

Myoepithelial carcinoma of the ear: A rare case report and review of the literature

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The aim of this study was to present a unique case report of myoepithelial carcinoma arising from the external auditory canal and presenting as a huge tympanomastoid mass along with a review of the literature.

A 52-year-old woman presented with a large periauricular swelling of a 3-year duration with a recent increase in size over the last 3 months along with pain and bleeding from the mass. The patient was evaluated by clinical examination, haematological and biochemical tests, and computed tomographic and MRI scan of the tympanomastoid region. An incision biopsy was performed before definitive management was initiated.

Computed tomographic scan and MRI were suggestive of a large tympanomastoid mass without any intracranial extension. The incisional biopsy report was suggestive of invasive adenocarcinoma. En-bloc excision of the mass including lateral temporal bone resection along with ipsilateral selective (levels I, II and III) neck dissection was performed under general anaesthesia. The resultant defect was reconstructed by a rotational scalp flap. Immunohistochemistry and histopathology of the excised specimen proved the diagnosis of myoepithelial carcinoma. After surgery, the patient was treated with adjuvant radiotherapy. At 1½ years of follow-up, the patient was doing fine, without any recurrence of disease.

Myoepithelial carcinoma of the ear has rarely been reported in the literature. Histopathology along with immunohistochemistry is the mainstay of diagnosing of this unusual lesion. Meticulous planning and proper execution of optimum surgical excision is the primary treatment modality, which should be supplemented with radiation therapy. Long-term disease-free survival, although rare, can be achieved as in the present case report.

Keywords:

immunohistochemistry; lateral temporal bone resection; myoepithelial carcinoma; scalp flap; tympanomastoid mass

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Introduction

Myoepitheliomas are tumours composed almost exclusively of neoplastic cells showing myoepithelial differentiation. Myoepithelial tumours, both benign and malignant, are described mostly in the context of salivary glands, but they also occur in the breast, soft tissue, skin, larynx and sinonasal region. Infratemporal fossa [1] and orbit [2] are other uncommon sites that may be involved by myoepithelial carcinoma. Myoepithelial carcinoma arising from the external auditory canal and mastoid has rarely been reported [3,4].

Materials and methods

This is a case report of a single case presenting with a rare condition in the Department of ENT, Head and Neck Surgery of a tertiary referral hospital in Eastern India.

Results and analysis

K.D., a 52-year-old woman, resident of Orissa, India, attended the Outpatients Department of ENT, R.G.

Kar Medical College, Kolkata, with a history of an irregular left-sided periauricular painless swelling that increased gradually in size over a period of 3 years, with a recent increase in size over the last 3 months along with pain and bleeding from the surface of the mass. On initial workup, the mass was found to be 8 × 7 × 5 cm in size, bony hard in consistency, with a bosselated surface, displacing the left pinna anteriorly and laterally encroaching upon the left mastoid region with overlying skin excoriation (Fig. 1). The mass was fixed to the skin as well as to the underlying structures. Facial nerve function was intact. High-resolution computed tomography (CT) of the tympanomastoid region showed a fairly large (8 × 7.5 × 5.5 cm) mixed attenuating heterogeneously enhancing partly necrotic mass lesion present in the left external ear with partial erosion of squamous temporal bone and mastoid air cells. The lesion was inseparable from the left pinna and extended anteroinferiorly towards the parotid gland along with partial destruction of the left temporomandibular joint. The medial wall of the middle ear and petrous process was uninvolved (Fig. 2).

MRI of the region showed a fairly large hypointense lobulated mass in T1 measuring 7.7 × 7.2 × 5.3 cm in size, with hyperintense foci in T2, arising from the left external ear with involvement of the left pinna, the glenoid fossa and posterior surface of the parotid gland. Middle ear ossicles could not be identified. Inner ear structures including the cochlea and the vestibule were free from the lesion (Fig. 3). Radiological impression was suggestive of a tympanomastoid neoplasm, probably malignant. There was no significant lymphadenopathy in the ipsilateral or the contralateral side of the neck.

Incisional biopsy from the mass showed cells with an epithelioid appearance arranged in trabeculae along with wide areas of necrosis and a diagnosis of adenocarcinoma of the ear was made. As a preoperative metastatic workup, skiagram of the chest and ultrasonography (USG) whole abdomen were advised,

both of which were normal. There was no scope for audiometry in the patient because of the huge mass occupying the auricular region on the affected side; hence, preoperative hearing status of the affected ear could not be assessed.

During surgery, ipsilateral supraomohyoid (levels I, II and III) neck dissection was performed and vascular control of the great vessels of the neck was established. Then, lateral temporal bone resection and total parotidectomy along with excision of the left pinna were performed for en-block resection (Fig. 4). The facial nerve had to be sacrificed for complete removal of disease. The resultant defect over the tympanomastoid region was covered with a transposition scalp flap and split-thickness skin graft (Fig. 5). The patient was referred to the Department of Physical Medicine after the surgery for rehabilitation of

Figure 1



Clinical photograph of the patient.

Figure 2



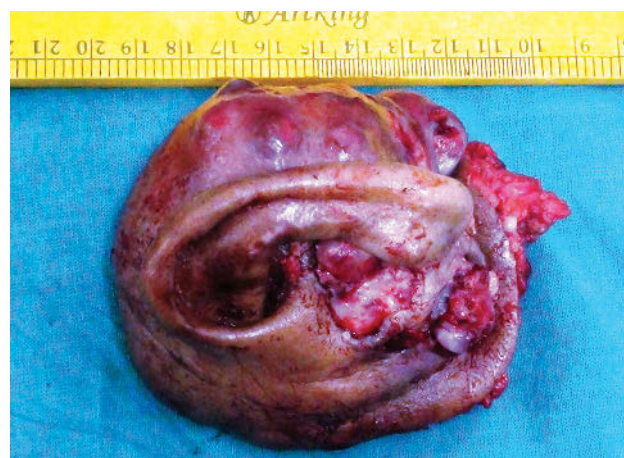
Computed tomographic scan of the lesion showing mixed attenuating heterogeneously enhancing partly necrotic mass lesion present in the left external ear with partial erosion of squamous temporal bone and mastoid air cells.

Figure 3



T2-weighted MRI image showing a tumour with a hyperintense signal with a multilobulated appearance arising from the left external ear region extending anteriorly to the posterior surface of the parotid gland.

Figure 4



Excised specimen.

the facial muscle paralysis arising because of facial nerve sacrifice during the surgery.

Histological report of the excised specimen indicated a tumour mass with a lobulated architecture separated by fibrous bands. The tumour was composed of spindle cells plasmacytoid and epithelioid in shape with a clear cytoplasm in most areas, with focal areas showing eosinophilic cytoplasm. Nuclei showed mild to moderate atypia. Cells were arranged in sheets and trabeculae with focal eosinophilic hyaline stroma. Sections showed many areas of coagulative necrosis. Increased mitotic activity (>7/10 hpf) was noted and there were focal areas of squamous metaplasia with keratinization. The overall histological picture was in favour of myoepithelial carcinoma with squamous metaplasia (Figs 6 and 7). Immunohistochemical analysis showed that tumour cells expressed cytokeratin, P-63, calponin, S-100 protein and glial

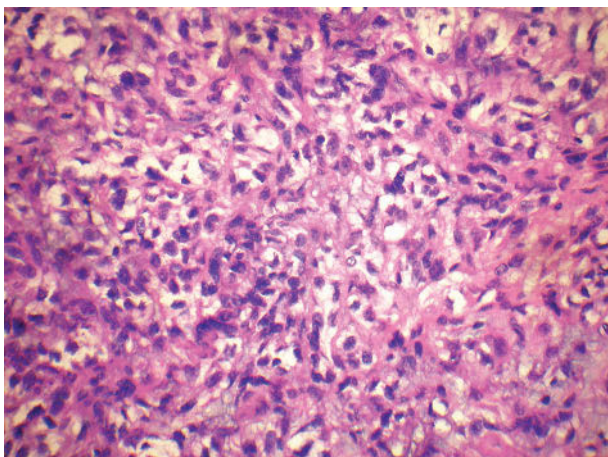
fibrillary acidic protein (GFAP) (Fig. 8), but were immunonegative for epithelial membrane antigen (EMA). Coexpression of cytokeratin along with P-63 in all tumour cells and absence of duct-lining cells expressing only cytokeratin and not P-63/calponin ruled out epithelial–myoepithelial carcinoma (EMC), which has a dual-cell population. The overall histopathological and immunochemistry thereby confirmed the diagnosis of myoepithelial carcinoma. The patient received postoperative external beam radiotherapy and is currently under follow-up.

Figure 5



Postoperative photograph of the patient 2 weeks after surgery.

Figure 7

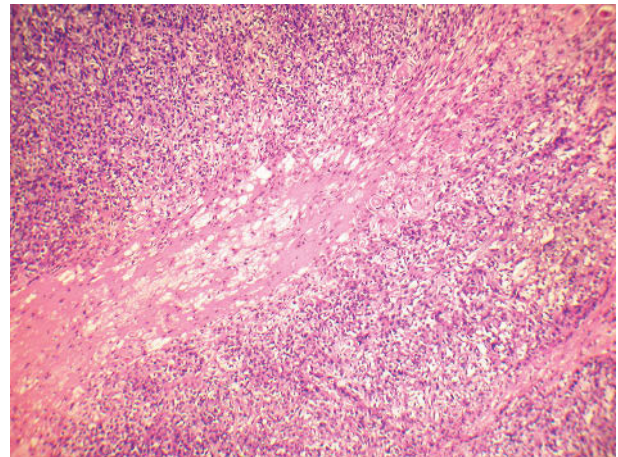


Histopathological image of the excised tumour (H&E stain, x400).

Discussion

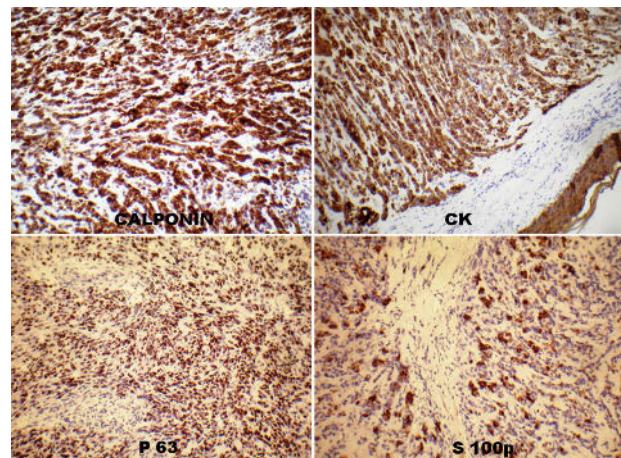
Myoepitheliomas or myoepithelial carcinomas most commonly involve the parotid glands, followed by other major and minor salivary glands, breast, skin and soft

Figure 6



Histopathological image of the excised tumour (H&E stain, x100).

Figure 8



Immunohistochemistry showing positivity for calponin, cytokeratin (CK), P-63 and S-100 protein (S-100p).

tissue. Myoepithelial carcinomas are of intermediate to high grade in nature, with the potential to behave aggressively. They can arise *de novo* or in a pre-existing pleomorphic salivary adenoma [5]. Other rare sites of myoepithelial carcinoma in the head neck region are the paranasal sinuses, nasopharynx, nasal cavity, the base of the tongue and infratemporal fossa [1,2]. Myoepithelial carcinoma arising from the external auditory canal and presenting as a tympanomastoid mass is quite rare in the literature and only five cases of myoepithelial tumours involving the ear have been reported so far.

The first case report of myoepithelioma of the external auditory canal was described by Chen [6]. Lau *et al.* [7] described a case of parotid myoepithelial carcinoma presenting with stenosis of the external ear canal in the absence of a palpable mass. The authors were of the opinion that surgery was the mainstay of treatment, with limited but evolving role of radiotherapy in these tumours. Joseph *et al.* [4] described a patient with malignant myoepithelioma of the mastoid arising from the postauricular soft tissue, but the patient was different from ours as he was young and died from systemic metastasis. Later, external ear myoepithelioma was described by Kong *et al.* [3] and Dirier *et al.* [8]. Middle ear involvement by myoepithelioma was described by Haggisawa *et al.* [9]. However, such a huge myoepithelial carcinoma of the ear involving the external auditory canal, part of the middle ear, mastoid and the parotid has never been reported in the literature before.

Myoepithelial tumours of soft tissue can occur in any age group (range 3–83 years), with equal sex predilection. This tumour can present as a subcutaneous as well as a deep-seated mass. Clinical presentation depends on its location. In this case, the anterolateral displacement of the pinna was most likely because of subdermal growth of the tumour after its exit from the external auditory canal by erosion of its wall. The facial nerve was not involved in this case because most likely it had arisen from the external auditory canal (EAC), subsequently encroaching upon the middle ear. Primary origin from the middle ear would have led to facial nerve palsy before extending to the EAC. A huge myoepithelial carcinoma such as in this case if arising from the parotid would have also involved the facial nerve, with resultant facial nerve palsy at the time of seeking medical advice.

In CT scan, myoepithelial carcinoma appears isodense to muscle and shows moderate homogenous enhancement with contrast. On MRI, it is hypointense on T1 and shows a dense homogenous enhancement. In the present case, the variegated appearance may

have been because of necrosis of the central part of the tumour.

The treatment of choice in myoepithelial carcinoma is complete excision [1]. However, in a case such as this, with involvement of the tympanomastoid bone and parotid, where multiple neurovascular structures pass through the surgical field, previous vascular control is necessary for complete excision of the mass. After excision of the mass, the resultant defect needs to be covered by a microvascular free flap or a myocutaneous flap. Local rotational flap, that is scalp flap was found to be a suitable alternative in this case because of the shorter operating time and reduced morbidity of the patient.

Histologically, myoepithelial carcinomas are distinguished from their benign counterparts by the presence of infiltrative growth, cellular atypia, increased mitotic activity and the presence of coagulative tumour necrosis. Usually, the tumours are unencapsulated, with pushing type of infiltration. The neoplastic cells may be spindle shaped, plasmacytoid, epithelioid or clear cells, arranged in solid sheets, trabeculae or fascicles. Cells may show squamous metaplasia with or without keratinization. The intervening stroma may be myxoid, collagenous or hyaline in appearance. To diagnose malignancy in tumours with exclusively myoepithelial differentiation, a mitotic activity of greater than 7/10 hpf or Ki-67 index of greater than 10% has to be found. In myoepithelial tumours with a relatively innocuous appearance and low mitotic activity, an infiltrative growth pattern remains the only feature to establish its malignant nature.

In the present case, although the overall histology strongly suggested a myoepithelial neoplasm, EMC and mucoepidermoid carcinoma of the ceruminous glands had to be considered in the differential diagnosis. EMC has to be differentiated whenever a histological diagnosis of myoepithelial carcinoma is made for a lesion predominantly showing a clear cell population as EMC is a relatively low-grade malignancy compared with myoepithelial carcinoma. EMC is a biphasic tumour with tubular structures lined by ductal cells surrounded by layers of clear cells. Squamous differentiation may also be observed in this tumour. Dual-cell population is indicated by the absence of myoepithelial markers in the cells lining the tubules, which show positivity for cytokeratin. Distant metastasis is very uncommon in EMC and tumour-associated mortality is low. However, because of the similarity in the histological pictures of the three diseases as well as the diversity of histopathological findings in individual lesions, immunohistochemistry is the investigation of choice to establish the final

diagnosis. Myoepithelial cells are usually positive for P-63, calponin, S-100 protein and CK-14. Cytokeratin, GFAP and actin are also often positive, but not carcino-embryonic antigen (CEA) or HMB-45. The present case showed positivity for cytokeratin, P-63, calponin, S-100 protein and GFAP on immunohistochemistry (IHC). Hornick and Fletcher reviewed 101 cases of myoepithelial tumours by immunohistochemistry; all cases were reactive for epithelial markers (keratins and/or epithelial membrane antigen). Ninety-three per cent expressed keratins (mostly AE1/AE3 or PAN-K), 87% expressed S-100 protein, 86% of the cases were positive for calponin, 63% reacted to EMA, 46% glial fibrillary acidic protein, 36% smooth muscle actin, 23% P-63 and 14% desmin. In their series, 41 cases were malignant myoepithelial tumours [10].

The histological or immunohistochemical features of these tumours cannot aid prediction of the prognosis. The only reliable clue to behaviour is the mitotic activity or the Ki-67 index [11]. Tran *et al.* [2] found the Ki-67 index to be between 30–50% and 5–25% in their report of two cases of MEC involving the orbit and both the patients died within a short time of diagnosis.

Recurrence of a benign myoepithelial tumour is uncommon if the excision is complete. Hornick and Fletcher [10] have reported an 18% recurrence rate in case of benign tumours, whereas the rate of recurrence in case of malignant tumours is 42% with 32% distant metastasis. Myoepithelial carcinoma can lead to recurrence, metastasis as well as tumour-related death. Recurrence and metastasis are more common in children even with a negative excision margin.

Conclusion

Myoepithelial carcinoma of the ear is a rare clinical entity and such a huge mass is yet to be reported in the literature. Histopathology and immunohistochemistry play a pivotal role in tissue diagnosis. Although local recurrence and distant metastasis are known to occur, the biological behaviour and growth pattern vary considerably as in the present case. Subdermal growth, involvement of the external auditory canal wall, the

presence of a tumour in the middle ear cavity and mastoid and parotid without involvement of the facial nerve itself are rare. Complete excision with a tumour-free margin remains the treatment of choice. The extent of surgical excision needs to be customized according to the spread of the disease. Selective neck dissection in myoepithelial carcinoma of the tympanomastoid region even in N0 neck seems to be a prudent decision in the long term. The authors intend to highlight the disease-free survival of the patient, which could be achieved in the present case through proper surgical management followed by radiation therapy. This report may be of importance in the management of this rare disease in future.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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