Case Report

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Malignant Fibrous Histiocytoma of Right Upper Alveolus

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Abstract

Malignant fibrous histiocytoma (MFH) is a high grade and aggressive sarcoma accounting for around 20% of soft tissue sarcomas. These tumors rarely arise in the head and neck. A case of malignant fibrous histiocytoma of the right upper alveolus in a lady is described. The clinical features, investigative modalities, diagnostic features and management is described.

Keywords: Histiocytoma; Sarcoma; Alveolus; Fibrous; Recurrence.

Introduction

Malignant fibrous histiocytoma (MFH) is a high grade sarcoma originally described by O'Brien and Stout in 1964 [1]. It is the most common soft tissue sarcoma occurring in late adult life between 50 - 70 years and is extremely rare in childhood. MFH occurs most commonly in the extremities (70 - 75%) with lower extremities accounting for 59% of cases followed by the retroperitoneum [1]. Tumors typically arise in deep fascia or skeletal muscle and rarely arise in the head and neck.

Case Report

62 yr old lady presented with history of growth right upper alveolus for 6months with comorbid conditions of diabetes and hypertension. She was a known tobacco chewer for the last 20 years. On examination she had an proliferative 4*3 cm growth right upper alveolus extending from right lateral incisor and posteriorly till right 2nd upper molar involving right gingivo buccal sulcus and right hard palate (Figure 1). Neck Right level II and III nodes 1*1 cm were palpable. Biopsy was suggestive of a poorly differentiated malignant mesenchymal neoplasm.

CECT scan (Figure 2) showed enhancing soft tissue density noted involving the right alveolus of the maxilla measuring about 3x 1.2 cm with minimal bony erosion. Multiple enlarged cervical lymphadenopathy involving Ib, II, III, was seen on the right side. PETCT showed metabolically active disease right upper alveolus (SUV max 10.63) with enlarged cervical nodes were seen at level IB,II,III right side with the largest right level II (33*30 mm, SUV max 13.17). There were no distant metastasis.

Disease was initially staged as Malignant mesenchymal neoplasm Right upper alveolusT4 N2bM0. Patient underwent Right Infrastructural maxillectomy (Figure 3) with Right Modified Neck Dissection Type III with reconstruction by Anterolateral thigh flap (Figure 4). Histopathology on gross showed proliferative tumor hard palate 2.4*2.3*2 cm. All cut margins were free, 10/19 nodes were positive.

Microscopic examination (Figure 5) showed highly pleomorphic spindle to polygonal cells with marked variation in size shape, marked nuclear

atypia with enlarged hyperchromatin nucleus, irregular nuclear membrane, prominent nucleoli and moderate amount of eosinophillic cytoplasm. 2-3 mitosis/10 high power field. These cells were arranged in bundles, fascicles arranged in herring bone pattern and arranged in variable direction in fibromyxoid stroma. Increased angiogenesis was seen. On IHC vimentin was strongly positive. Rest markers cK, Cd31, Cd34, LCA, EMA, HMB, Desmin all were negative. On followup of 3 years patient is asymptomatic with no recurrence.

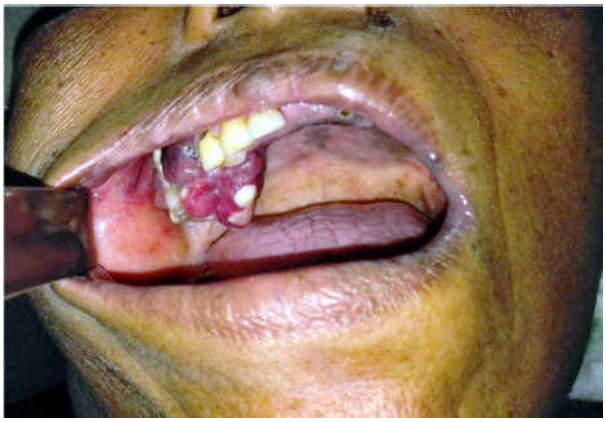

Fig. 1: Showing the tumor involving the Right hard palate and right upper alveolus

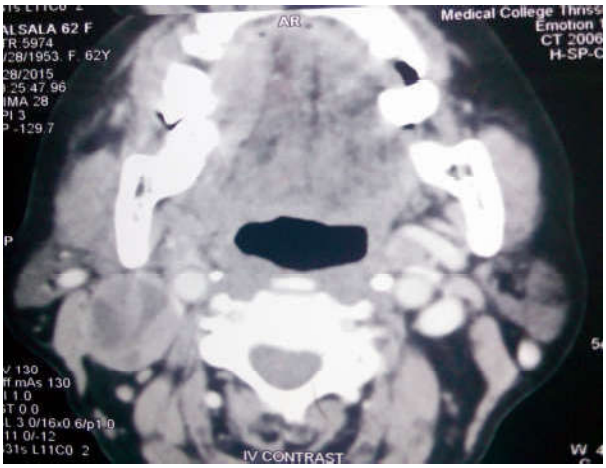

Fig. 2: CECT scan showing the tumor involving the right upper alveolus

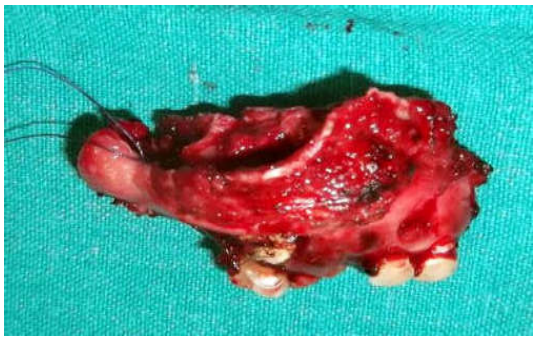

Fig. 3: Showng Right infrastructural maxillectomy specimen

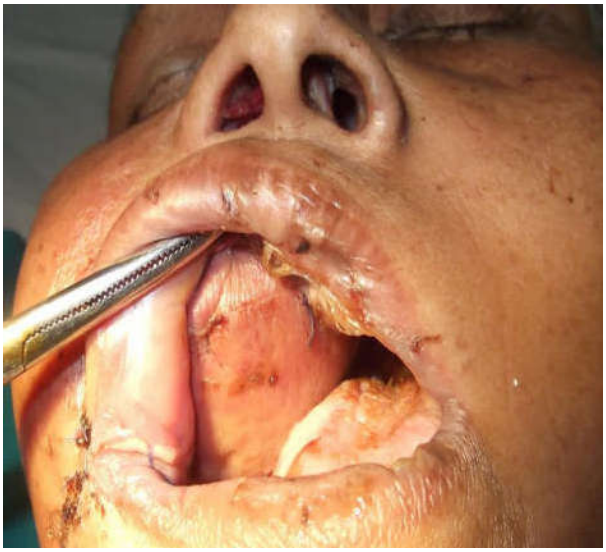

Fig. 4: Showing reconstruction of maxillarydefect by anterolateral thigh flap

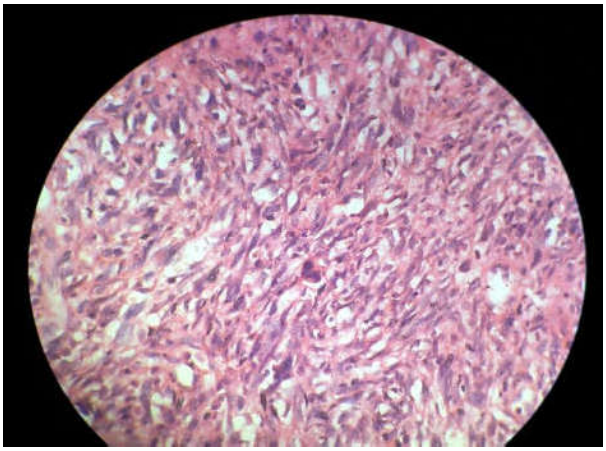

Fig. 5: Photomicrograph high power showing storiform pattern of MFH

Discussion

Malignant fibrous histiocytoma tumors (MFH) rarely occur in the head and neck with an incidence of 1-2% [2]. In this region, prevalence of the tumor in the nasal cavity and paranasal sinuses is 30%, craniofacial bones is 15-25%, larynx and soft-tissue of neck is 10-15% each, and oral cavity 5-15% [3]. In our case, it has affected the maxillary alveolar ridge and the hard palate.

Male predominance has been reported for this neoplasm with a male: female ratio of 2:1 However Chen et al [3] found a female predominance with male: female ratio of 1:2. In our case, it was a primary tumor reported in a lady.

MFH tumors can be either primary or secondary. Primary tumors are less aggressive and more

common than secondary tumors [4]. Secondary tumors are generally associated with underlying conditions such as prior radiotherapy prior trauma, fibrous dysplasia or benign bone tumors like enchondroma. MFH may occur at any age, with a peak incidence in the fifth to seventh decade. MFH is one of the most common Radiation-induced sarcoma of the head and neck. The period of latency between initial radiation therapy and diagnosis of Radiation-induced sarcoma ranged from 9 - 45 years with a median of 17 years [5].

The exact histogenesis of MFH remains controversial. The majority of investigators have suggested that histiocytic and fibrocytic cell lines are derived from the small numbers of undifferentiated mesenchymal cells [6]. In general, the tumor contains both fibroblast like and histiocyte like cells in varying proportions, with spindled and rounded cells exhibiting a storiform arrangement.

Enzinger and Weiss have defined five histological subtypes of MFH as follows: storiform-pleomorphic, myxoid, giant cell, inflammatory, and angiomatoid. The storiform-pleomorphic and myxoid variants are the most common type. MFH is generally seen between the ages of 50 and 70 years. An exception is the angiomatoid variant that usually affects individuals who are younger than 20 years old [1-3]. Tumors with angiomatoid and myxoid patterns are often associated with a more favorable prognosis as they metastasise late and respond well to surgery [2]. The inflammatory and, the pleomorphic types are more aggressive, metastasise early and respond less favorably to surgery alone [7]. Our patient showed malignant tumor arranged predominantly in form of in storiform pattern.

In recent times MFH is considered as a form of fibrosarcoma and the tumor is very likely over diagnosed by some pathologists. This is because the more pleomorphic the tumor, the more difficult is to distinguish from other types of sarcomas, such as spindle cell carcinoma, pleomorphic leiomyosarcoma and pleomorphic liposarcoma Distinction among these soft tissue tumors is best achieved by a joint immunohistochemical and ultrastructural study.

Fibrous histiocytoma is typically immunoreactive for vimentin, and sometimes for smooth muscle actin or alpha-1antitrypsin, but not for desmin, keratin, epithelial membrane antigen, S-100 protein, factor VIII-related antigen, CD34, or carcino-embryonic antigen, supporting the hypothesis that the tumor cells are of mesenchymal origin.

Surgery is the best treatment for this aggressive neoplasm. The advent of more advanced reconstructive techniques including free tissue transfer has made more aggressive surgical resection of these tumors possible. Local recurrence rate of MFH after initial local excision ranges between 16% and 52%. Block et al [7] reported local recurrence or distant metastasis in 55% of cases of MFH. Recurrence is related to size, depth of invasion, and microscopically positive surgical margins. The presence of positive surgical margins after definitive treatment is the single most important factor relating to local recurrence [8].

Owing to the high incidence of local recurrence, postoperative radiotherapy is generally advocated. Incidence of distant metastases is 25-35% in patients with MFH of head and neck region with pulmonary being most common (90%) followed by bone (8%), and liver [3]. The 5-year survival rate ranges from 35-60%. The primary tumor factors associated with a worse prognosis are the histiologic subtype, necrosis, a high mitotic count, and blood vessel invasion [9]. The clinical stage of the tumor, which is defined by tumor grade, size, and presence of distant metastases, is the most important prognostic factor. Other 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 [2,10].

Conclusion

Malignant fibrous histiocytoma of the head and neck is an aggressive tumor. Inadequate resection is related to a higher local recurrence rate and worse prognosis and hence these tumors need to be resected as extensively as possible.

Conflicts of Interest: Nil

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