Malignant myoepithelioma of the parapharyngeal space – a case report

K. Ramachandran · M. Venugopal · R. Suma · K. C. Naseer

Abstract Malignant myoepithelioma (MM) of parapharyngeal space provide both a diagnostic and therapeutic challenge. Malignant myoepitheliomas are rare tumour of salivary glands. Most occur in the parotid gland, few other sites of origin are described. Malignant myoepithelioma of the parapharyngeal space are extremely rare with only a few cases has been reported. We present a case of malignant myoepithelioma in the parapharyngeal space and discuss its diagnostic and therapeutic aspects.

Keywords Malignant Myoepithelioma · Parapharyngeal space

Introduction

Neoplasms arising in the parapharyngeal space constitute 0.5% of all head and neck tumours. The majority of these are benign (70%) and 45% are of salivary gland origin [1]. Myoepitheliomas are rare tumours that account for about

1% of all salivary gland tumours. Most are benign, but some have been malignant. The latter, even rarer, entity has been termed malignant myoepithelioma (MM) or myoepithelial carcinoma and represents about 10% of myoepitheliomas. In 1943, Sheldon first used the term myoepithelioma and in 1975 Stromeyer et al [2] reported the first case of MM. Most common site of MMs are in parotid gland, other sites such as the oral cavity, palate, retromolar area, cheek, lacrimal gland, breast, larynx, nasopharynx has been involved.

We present a case of Malignant Myoepithelioma in the parapharyngeal space and discuss its therapeutic aspects.

Case report

A 28-year-old women presented with a four month history of a painless mass in right tonsillar region with fullness in right retromandibular groove. She reported no dysphagia nor history suggestive of upper airway problem. On examination there was a smooth bulge involving (R) side of soft palate and (R) tonsil pushed medially. The swelling was firm, mobile and covered with intact mucosa.

Computed tomography (CT) showed a mixed density mass in the (R) parapharyngeal space with no obvious lymphnode enlargement (Fig. 1). A fine needle aspiration cytology from swelling showed clusters of dark staining cells in the myxoid back ground and pathologist advised excision biopsy.

Patient underwent excision of the mass via combined transoral and transcervical approach. Tumour was well encapsulated and it was then enucleated from the parapharyngeal space. The patient made an uneventful post operative recovery and post operative chemo-radiation was given.

The first histopathological examination with low power showed numerous sheets and cords of neoplastic cells in a myxoid background with area of central necrosis (Fig. 2). High power showed both spindle-shaped and epitheloid neoplastic cells with mitotic activity suggestive of malignant myoepithelioma (Fig. 3).

K. Ramachandran¹ · M. Venugopal² · R. Suma² · K. C. Naseer³

¹Professor and Head,

²Assistant Professor,

³PG Student

Department of ENT,

Medical College,

Calicut, Kerala

India

K. Ramachandran (✉)

Ph: +91 / 9446463170

e-mail: drkrc_ent@yahoo.co.in

Fig. 1 An axial contrast CT scan showing mixed density mass in the (R) parapharyngeal space

Fig. 2 Hematoxyline-eosin staining (10x) showing numerous sheets and cords of neoplastic cells in a myxoid background with area of central necrosis

Fig. 3 Hematoxyline-eosin staining (40x) showing both spindle-shaped and epithelioid neoplastic cells with mitotic activity

Discussion

Salivary gland neoplasms composed of exclusively of myoepithelial cells (myoepitheliomas) are unusual and intriguing. A majority of the myoepitheliomas described in the literature has been benign and the malignant counterpart has been recognized recently. However, the precise criteria for inclusion of a tumour in this category still remain controversial. Some authors consider it a variation of the pleomorphic adenoma. Others define it as a tumour composed solely of myoepithelial cells. Myoepitheliomas and MMs are considered as two separate clinical pathological entities. MMs are tumours of epithelial origin that may occur with pre-existing benign lesion like pleomorphic adenomas or benign myoepitheliomas but then also may arise *de novo*.

MM are usually a slow-growing painless mass that originates in the parotid gland, however, other sites such as the palate, nasopharynx, lacrimal sac, larynx, cheek and breast have also been reported. Usually affects patients over 50 years old, with no gender preference. Because this seldom associated with pain, their diagnosis can be delayed by months or even years.

When these involve the pharynx, MMs produce symptoms like dysphagia, respiratory problems. Retro-mandibular swelling or medial displacement of tonsil or lateral wall of pharynx has been found on physical examination.

Physical examination of lesion in the parapharyngeal space is limited by their location and so radiological imaging is critical in their evaluation (CT and MRI). CT allows the site and the extension of tumour. MRI is more precise in defining the tumours relationship with the adjacent structures and internal carotid A.

Histologically, two basic growth patterns were identified in these neoplasms. These include a multinodular growth pattern and a diffuse sheet-like arrangement of tumour cells. A wide variety of morphologic tumour cell types was found in these MMs. More than one cell types are seen in majority of tumour. However in the majority of the neoplasms one cell type prevailed. The various cell types present in these tumours were categorized as follows: Epithelioid, Clear, Hyaline (plasmacytoid), Spindle and Mixed [3]. Immunohistochemically the tumour cells in malignant myoepitheliomas are immuno reactive to cytokeratin, smooth muscle actin, and S-100 protein, vimentin, and muscle-specific actin.

There are no discernible histologic features that correlate clearly with behaviour. Overall, most tumours that display marked histologic aggressiveness (cytologic atypia, high mitotic rate and necrosis) behaved adversely, but others with similar histologic features have been relatively indolent [3]. Conversely, occasional low-grade tumours with minimal mitotic activity have metastasized, and a few have been caused patients demise [4]. Di Palma and Guzza [5]

considered MMs to be low grade when it arose in a pleomorphic adenoma and high grade when it arose de novo.

As far as treatment of MMs is concerned, there is limited experience but complete surgical excision is the mainstay of therapy. Several surgical approaches for the management of para pharyngeal mass have been described included transcervical, transcervico submaxillary, transmandibular, transparotid, transoral and infratemporal [6].

In our patient, tumour mass was bulging to oropharynx, therefore, we adopted the combined transoral and transcervical approach. It enabled complete resection of the tumour and there have been no signs of recurrence during the 2 year follow-up period.

Conclusion

MM are rare tumours of salivary glands. The diagnosis of a malignant myoepithelial tumour should be considered wherever one encounters an unusual salivary gland neoplasm, having the histological characteristics described. As far as the treatment of MMs is concerned there is limited experience, but complete surgical excision is the mainstay

of therapy. The role of radiation therapy and chemotherapy is not yet established.

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