CASE REPORT

"Undifferentiated Pleomorphic Sarcoma: Report of a Rare Case"

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Abstract-"Undifferentiated Pleomorphic Sarcoma (UPS), previously called Malignant Fibrous Histiocytoma, is a highly aggressive soft tissue sarcoma. It is the most common soft tissue sarcoma reported in later adult life. The head and neck region, especially maxillary sinus is a very rare site for UPS. Surgical excision with wide margins is the treatment of choice. Even excision of clinically normal soft tissues has been done, since it has been found to infiltrate adjacent tissues leading to recurrences. A local recurrence rate of nearly 50% have been reported.

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Introduction

"Undifferentiated Pleomorphic Sarcoma (UPS), previously called Malignant Fibrous Histiocytoma, is a highly aggressive soft tissue sarcoma originally described by O'Brien and Stout in 1964.[1] It is the most common soft tissue sarcoma reported in later adult life. The head and neck region, especially maxillary sinus is a very rare site for UPS.[1,2,3,4]

Case Report

A 32 year old male patient, farmer by occupation, came to Dept. of Oral Medicine and Radiology with a swelling in upper right back teeth region since 4 months. The swelling started gradually at first, small

in size initially, and reached its present size over a period of 4 months. Initially he approached a dentist for the swelling and because there was mobility of upper back teeth, he underwent extraction. However after extraction the swelling persisted and enlarged in size. There was no history of pain, bleeding, pus discharge or numbness. There was no significant medical history. He had a habit of beedi smoking one packet daily since 20 years. He was from a poor socioeconomic background. On extraoral examination, mild facial asymmetry was present because of the swelling in middle third of the face, 3 × 1.5cm in size, and the skin over the swelling appeared normal, non-pulsatile, non-tender, non-reducible and firm in consistency. Right side submandibular lymph nodes were palpable, tender and enlarged, approximately 1.5×1.5cm in diameter.

Intraorally, a swelling was present on the right maxillary edentulous ridge in the region of 13 to 18, extending into the hard palate and buccal vestibule, 4 × 2.5cm in size, extending anteroposteriorly from 13 to 17 region, which was oval in shape, not associated with bleeding or pus discharge, normal in color as surrounding tissues (Fig. No.1). there was expansion of the buccal as well as palatal cortex. It was non-tender on palpation, soft to firm in consistency, having a smooth surface and was non compressible, non-pulsatile, non-reducible and non-fluctuant.the borders were smooth and indistinct. Other hard tissue findings were non-significant with 26 being caries exposed and 28 having amalgam restoration. A provisional diagnosis of salivary gland tumor was made and differential diagnosis of periapical cyst and non-odontogenic tumor was made.

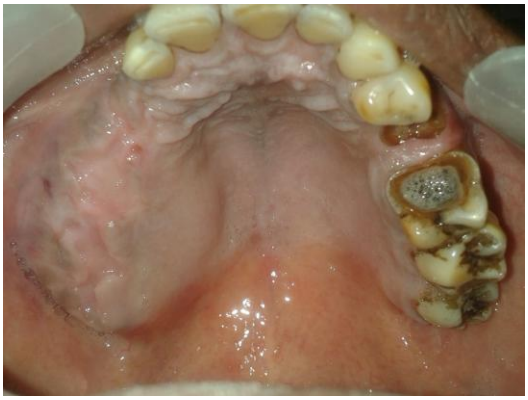

Fig. No. 1: Intraoral view showing swelling extending from 13 to tuberosity region.

Aspiration of the swelling was done and it was non-productive. Conventional radiographs revealed a radiolucent area in the right side of the maxilla, eroding the alveolar cortex of edentulous region of 13 to 18, with soft tissue extending into maxillary sinus (Fig No. 2). CT scan showed homogenous area in

maxillary sinus suggestive of soft tissue mass which was indicative of extension to the sinus also (Fig No. 3). The patient was sent for Incisional biopsy.

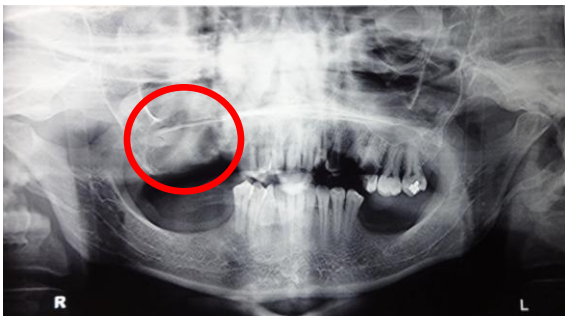

Fig. No. 2: OPG showing radiolucency involving right maxilla and eroding the sinus floor.

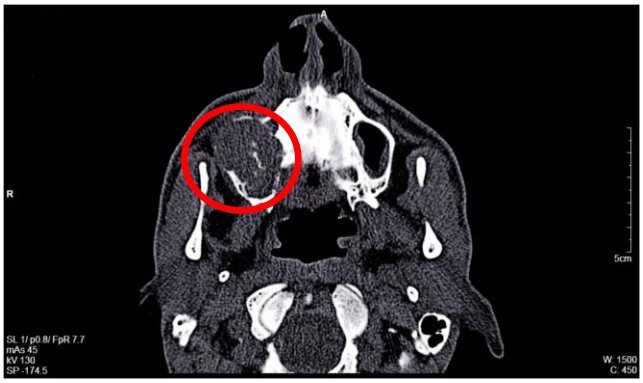

Fig. No. 3: CT view showing the extent of the lesion.

An Incisional biopsy from the region showed a highly cellular connective tissue stroma showing spindle shaped fibroblast-like cells. Interspersed between these was a population of large, ovoid histiocyte-like cells with clear cytoplasm. There were also multiple giant cells scattered throughout the stroma. At areas the spindle cells seemed to be having a whorled or storiform pattern. Based on these findings a diagnosis of Malignant Fibrous Histiocytoma (MFH) or Undifferentiated Pleomorphic Sarcoma (UPS) was established (Fig No. 4) and the patient was sent for surgical excision of the lesion. A maxillectomy was done and the patient had an uneventful period of healing and is currently under strict follow up.

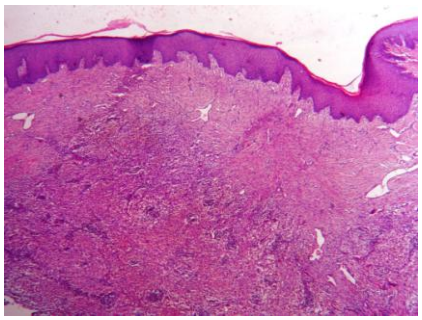

Fig. No. 4: Photomicrograph at 10X magnification.

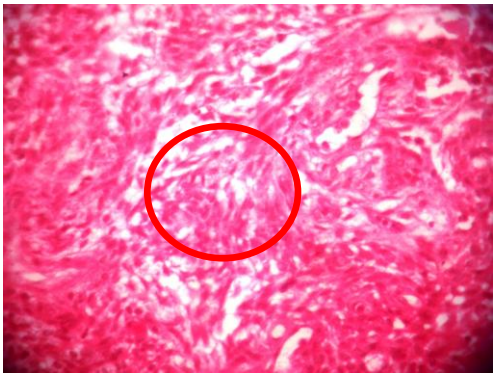

Fig. No. 5: Photomicrograph at 40X magnification.

Discussion

"Undifferentiated Pleomorphic Sarcoma (UPS), previously called Malignant Fibrous

Histiocytoma (MFH) is a highly aggressive soft tissue sarcoma originally described by O'Brien and Stout in 1964.[1] There are more than 10,000 sarcoma cases reported each year, originating from any histologic types like bone, cartilage, fat, muscle, blood vessels, or other connectives.[5] Because of the complexity of origin, and the numerous histological subtypes, the histopathology classification of sarcomas is difficult. UPS is a subtype in this sarcoma group.[6] The term Malignant Fibrous Histiocytoma implied that the tumor cells were of fibroblastic and histiocytic origin; however, the precise origin and fibrohistiocytic differentiation of MFH cells has been questioned since then.[6,7] There are various subtypes of MFH, and since the morphologic pattern seen with pleomorphic type of MFH is shared by a variety of poorly differentiated malignant neoplasms,[6] the World Health Organization (WHO) in 2002, modified the terminology and classification of MFH and its subtypes. Pleomorphic sarcoma is the alternate name by WHO, as it provides an accurate description of the tumor without implying the origin of the tumor cells.[6]

Pleomorphic sarcoma (PS) is most common soft tissue sarcoma, and accounts for 20 - 24% of soft-tissue sarcomas. It is extremely rare in childhood, and is very common in late adult life between 50 - 70 years. It is more commonly seen in males than in females. Extremities (70 - 75% are the common sites in body, particularly, lower extremities (59%).[1] It has been reported to occur in various sites from lungs, kidney, to CNS and oral cavity.[8]

Head and neck region is a very rare site for UPS, with only 1- 3% occurring in this region. [2,3,4] The most common sites in the head and neck are the sinonasal tract, soft tissues of the neck, craniofacial bones, and salivary glands.[9] According to some authors, mandibulomaxillary was the most common site for MFH of the skull followed by calvarium, the reported cases are very few.[10] Primary PS of maxilla and maxillary sinus is very rare, and only around 62 well documented cases have been reported in literature under the name MFH, in maxilla and maxillary sinuses region[11] and only 23 cases in maxillary sinus alone.[2,3,12,13,14,15,16] Some cases can extend into alveolar ridge or present as oral cavity lesions as our case.[17]

In patients with UPS in maxillary sinus, the frequent symptoms at the time of presentation are swelling of the cheek, nasal obstruction, nasal

discharge, and epistaxis.[4,18] Radiographs usually show illdefined radiolucency. The diagnosis of MFH is based on positive immunostaining with vimentin, α1-antitrypsin, and α1-antichymotrypsin.[19, 20,21]

Surgical excision with wide margins is the treatment of choice.[22,23] even excision of clinically normal soft tissues has been done, since it has been found to infiltrate adjacent tissues leading to recurrences.[4,22,23,24] A local recurrence rate of nearly 50% have been reported.[25] The clinical predictors of a poor outcome include advanced age, male gender, underlying systemic illness, large primary tumors, tumors arising from the bones, deep-seated tumors, and a history of previous radiation.[22,26,27,28]

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