Primary malignant mesenchymoma arising from submandibular salivary gland: A rare case report

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ABSTRACT:

Malignant mesenchymoma is a rare tumour in which there are two or more than two distinct mesenchymal components. These are generally considered as high grade neoplasms and are associated usually with poor prognosis. Here, we report a case of malignant mesenchymoma containing undifferentiated spindle cell sarcoma, leiomyosarcoma, chondrosarcoma, osteosarcoma and areas with rhabdoid differentiation in a 54 year old male. The primary tumour measured 5.5 x 4 x 3 cm in size; weighing 135 gm arising from left submandibular salivary gland. Fine needle aspiration cytology showed presence of pleomorphic spindle cell clusters with atypia and myxoid stroma. An impression of malignant salivary gland neoplasm was given. Diagnosis of malignant mesenchymoma was made on histopathological examination supported by immunohistochemistry showing strong positivity with p53, Ki-67 and focal positivity for SMA, S-100, Desmin and negativity for CK. The exact histogenesis of malignant mesenchymoma as well as the optimal management strategy to decide prognosis still remains uncertain as it is a very rare tumour.
KEY WORDS: Malignant mesenchymoma; salivary gland; mesenchymal component

INTRODUCTION:

Malignant Mesenchymoma (MM) is a rare malignant soft tissue neoplasm which usually contains two or more different mesenchymal components. This term was first used by Stout in 1948. Only few cases of MM have been described in the literature till date and as per our knowledge, none in the salivary gland. Other mesenchymal tumours with two or more different morphologies like dedifferentiated liposarcoma, dedifferentiated chondrosarcoma and liposarcoma with cartilaginous, smooth muscle and osseous differentiation should be excluded for correct diagnosis. Immunohistochemistry (IHC) plays major role in accurate diagnosis of this entity. These tumours are usually high grade and are seen in different organs.

We report a rare case of MM in submandibular salivary gland in an elderly patient.

CASE REPORT:

A 54 year old man presented to Outpatient Department of Otorhinolaryngology with swelling under left jaw increasing in size for last 1 month. Initially, it was about 0.5 x 0.5 cm which gradually increased to the present size of 7 x 4 cm [Figure 1]. Patient was addicted to tobacco since 30 years. There was no associated fever, pain, voice change, breathing difficulty, dysphagia, weight loss and/or history of exposure to tuberculosis.
Local examination revealed presence of left sided neck swelling of 6.5 x 4 cm extending from lower border of mandible to the submandibular area. The overlying skin was tense but local temperature was not raised. There was no redness or ulceration. The swelling was immobile and was firm in consistency. On general physical and ear, nose, throat examination, no abnormality detected. Ultrasonography neck showed the evidence of a hypoechoic, irregular, lobulated lesion of size 4.8 x 4.1 x 3.3 cm in the left submandibular gland. There was also evidence of multiple hypoechoic lesions of average size 1.3 x 0.5 cm. The rest structures in the neck were normal. A suspicion of malignancy was raised and histopathological correlation was advised. We could not do further preoperative evaluation in the form of

Figure 1: Swelling under left jaw of present size of 7 x 4 cm.
magnetic resonance imaging as patient did not give consent for the same due to financial constraints.

Fine needle aspiration cytology (FNAC) was performed using a 26G needle and 10 mL syringe. Smears were stained with Papanicolaou and Giemsa stains. Smears showed presence of tightly clustered pleomorphic spindle cells. The spindle cell clusters showed nucleomegaly along with atypia with presence of myxoid stroma [Figure 3A]. The lesion was suspected as malignant salivary gland neoplasm with two differential diagnoses of pleomorphic ex carcinoma or malignant myoepithelioma and histopathological confirmation was advised.

Intraoperatively, marginal mandibular nerve was stretched. During surgical excision of the tumour, blunt dissection was performed taking care to not to damage marginal mandibular nerve and lingual nerve. Whole tumour mass was excised and sent for histopathology. On gross examination, the mass measured 5.5 x 4 x 3 cm in size. External surface was grey white and firm in consistency. Cut surface showed homogenous greyish white appearance without necrosis. [Figure 2]. Histological examination revealed presence of malignant spindle cell component [Figure 3B]. The tumour cells were spindle in shape with eosinophilic cytoplasm. There was proliferation of poorly differentiated immature mesenchymal cells with hyperchromatic nuclei with marked atypia. The mitotic figures were higher than 5 mitoses per 50 high power fields. There was marked cellular polymorphism along with numerous typical and atypical mitotic figures [Figure 3C,D]. There were also areas of chondrosarcoma having atypia with abundant cartilaginous matrix with hyperchromatic nuclei [Figure 3E]. The section also showed areas of osteosarcoma showing pleomorphic spindle cells producing eosinophilic osteoid [Figure 3F]. There was also component of leiomyosarcoma containing hypercellular spindle cells with nuclear atypia and abnormal mitotic figures [Figure 3G]. There was presence of rhabdoid differentiation at places showing moderately cellular areas with sheets of small spindle cells having deeply eosinophilic cytoplasm with elongated tails (tadpole cells) and
inconspicuous nucleoli [Figure 3H]. The spindle cells showed focal positivity to desmin and smooth muscle actin (SMA) [Figure 4A,B]. S-100 was positive in chondrosarcomatous region [Figure 4C]. p53 and Ki-67 were strongly positive throughout [Figure 4E,F]. The tumour was negative for pancytokeratin [Figure 4D]. On the basis of morphology and IHC, a diagnosis of MM was made. Post-operatively, after receiving radiotherapy, patient was disease free at 3, 6, 12, 18 and 24 months of follow up.

Figure 2: Cut surface of submandibular salivary gland showing homogenous greyish white appearance without necrosis.
Figure 3: (A) FNAC showing spindle cell clusters with prominent nuclear enlargement with atypia and myxoid stroma (Giemsa, x400), (B) Predominant spindle cell sarcoma (H&E, x100), (C) Marked cellular polymorphism with typical and atypical mitotic figures (H&E, x400), (D) Spindle shaped tumour cells with abundant eosinophilic cytoplasm (H&E, x400), (E) Areas of
chondrosarcoma (H&E, x100), (F) Areas of osteosarcoma with osteoid production (H&E, x400), (G) Areas of leiomyosarcoma (H&E, x400), (H) Areas of rhabdoid differentiation (H&E, x100).

**Figure 4:** (A) Focal positivity for Desmin (IHC, x400), (B) Positivity for Smooth Muscle Actin (IHC, x400), (C) S-100 positivity in chondrosarcomatous region (IHC, x400), (D) Negativity for pancytokeratin (IHC, x400), (E) Strong positivity for p53 (IHC, x400), (F) Strong positivity for Ki-67 (IHC, x400).
DISCUSSION:

MM is a unique tumour which consists of two or more different malignant mesenchymal components like osteosarcoma, chondrosarcoma, leiomyosarcoma and liposarcoma. It can occur at all locations in the body which includes the retroperitoneum, soft tissue of lower limbs, heart, pleura, liver, orbit, larynx, thyroid, bone, mediastinum and urinary bladder. The occurrence of this tumour in submandibular salivary gland is unusual and as per our knowledge, has not been reported till date in English literature. It appears to arise from primitive mesenchymal cells which have capacity for totipotent differentiation; but the histogenesis still remains uncertain. While diagnosing MM, we have to take into consideration that some sarcomas have osteoid and chondroid differentiation that may imitate this entity.

On cytology, diagnosis was given as a malignant salivary gland neoplasm with possibilities of pleomorphic ex carcinoma or malignant myoepithelioma. But on histology, there was presence of four distinct malignant mesenchymal components. Further on IHC, the tumour cells were negative for pancytokeratin which excluded epithelial component. Partial positivity for smooth muscle actin and desmin showed myoblastic differentiation. The chondrosarcomatous region was neoplastic and showed positivity for S-100. The differential diagnoses can be spindle cell sarcoma with osseous and cartilaginous regions and malignant peripheral nerve sheath tumours (MPNST) wherein cartilaginous and bone formation is common. But in MPNST, the osseous and cartilaginous component is reactive while in present case, it was malignant. Further, S-100 was negative in spindle cell component being positive only in chondrosarcomatous region ruling out MPNST. The other tumours which mimic MM with presence of two or more components include liposarcoma with smooth muscle component; cartilaginous or osseous differentiation dedifferentiated liposarcoma and dedifferentiated chondrosarcoma. All these entities can be differentiated from MM in that the myoblastic component in MM is not dedifferentiated and rest all components are neoplastic.
As the tumour consisted of different components despite a predominant homogenous macroscopic finding on cut surface, it is important to study all areas of tumour by IHC for correct histological diagnosis of MM.

The prognosis of MM is still controversial, according to some authors, it is a high grade malignant neoplasm with poor survival rate.\textsuperscript{4,5} Brady MS et al studied cases of MM and found that 2 and 3 year survival rate as 75% and 37% in femoral and retroperitoneal cases.\textsuperscript{4} Sharma TC et al in his study reported that all 4 patients of MM died of disease within two years.\textsuperscript{5} In the present case, the patient was uneventful till 3, 6, 12, 18 and 24 months follow up period. The presence of different components may suggest outcomes as reported by few authors. Adachi studied a case of MM a 40 year old patient and reported that presence of a rhabdomyosarcomatous component correspond to a poor prognosis. He also suggested that there is no significant difference in prognosis with respect to tumour size, site and gender.\textsuperscript{6}

There are insufficient data to suggest the best treatment option or modality to cure the condition as the tumour is very rare.\textsuperscript{7,8,9} Only chemotherapy or radiotherapy were found to be ineffective. Thus, a multidisciplinary approach including surgery, chemotherapy and radiotherapy may be useful for such tumours as in our case.\textsuperscript{9}

**CONCLUSION:**

We reported a case of malignant mesenchymoma containing undifferentiated spindle cell sarcoma, leiomyosarcoma, chondrosarcoma, osteosarcoma and areas with rhabdoid differentiation in a 54 year old male arising from submandibular salivary gland. As it is a very rare tumour and probably first case in salivary gland, more literature is needed to decide management and prognosis of this extremely rare tumour.
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