

Case Report

Largest mucoepidermoid carcinoma of parotid gland in the world

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ABSTRACT

Mucoepidermoid carcinoma (MEC) is the most common malignant neoplasm of the major salivary glands, accounting for 15.5% of all cases benign and malignant.¹ It has multiple types of morphological variants and thus poses a significant challenge in diagnosis on Fine needle aspiration cytology (FNAC). The aim of this article is to report a case of MEC in a 70-years-old male patient who presented with a painless firm fluctuant swelling in left preauricular area on left cheek to Karnataka Institute of Medical Sciences, Hubli. Detailed history was noted, and the swelling was examined clinically. Radiological investigations were carried out. Fine-needle aspiration cytology was done for the lesion and report suggested tumor of the parotid gland. Total parotidectomy procedure was done. There were no intra operative complications and post operatively there was grade 4 Housemann Brackmann facial nerve palsy on left side. The weight of the tumor was 1.45 kg. The histologic picture confirmed that the tumor was MEC of parotid gland. Through this article we want to report the management of giant MEC, the largest to be reported in world literature.

Keywords: Fine needle aspiration cytology, Mucoepidermoid carcinoma

INTRODUCTION

Mucoepidermoid carcinoma (MEC) of the salivary gland is believed to arise from pluripotent reserve cells of the excretory ducts that are capable of differentiating into squamous, columnar, and mucous cells. Stewart et al in 1945 described it first as a salivary gland tumor.¹

Slightly more than 50-60% of these tumours arise in the major salivary glands, with more than 80% occurring in the parotid gland, 8-13% in the submandibular gland, 2-4% in the sublingual gland and the remainder in minor salivary glands, mostly in the palate.⁶ Other sites are the buccal mucosa, tongue and retromolar area. It has a female preponderance (3:2).² It is more common in adults in their fourth to sixth decades, with the highest prevalence during the fifth decade of life.²

The most common presenting symptom is a slowly enlarging painless mass of several years' duration clinically mimicking a pleomorphic adenoma or other benign neoplasm. Pain or facial nerve palsy may develop, usually in association with high-grade tumours. The neoplasm is composed of three cell types: mucous, epithelial and intermediate in nature. A significant amount of MEC arise from a fusion product of the CRTC1-MAML2 gene that has undergone t(11:19) (q21;p13).^{3,4} There has been a suggestion to a possible linkage to cytomegalovirus and papillomavirus.

CASE REPORT

A 70-year-old male presented to our institute with a large swelling over left side of face in preauricular area for last 35 years but started increasing in size rapidly since 1 year with associated pain. It was firm to hard in consistency.

The swelling was measuring 20 cms×18 cms×12 cms in dimensions. There was no skin and facial nerve involvement on clinical examination.



Figure 1: A large swelling on left side at the time of presentation.



Figure 2: CT scan showing a tumor of in homogenous low to intermediate signal intensity with ill-defined margins with loss of planes and infiltration into the deeper structures.

USG revealed multiple mixed echoic focal lesions with cystic components in left parotid region, largest measuring 3.1×4.6 cms. CT scan showed a large well defined inhomogeneously enhancing soft tissue density mass measuring 16×14×11 cms lesion on left side cheek. Superficial lobe of left parotid gland not seen clearly with loss of fat planes between the lesion and masseter muscle. Deep lobe was involved with no infiltration to facial nerve.

FNAC showed features suggestive of mucoepidermoid carcinoma. The patient underwent total parotidectomy with elliptical incision. Intraoperatively, the tumour was well encapsulated with a pedicle with feeding vessel noted. Tumour had focal areas of invasion to deeper masseter muscle. The marginal mandibular branch of the facial nerve was infiltrated and was sacrificed to achieve R0 resection. The skin was closed in 2 layers and a suction drain was placed. Specimen was sent for biopsy. The weight of this tumour was 1.45 kg.



Figure 3: Preoperative skin incisions marked as an elliptical fashion and later flaps raised.



Figure 4: Intraoperative photo of the tumor after being removed.

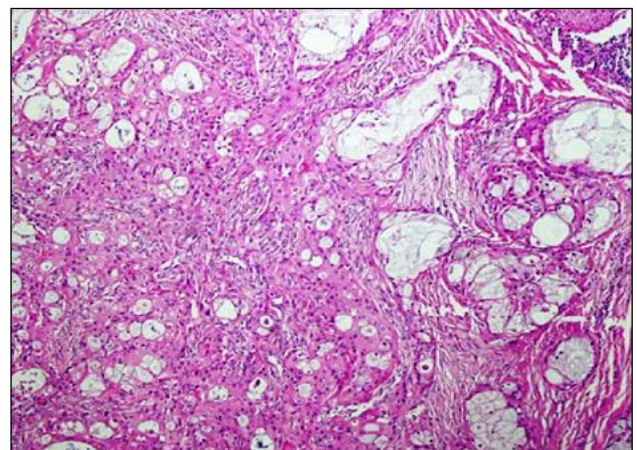


Figure 5: H and E staining of the excised tumor at 10X showing three distinct cell lines; epidermoid cells, mucous cells and intermediate cells.

Histopathology report was consistent with features of MEC-intermediate grade. All the margins were tumor free and parotid lymph nodes did not have metastasis.

Post-operative day one patient developed, left side facial nerve neuropraxia. He had incomplete closure of the left eye and deviation of the angle of the mouth. Drain was removed post op day 3 and patient was discharged on day 7. Patient was followed up.



Figure 6: Post-operative day 2 showing the patient with sutures and romovac drain in situ.



Figure 6: Post-operative day 10 showing houseman Brackmann grade 4 facial nerve paralysis.

DISCUSSION

MEC of the parotid classically present as a painless swelling, usually 2-3 cms.⁴ The tumor grades are classified as low, intermediate, or high on the basis of the traditional grading criteria of architectural pattern (cystic vs solid), cell type (mucous vs epidermoid), and nuclear pleomorphism.

Grading of mucoepidermoid lesions relates in part to the ratio between epidermoid and glandular elements, the high-grade tumors having a larger proportion of the former.

MEC is unique in that it demonstrates a broad spectrum of aggressiveness, from the low grade that rarely kills to its high-grade counterpart that frequently does.

Low-grade MEC tend to create mostly local problems and can have a long natural history. A locally aggressive surgical approach is usually associated with cure. The high-grade and, to a great extent, the intermediate grade mucoepidermoids are often troublesome because they are locally aggressive and are prone to invasion of nerves and vessels as well as to early metastasis. Because of the propensity for regional nodal metastasis in high-grade mucoepidermoid carcinomas, regional nodal dissection combined with generous local or primary resection plus postoperative radiation therapy is important in the treatment of these lesions.

44% of the previously untreated patients with intermediate or high-grade mucoepidermoid parotid tumors develop nodal involvement at some stage.⁵ Prognosis is related to TNMH.

Superficial parotidectomy is the treatment for low and intermediate grade in absence of involvement of deep lobe and parotid lymph nodes. Nodal involvement mandates conservative total parotidectomy along with selective neck dissection.⁶⁻⁸ Also, a dissection of regional lymph node basins should be performed in cases of suspicious adenopathy and in those with more advanced-stage and higher-grade lesions. When treated in this fashion, parotid MEC appears to be a highly curable disease.⁶⁻⁸

Nonetheless, follow-up should continue for an extended time to rule out late recurrence. The overall 5-year survival rates ranged from 0%-43% for patients with high grade MEC, 60%-92% for intermediate grade MEC and 92%-100% in patients with low grade tumours.⁸⁻⁹

CONCLUSION

This rare case presentation of mucoepidermoid carcinoma in the left preauricular region in a 70-year-old man is thought to be the largest described in the literature. It is a atypical case as mucoepidermoid carcinoma are usually of short duration. Despite the availability of treatments such as micro-vascular surgery, wide surgical excision of the tumour is of prognostic value in patients from low socioeconomic backgrounds which is of much value in a country like India.

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