Giant Mucoepidermoid Carcinoma: A Diagnostic Dilemma

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ABSTRACT

Mucoepidermoid carcinoma (MEC) is the most common malignant neoplasm of salivary glands, and accounts for 30% of parotid gland malignancies. The presence of mucoepidermoid carcinoma arising from pleomorphic adenoma (PA) is rarely reported. We present a case of 50 year old female with huge parotid swelling measuring 20cm × 15cm. Immunohistochemical staining helped in making final diagnosis.

Key words *Giant mucoepidermoid tumor, Parotid gland, Malignant salivary gland tumor.*

INTRODUCTION:

Tumors of the salivary glands are rare with an incidence of 5.5 cases per 100,000 individuals.[1] Approximately 80% of all salivary gland tumors arise in the parotid gland and roughly 80% of these are benign.[1] Pleomorphic adenoma is the most frequent salivary gland tumor with an incidence of 67.5%. Benign PA can transform into a carcinoma ex pleomorphic adenoma. Carcinoma ex pleomorphic adenoma (Ca ex PA) is defined as a carcinoma arising from a primary (de novo) or recurrent benign pleomorphic adenoma.[2] This condition is uncommon and often poses a diagnostic challenge.

Presence of mucoepidermoid carcinoma arising from pleomorphic adenoma is rarely reported.[3] A high grade salivary gland adenocarcinoma that is difficult to classify should include Ca ex PA in its differential diagnosis. Prevalence of this tumor is highest in 3rd to 6th decade of life with equal gender distribution. In this study we report our experience of managing such a case.

CASE REPORT:

A 50 year old female presented with a slowly growing huge swelling in right parotid region for the last 15 years. Pain and foul smelling discharge from the mass noted about six months back (Fig I). On examination a firm 20cm × 15cm fungating, nodular mass involving right side of the face, at the parotid region found. Ear lobule was pushed up with obliteration of retromandibular groove. It was firm to hard, lobulated with well-defined margins, not adherent to underlying structures having little mobility. Curtain sign was positive. Skin ulceration of 5cm × 5cm with yellowish discharge on lateral aspect of swelling with everted margins, was noted. Facial nerve was intact on examination. Lymphnodes were not palpable (Fig II).

CT scan revealed a 21cm × 15.5cm × 18cm heterogenously enhancing encapsulated mass lesion involving the deep lobe of the right parotid gland. Right mandibular angle and the mandibular cortex were intact. There was no abnormal perineural or peri vascular enhancement. No lymph nodes enlargement was noted. FNAC showed clustered

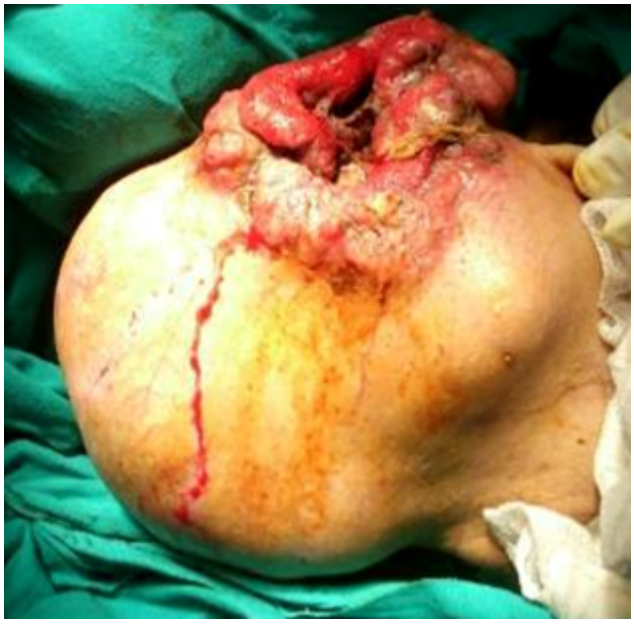

Fig I: A large fungating mass in parotid region

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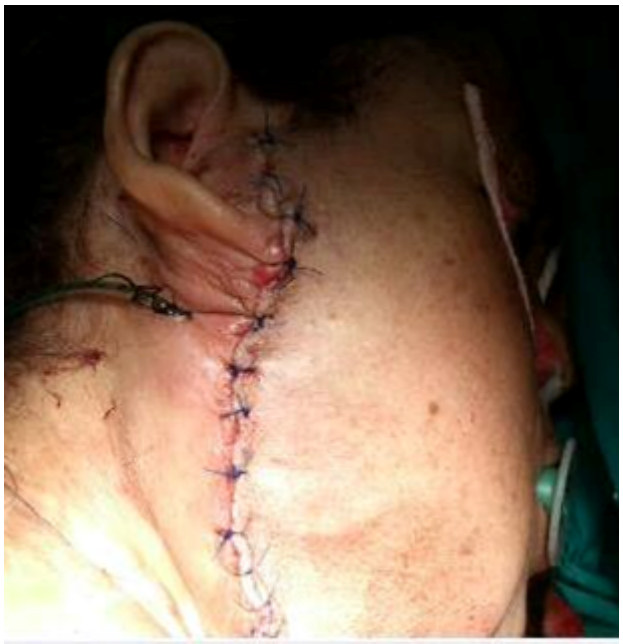

Fig II: Postoperative result.

epithelium-like cells and scattered cells in a necrotic background. The cells, especially the latter, exhibited significant cellular pleomorphism and had irregularly shaped nuclei. Myxoid stroma-like cell clusters with cellular atypism also seen.

Patient underwent conservative parotidectomy through transparotid approach with facial nerve preservation. Histopathological report revealed neoplastic lesion arranged in lobules surrounded by fibrous septa, admixture of epithelial and myoepithelial cells with mucoid and chondromyxoid background. Sheets of fat and calcification and bone formation, having plasmacytoid morphology showing ciribriform architechture with central comedo necrosis seen. Stroma was desmoplastic.

A diagnosis of carcinoma ex pleomorphic adenoma was made, but after immunohistochemical stains that showed CKAE1/AE3, CK7, CK20, S100 positive, changed to mucoepidermoid carcinoma, high grade with perineural invasion and necrosis. The lesion was 0.2 cm away from the lateral borders and from deep resection margins. Lesion was away from medial and inferior border. The patient was referred to radiation oncology for radiation therapy. Patient remained well till date without recurrence.

DISCUSSION:

Salivary gland carcinomas are a rare neoplasms. The pleomorphic adenoma is the most frequent with an incidence of 67.5% . The benign PA can transform into a carcinoma ex pleomorphic adenoma.[1] The risk of malignant transformation in PA varies from 5% to 10%.[4] Many authors claim that the risk of malignant transformation increases with the duration of disease as well as with tumor size. Carcinoma ex-pleomorphic adenoma is an un-common malignancy, accounting for roughly 11% of primary tumors of the salivary glands.[1]

Patients either present with a low or high stage disease depending upon duration of tumor.[5] In our case lead time was 15 year and tumor of high grade. Many subtypes of salivary gland carcinoma have been reported in association with PA. Adenocarcinoma is the malignant component in the majority of cases. The existence of MEC arising from PA is rarely reported.[6] Mucoepidermoid carcinoma arise from the pluri-potent reserve cells of salivary gland ducts. Presence of mucoepidermoid carcinoma in pleomorphic adenoma is rare.[7] MEC is a rare disease of the salivary glands usually occurring as a small tumor. Skin fixation, ulceration or fixation to adjacent structures may indicate malignancy.[8]

The histopathological features of this tumor is nearly identical to mucinous eccrine carcinoma of skin, mucinous carcinoma breast and colloid carcinoma of intestine.[9] The immuotyping of CK7/CK20 can aid to determine the tumor origin based on careful clinical history and evaluation (CK7+/Ck20- phenotype may show salivary gland tumor, whereas CK7- /CK20+ profile may be a clue to an intestinal origin).[9] Overall, none of these preoperative diagnostic measures are of high accuracy when used alone. Hence, a combination of diagnostic tool should be available.

Complete surgical resection is the mainstay of treatment for all grades of MEC. Large size of the tumor may be an obstacle in resection during an operation. Chemotherapy as single modality of therapy has not been proven to be effective. Radiation therapy is considered the cornerstone of adjunctive therapy and plays an important role in those patients with advanced stage disease.[8]

Low-grade MEC behaves less aggressively and is typically treated with surgical excision alone. High-grade tumors are generally treated with wide surgical excision with lymphadenectomy and adjuvant radiotherapy. Intermediate-grade tumor treatment is not well established. According to Wright et al these tumors have high incidence of recurrence within 2 to 3 years of treatment.[10] A long term follow up in these patients with a minimum period of 5 years is therefore recommended. This case emphasizes the importance of the correlation among clinical, histopathological and immunohistochemistry

in order to reach an appropriate diagnosis.

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