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A Case of Salivary Gland Carcinosarcoma- Monophasic Sarcomatous Pattern on Tumor Recurrence

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Abstract **Case Report**

Carcinosarcoma is an aggressive, biphasic high grade malignant neoplasm. Carcinosarcoma of salivary gland is an extremely infrequent neoplasm and only less than 80 cases were reported in the literature since the original description by Kirklin *et al* in 1951. We report a case of Carcinosarcoma ex pleomorphic adenoma in deep lobe of parotid in a 64 year old lady. This case is reported because of the rarity of carcinosarcoma in salivary gland, and to make aware of the unusual monophasic sarcomatous pattern of tumor recurrence, else an erroneous diagnosis of second malignancy may be made.

Keywords: Carcinosarcoma ex pleomorphic adenoma, Malignant mixed tumor, Parotid gland.

INTRODUCTION

The global annual incidence of salivary gland tumors varies from 0.4 to 13.5 cases per 100,000 populations and the frequency of malignant salivary gland tumors ranged from 0.4 to 2.6 cases per 100,000 populations [1]. The commonest type of salivary gland tumor is benign mixed tumor (pleomorphic adenoma) accounting for about 50% of all salivary gland tumors. Malignant mixed tumor is a rare entity, which comprises four distinct types of lesions [1]. Since the original description of mixed carcinoma and sarcoma of parotid gland by Kirklin *et al*. in 1951, only less than 80 cases were reported in the literature till date [2, 3].

A 64 year old female patient reported to the ENT outpatient clinic in 2015 with a complaint of change in voice of two weeks duration. Clinical examination showed a bulge involving the right anterior pillar and soft palate with shifting of uvula to the left. There was no external swelling in right pre-auricular region. In CECT of neck, a soft tissue density enhancing lesion was identified in right parapharyngeal space. No bony erosion or enlarged cervical lymph node was identified. Per-oral FNAC from the same lesion showed findings suggestive of pleomorphic adenoma, but no surgical treatment was done. Within a period of 2 years she developed difficulty in swallowing due to a hard mass involving right side of soft palate and oropharynx. CT of neck showed a soft tissue lesion in the right parapharyngeal space, measuring 9x6x8 cm, with cystic and necrotic changes. Lesion extended from base of skull superiorly, to upper border of C5 vertebra inferiorly, obliterating nasopharynx medially and displacing carotid vessels laterally. No bone involvement, restriction of tongue movements or lymphadenopathy was identified. The right parapharyngeal mass was removed in multiple pieces through a per-oral incision made on soft palate. Excision specimen together measured 10x6x3 cm and weighed 200 gm. It had yellowish white appearance with chondroid areas. Microscopic examination showed salivary gland tissue and mucosal tissue with an infiltrating biphasic neoplasm composed of epithelial and mesenchymal cells (Figure 1) Epithelial cells were arranged as solid nests, tubules, and cords along with myoepithelial cells and foci of squamous metaplasia, separated by chondromyxoid mesenchymal stroma, consistent with pleomorphic adenoma. In some areas these epithelial cells had high nucleo-cytoplasmic ratio and hyperchromatic nucleus, suggestive of malignancy. In other areas, the neoplasm showed a lobular architecture where chondromyxoid stroma along with small round to ovoid cells in cribriform or psuedoacinar pattern was observed. These small cells had moderate amount of eosinophilic cytoplasm, vesicular round nucleus and prominent single nucleolus. Since the same neoplasm had areas of pleomorphic adenoma and cells with malignant features in the form of undifferentiated carcinoma and myxoid chondrosarcoma, a diagnosis of Malignant mixed tumor of salivary gland was made.

Within 6 months of surgery, the patient developed a firm to hard irregular 8x6 cm sized

swelling in right pre-auricular region which was fixed to mandible. Recurrence of the tumor was suspected, and so right radical parotidectomy, along with right hemimandibulectomy and right submandibulectomy was done to remove the tumor. Peroperatively, the facial and lingual nerves were infiltrated by the tumor. The excision specimen together measured 10x6x6 cm and weighed 200 gm. Ramus of mandible appeared bulged out. Cut section of specimen showed a grey-white glistening neoplasm involving the ramus of mandible and adjacent soft tissue (figure 2). Microscopic study showed an infiltrating neoplasm with lobular architecture, involving ramus of mandible, periosteum and attached gingival soft tissue. Within the lobules, chondromyxoid stroma and small round to oval neoplastic cells were seen, which resembled the myxoid chondrosarcomatous component of the previous biopsy. No evidence of pleomorphic adenoma or malignant epithelial component was seen in the specimen even after thorough sampling. Both resected ends of the bone, submandibular gland and residual parotid gland were free of neoplasm. So, the recurrence of tumor showed only the malignant mesenchymal element in the form of myxoid chondrosarcoma.

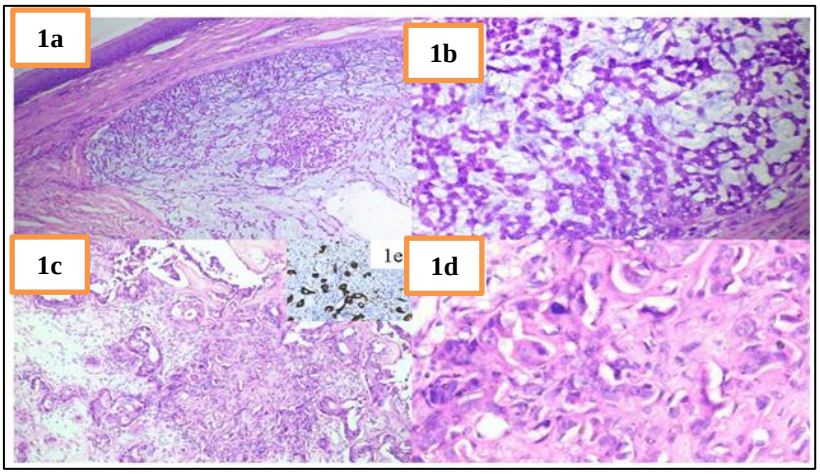

Fig-1

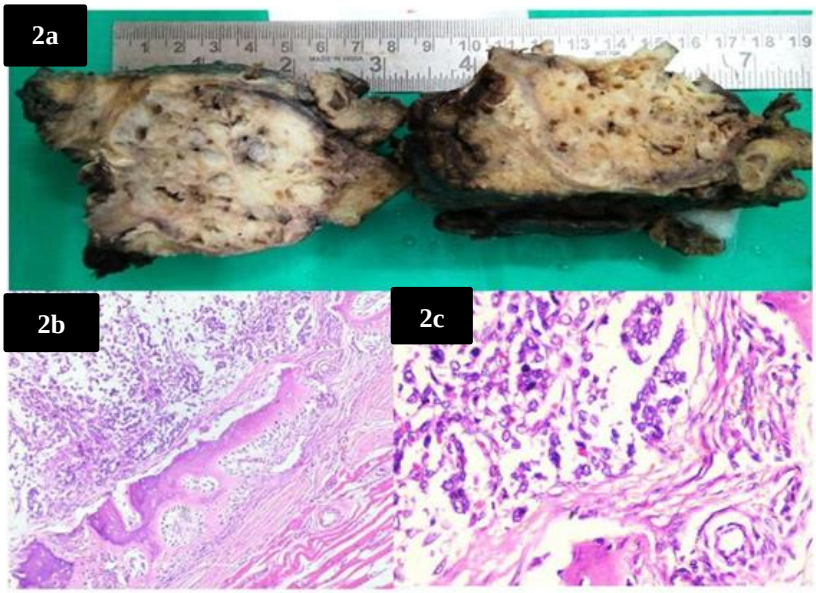

Fig-2

Figure 1
a) Mucosa with tumor having features of pleomorphic adenoma (H&E x100)
b) Focal myxoid chondrosarcomatous component(H&E x100)
c) Focal undifferentiated carcinomatous component(H&E x200)
d) Undifferentiated carcinomatous component(H&E x400)
e) Undifferentiated cells positive for cytokeratin (x200)

Figure 2
a) Mandible with infiltrating neoplasm having lobular architecture
b) Myxoid chondrosarcomatous tumor infiltrating the bone(H&E x100)
c) Myxoid chondrosarcomatous tumor infiltrating the bone(H&E x400)

DISCUSSION

The term Malignant mixed tumor was first coined by King *et al.* in 1967[4]. Malignant mixed tumor is a rare disease and comprises four distinct types: 1. Metastasizing mixed tumor is a histologically benign pleomorphic adenoma that inexplicably manifests local or distant metastasis. 2. Carcinoma ex pleomorphic adenoma is a pleomorphic adenoma from which an epithelial malignancy is derived. 3. Carcinosarcoma ex pleomorphic adenoma is a pleomorphic adenoma from which a malignant tumor composed of a mixture of both carcinomatous and sarcomatous elements are derived. 4. Carcinosarcoma or true malignant mixed tumor is a malignant tumor composed of a mixture of both carcinomatous and sarcomatous elements without histological evidence of pre -existing pleomorphic adenoma [1].

Carcinosarcoma of salivary gland is an extremely rare neoplasm. Gnepp *et al.* in 1993 published a review of 43 cases of carcinosarcoma of salivary glands. Majority were reported in parotid, followed by submandibular and minor salivary glands in palate [5, 6]. Generally these are poorly circumscribed and infiltrative neoplasms, histopathology being the gold standard for diagnosis. In this biphasic tumor, the relative proportion of carcinomatous and sarcomatous component is variable and at times one of the components is evident only on thorough searching. Immunohistochemistry is helpful in confirming the diagnosis. The carcinomatous component may be adenocarcinoma, squamous cell carcinoma, undifferentiated carcinoma, salivary duct carcinoma, mucoepidermoid carcinoma, adenoid cystic carcinoma or epithelial myoepithelial carcinoma. Sarcomatous component is usually chondrosarcoma. However, cases with fibrosarcoma, osteosarcoma, myxoid sarcoma, undifferentiated sarcoma and rarely rhabdomyosarcoma are also reported [6-9]. Destructive local infiltration, angioinvasion and perineural invasion are common in carcinosarcoma. Treatment is wide surgical excision and radiotherapy. However local recurrence and or metastasize to lung, bone or brain occur in 60% of patients within a 30 month period. In our case regular follow up of the patient was available for a period of six months only.

Usually both malignant epithelial and mesenchymal components will be present in tumor when it recurs or metastasizes. In 2016, Mansour *et al.* reported a recurrent tumor with pure carcinomatous pattern in an old case of parotid carcinosarcoma [10]. In our case, pure sarcomatous pattern was observed in recurrent tumor.

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