Malignant Myoepithelioma of the mastoid region – A case report

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Abstract

A 23-year-old male patient presented with a post auricular mass and an attic antral type of chronic suppurative otitis media. He underwent excision of the mass and canal wall down mastoidectomy. Histopathological diagnosis of malignant myoepithelioma was made. The patient refused treatment and later he died with systemic metastasis. This case is reported for the rarity of the site, young age of presentation and aggressive behaviour.

Keywords Malignant myoepithelioma • Mastoid • Cytokeratin • S100

Introduction

Malignant myoepithelioma or myoepithelial carcinoma is a rare tumor, occurring more often in the salivary glands. In 1943, Sheldon first used the term myoepithelioma and in 1975, Stromeyer et al, reported the first case of malignant myoepithelioma. These tumors are mainly seen in the parotid glands, but few cases have been reported in the soft palate, minor salivary glands, nasopharynx, paranasal sinuses, breast, bronchus and lung. They are aggressive neoplasms

with a tendency to invade adjacent structures. Involvement of the ear and mastoid process by myoepithelioma is very rare.

Case report

A 23-year-old male patient presented with a post-auricular mass on the left side. He complained of discharge from the left ear and giddiness for the past one year and pain for the past one month. The patient underwent myringoplasty in that ear in another hospital three months earlier. There was an asymptomatic period of one month following which the symptoms recurred. On local examination the pinna was pushed laterally, with hard, non tender mass in the post auricular mastoid region with obliteration of the sulcus. The auditory canal was also narrowed. There was a mucopurulent discharge from the ear. The other ear showed attic retraction with cholesteatoma flakes and a perforation. Throat and nose were within normal limits. The Pure tone audiogram revealed a conductive hearing loss of 50 db on the right side and 46.6 db on the left side.

Revision mastoidectomy was done and the post auricular mass was excised completely. It was arising from the postauricular soft tissue. Bone was not involved. Histopathological examination of the excised mass showed skin with the deep dermis showing sheets of tumor cells with clear cytoplasm, large pleomorphic vesicular nuclei, some of them with prominent nucleoli (Fig. 1) Brisk mitotic activity and areas of necrosis were also noted. The tumour cells showed strong cytoplasmic positivity for cytokeratin, EMA (Epithelial Membrane Antigen) and S100. The immunostains for LCA, CD 31, CD 1a, HMB 45 and SMA were negative. A diagnosis of malignant myoepithelioma was made. The patient came for follow up for two months and was advised chemotherapy and radiation but refused any form of treatment. Two months later he came with multiple metastasis in the scalp (Fig. 2) and lungs and expired after 4 months

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Fig. 1 Photomicrograph showing the tumour cells with clear cytoplasm and vesicular nuclei (400X)

Fig. 2 Figure showing metastatic deposit in the scalp.

Discussion

Malignant myoepitheliomas are rare tumors arising from the myoepithelial cells of glands. These tumours have been reported in various sites of which salivary gland is a common site, forming 1% of all salivary gland tumors. Among the salivary glands, parotid gland is affected more often¹. Most of these patients have a good prognosis, but several instances of recurrences have also been noted. These tumors have been reported in the minor salivary glands also either denovo or arising in a preexisting pleomorphic adenoma. Tomakazu et al reported a case of malignant myoepithelioma arising from a recurrent pleomorphic adenoma of the minor salivary gland. The patient had two recurrences of which the second one was malignant with massive growth and bone invasion. But with adequate surgical resection the patient has been free of tumour for the past 20 months⁹.

It has also been reported in the soft palate^{2,3}. In the soft palate the tumour was seen in a background of pleomorphic adenoma, but the recurrence was more aggressive with an infiltrating growth pattern and a high mitotic rate. These patients were also free of tumour after an adequate surgical clearance. Few cases have also been reported in the pharynx and nasopharynx. In the pharynx it presents with non specific symptoms like dysphagia, foreign body sensation or respiratory problems and in the nasopharynx, with nasal obstruction, ear fullness, serous otitis media and conductive hearing loss.

In few cases of rhinopharyngeal myoepithelioma, two patients were treated with surgery, followed by radiotherapy and no recurrence was noted. Cheek, retromolar area, oral cavity and breast are the other sites where this tumour is reported⁵. Many pulmonary epithelial myoepithelial carcinomas and few salivary gland myoepithelial carcinomas showed local recurrence and metastasis. In view of the variation in behaviour it is suggested that these tumours should be designated as having uncertain malignant potential. Our case is rare as it occurred in the post auricular region (mastoid)-a non salivary gland origin. Malignant myoepitheliomas show cytological atypia and have a potential for aggressive behavior. The features of malignancy are infiltrative borders,

cellular atypia, frequent mitosis and coagulative necrosis⁶. In a study by Saveria et al the presence of tumor infiltration into the adjacent tissues is the most notable feature and should be considered as the minimum requirement for myoepithelial carcinoma⁷. In the salivary glands, the clinical behaviour and outcome are intriguing. Most tumours displayed marked histologic aggressiveness but some were indolent.

The factors which indicate a worse prognosis are marked cellular pleomorphism, perineural invasion, high mitotic count (>7/10 HPF), high Ki 67 labelling index (>50%) and p53 protein overexpression⁸. Use of electron microscopy or immunohistochemistry is mandatory to substantiate the myoepithelial nature of this tumor. The tumor cells show positivity for cytokeratin (90%), CK14 (53%) Smooth Muscle Actin (50%), calponin (75%), S100 (100%), GFAP (31%), EMA (21%) CEA (0%) and HMB 45 (0%).

Electron microscopy shows desmosomes, array of cytoplasmic microfilaments in the absence of dense bodies and pinocytotic vesicles with basal lamina separating the tumor cells from the stroma. Our patient presented with malignant myoepithelioma of the mastoid region. After surgery he refused further therapeutic intervention, had a rapid downhill course and died within 15 months post surgery with local recurrence and lung metastasis. This reinforces the fact that malignant myoepithelioma should be treated as potentially aggressive neoplasms and more radical therapeutic modalities such as chemoradiotherapy have to be administered.

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