and well with no previous medical history of note. He was taking no regular medication and was a non-smoker with minimal alcohol consumption.

INVESTIGATIONS

The patient underwent initial ultrasound of the gland, which demonstrated a 3 cm, mixed texture mass in the superficial lobe of his right parotid gland in a preauricular region (figure 1). Although this lesion had no definitive malignant characteristics, the margins appeared ill-defined in part, and the internal architecture of the lesion was heterogeneous. He proceeded to USCB for histological diagnosis of this lesion, on ultrasound criteria, consistent with a likely tumour and with some suspicious clinical and sonographic features.

Histological examination of the core biopsy reported a neoplasm with areas containing cells with both epithelial/myoepithelial appearances. There was no nuclear pleomorphism or mitotic activity seen to suggest a malignant process, and a differential diagnosis of basal cell adenoma and benign myoepithelioma was suggested (figure 2). No immunohistochemistry was carried out on the initial core biopsy.

Following discussion of the case at the regional multidisciplinary meeting, the patient proceeded to superficial parotidectomy. Histological analysis of the excised specimen showed a more myoepithelial appearance than in the initial core biopsy with no significant nuclear pleomorphism but there were areas with numbers of mitotic figures (figure 3A). More importantly the excision specimen revealed both capsular (figure 3B) and perineural invasion (figure 4) at the edge of the lesion, though excision was complete and surgical margins were clear.

Figure 1 Ultrasound image of the mass.

CASE PRESENTATION

A 57-year-old man presented with a 6-week history of a firm, non-fixed, slowly enlarging mass in his right parotid gland. His head and neck examination was otherwise unremarkable, with facial nerve function intact and no palpable cervical nodes

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Figure 2 Core biopsy histology. Note the relative lack of nuclear pleomorphism, atypia or other features of malignancy.

Immunohistochemistry showed widespread strong expression (including in the invasive areas) of the myoepithelium associated markers smooth muscle actin, p63, vimentin, CD10 and myosin as well as CK14 and S100 protein (figures 5A, B and 6A, B). There was also expression of epithelial markers CK7 and broad spectrum cytokeratins (MNF116, CAM5.2 and AE1/AE3). Epithelial membrane antigen was generally negative. Following the criteria described by Barnes *et al*² for the classification of salivary tumours the combination of immunohistochemical evidence of myoepithelial differentiation along with unequivocal evidence of capsular and perineural invasion led to a final histological diagnosis of well-differentiated myoepithelial carcinoma (malignant myoepithelioma).

DIFFERENTIAL DIAGNOSIS

- ▶ Benign myoepithelioma
- ▶ Basal cell adenocarcinoma
- ▶ Atypical pleomorphic adenoma, oncocytoma, basal cell adenoma
- ▶ Cystadenolymphoma (Warthin's tumour)
- ▶ Primary/secondary parotid malignancy

TREATMENT

The case was discussed in a postoperative multidisciplinary meeting. Postoperative MR neck and CT thorax scans revealed

no evidence of any regional nodal or distant metastatic disease. The lesion was confirmed as widely excised with clear surgical margins, and the case was presented again at the regional head/neck cancer meeting. Treatment options for this high-grade malignancy include complete parotidectomy, radiotherapy or regular surveillance. The patient declined further surgery, opting for close MRI surveillance. There was no indication for radiotherapy in view of the wide excision margins.

OUTCOME AND FOLLOW-UP

There were no postoperative issues, and the patient recovered with no complications. He remains well on surveillance for 12 months to date.

DISCUSSION

Originally described in 1975, myoepithelial carcinoma (malignant myoepithelioma) is a rare soft tissue malignancy that has been reported in multiple sites including the breast, skin and lung. Less commonly, it presents as a head and neck tumour, the most common site being the parotid gland.³ Of all salivary gland neoplasms, it has a reported incidence of 0.2%, with those affected usually aged between 40–60 years old.⁴ Current knowledge of myoepithelial carcinoma is limited, and with less than 50 cases on record, there is little available literature on the behaviour or prognosis of this entity.⁵

It is generally considered that in most cases, the tumour arises *de novo*, however, malignant transformation within a recurrent pleomorphic adenoma or benign myoepithelioma is also reported. In general, myoepithelial carcinomas are regarded as a high-grade malignancy, with a poor prognosis. There may be extensive local growth and infiltration of the surrounding tissues observed. There is a high risk of recurrence, and distant metastasis to the lungs, liver and bone is thought to be common, however local metastasis to cervical nodes is infrequent. Some well-differentiated examples of the lesion may be slow-growing, and therefore less aggressive, although it is generally considered that any neoplasm containing myoepithelial cells should always be regarded as potentially high grade.⁶

Surgery is the first choice of treatment for myoepithelial carcinoma, with the requirement for good clearance due to high rates of recurrence. Radiotherapy is considered where primary surgical excision margins are close or incomplete. It is

Figure 3 (A) H&E ×400. Excision biopsy showing fairly uniform cells with myoepithelial appearance showing mitotic activity (Arrows) (B) H&E ×200. Excision biopsy showing islands of tumour cells (Arrows) invading fat at the edge of the lesion.

Figure 4 H&E $\times 200$. Excision biopsy showing perineural invasion (Arrows) outside the capsular edge of the lesion.

recognised that the diagnosis of myoepithelial carcinoma with FNAC can be challenging because, as with all tumours containing a myoepithelial component, there may be a diverse range of

cytological features, some of which may not be overtly malignant. These may include spindle, plasmacytoid, epithelioid and clear cell types.⁷ These features of myoepithelial differentiation are seen within both myoepithelial carcinoma, and benign myoepithelioma. If the lesion arises within a benign pleomorphic adenoma, the additional cytological features can add to the diagnostic difficulty.⁸

Recent publications have demonstrated high diagnostic accuracy of USCB in parotid diagnosis, although examples of false-negative USCB are not well described. USCB has the potential advantage over FNAC in that it provides a core of tissue that can be used for immunohistochemical analysis, allowing improved typing and grading of tumours.⁹ Common immunohistochemical markers found in myoepithelial carcinoma include cytokeratins and vimentin (figure 5B), although over time, the neoplastic transformation of myoepithelial cells can result in changes to their smooth muscle phenotype, and there may be variable positivity for S100 (figure 5A), calponin, actins, p63 protein, smooth muscle myosin, glial fibrillary acidic protein and CD10.¹⁰ However, one major disadvantage of both FNA and core biopsy techniques is that the sample obtained

Figure 5 (A) Immunostain for actin $\times 400$ on excision biopsy. Showing strong actin positivity in the neoplastic cells. (B) Immunostain for myosin $\times 400$ on excision biopsy. Showing strong myosin positivity in the neoplastic cells.

Figure 6 (A) Immunostain for S100 protein $\times 400$ on excision biopsy. Showing strong positivity in neoplastic cells invading fat at the edge of the tumour. (B) Immunostain for CD10 $\times 400$ on excision biopsy. Showing strong positivity in neoplastic cells invading fat at the edge of the tumour.

may not contain areas with all cytological features of the mass, including perhaps the most important feature for diagnosis of malignancy; the capsular margin of the tumour.

In this case study, the crucial diagnostic feature to indicate that this was a myoepithelial carcinoma rather than a benign myoepithelioma was capsular and perineural invasion at the margin of the lesion. This was only identifiable on histology of the excised specimen postparotidectomy. Poorly-differentiated salivary gland malignancies are usually easily recognised as malignant on histology of core biopsy as they show necrosis, nuclear pleomorphism and mitotic activity. However, in well-differentiated salivary gland malignancies (including myoepithelial carcinoma and basal cell adenocarcinoma) which lack such nuclear pleomorphism and necrosis, the ability to demonstrate evidence of infiltrative growth such as capsular or perineural invasion is essential for identifying the malignant nature of the neoplasm.²

Learning points/

- ▶ Myoepithelial carcinoma is an unusual malignant salivary gland tumour, but increased awareness of the neoplasm can lead to a rapid, more accurate diagnosis.
- ▶ Ultrasound-guided core biopsy is favoured over fine-needle aspiration cytology to allow for more accurate immunohistochemical analysis to be undertaken in addition to cytological assessment.
- ▶ Preoperative diagnosis of myoepithelial carcinoma may be difficult, as certain cytological features of the tumour are seen in other pathologies.
- ▶ Some cases of myoepithelial carcinoma may only be diagnosed postoperatively when analysis of the full-excised specimen can be undertaken.
- ▶ A multidisciplinary approach involving triple assessment is essential in the investigation and management of parotid tumours.

If the capsule/margin of the lesion appears poorly defined on initial ultrasound examination, USCB can potentially be targeted to sample the capsular area. This may improve diagnostic yield, however, it must be remembered that this method will not reliably demonstrate capsular infiltration, which, being very focal, is usually only apparent on the full-excised specimen.

This case serves to raise awareness of this unusual, but increasingly recognised malignant salivary gland tumour. By recognising the potential difficulties that can arise in making a preoperative diagnosis of myoepithelial carcinoma, the importance of the triple assessment (clinical, radiological and immunohistochemical analysis) is highlighted.

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Competing interests None.

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