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Case Report

Recurrent Malignant Fibrous Histiocytoma of Maxillary Sinus: A Rare Case Report

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Abstract

Malignant fibrous histiocytoma is a high-grade and aggressive sarcoma. It is a type of histiocytoma that is the most common soft tissue sarcoma of late adult life. These tumors rarely involve the head and neck region. Wide excision followed by postoperative radiotherapy is believed to be the treatment of choice for malignant fibrous histiocytoma. We present a rare case of malignant fibrous histiocytoma of the maxillary sinus in a 72 year old female with recurrence.

Keywords: Neoplasms; Malignant fibrous; Histiocytoma; Sarcoma; Fibrohistiocytic Tumors; Maxillary Sinus.

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Introduction

Malignant fibrous histiocytoma (MFH) is a high grade and aggressive sarcoma originally described by O'Brien and Stout in 1964.[1] MFH accounts for 20 - 24% of soft-tissue sarcomas, making it the most common soft tissue sarcoma occurring in late adult life between 50 - 70 years and extremely rare in childhood. It is more commonly seen in males than in females and is seen more often in Caucasian than those of African or Asian descent. 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. MFH has been reported to occur in the lung, kidney, bladder, scrotum, vas deferens, heart, aorta, stomach, small intestine, orbit, CNS, paraspinal area, dura mater, facial sinuses, nasal cavity, oral cavity, nasopharynx, and soft tissues of the neck.[2]

It has been estimated that approximately 1-3% of these tumours occur in the head and neck region.[3] Although previous large retrospective studies indicating a local recurrence rate of nearly 50% have been reported, recent studies from cancer centres indicate a lower rate. Data derived from M.D.Anderson Cancer Center & Memorial Sloan showed a recurrence rate of 19 - 31% and a metastatic rate of 31 - 35%.[3]

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.[4] Microscopically, MFH can be classified into five types: pleomorphic storiform, giant cell, inflammatory, angiomatoid and myxoid. The combination of infrequent occurrence, varied pathologic features, uncertain histogenesis, subtypes and the many potential sites of presentation make these tumors a challenge for the diagnostician, surgeon and oncologist. We present a rare case of malignant fibrous histiocytoma of the maxillary sinus in a 72 year old female with recurrence.

Case report

A 72 year old female visited the Oral & Maxillofacial Pathology Department with a chief complaint of a lump and intermittent pain on the right side of the face since 2 months. Her medical history was non-contributory. Her past medical history was insignificant to the current lesion. Patient was ambulatory. On extraoral examination a diffuse ill-defined oval swelling measuring 3 x 4 cm was seen on the right side of the face extending from the inferior border of the zygomatic arch superiorly to 1 cm below the ala of the nose inferiorly obliterating the nasolabial fold. It extended anteriorly from the anterior wall of the right maxillary sinus and 2 cm short of the tragus posteriorly. Epistaxis and nasal discharge was noticed in the right nostril. Intraoral examination revealed a small bulge in the right palatal region measuring 3 x 3.5 cm oval in size extending onto the alveolus and obliterating the buccal vestibule. On palpation the mass was firm, non-fluctuant, non-tender and immobile. No discharge was present. No lymph nodes were palpable.

Orthopantomograph showed completely edentulous maxillary and mandibular arches,

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the right maxillary sinus was hazy and cloudy in appearance. Invasion of the lateral wall of the nose and deviation of the nasal septum was seen. Destruction of the floor of the sinus and encroachment onto the orbital floor was observed. A provisional diagnosis of carcinoma of the maxillary sinus and a differential diagnosis of a low grade sarcoma was made respectively. Routine blood investigations showed a haemoglobin level of 9 mg/dl with mild leucocytosis. The platelet count, fasting and post prandial blood sugar levels and urine examination was normal with no significant findings.

An incisional biopsy was performed under local anaesthesia and the tissue subjected to histopathological examination which revealed a highly cellular lesion with streaming and sweeping fascicles of dark spindle shaped cells arranged in a storiform pattern and pale round and ovoid cells in the background (Fig 1). High power view showed pleomorphic spindle shaped fibroblasts which had features hyperchromatism, altered nuclear cytoplasmic ratio and mitotic figures with multinucleated giant cells. Large atypical pleomorphic histiocytes were seen in the background (Fig 2). A provisional histopathological diagnosis of malignant fibrous histiocytoma was made. The histopathological differential diagnosis included fibrosarcoma and malignant peripheral nerve sheath tumor. The diagnosis was confirmed after subjecting the tissue for Immunohistochemistry (IHC) which showed positivity for vimentin confirming its mesenchymal origin (Fig 3). Other IHC markers when used, showed positivity for alpha-1 antitrypsin (Fig 4) and negativity for S100 protein (Fig 5). A final diagnosis of malignant fibrous histiocytoma was established.

Surgery was carried out under general anaesthesia and the mass was excised. The gross specimen was received by oral & maxillofacial pathology department which was shiny tan white and firm to palpate. On follow up, after 2 months patient showed a recurrent mass in the same region (Fig 6).

Computed tomography revealed a radiodense mass in the right maxillary sinus with extension into the adjacent tissues. The recurrent tumor showed destruction of the orbital floor and erosion of the floor of the right maxillary sinus, extension into the ethmoidal sinus was seen. There was destruction of the nasal septum. Surgery with radiotherapy has been planned again.

Discussion

Malignant fibrous histiocytoma is a highgrade and pleomorphic soft tissue sarcoma, it has been suggested that from 25-35% of patients with MFH of the head and neck will develop metastasis, most often to the lung.[5] This finding suggests that the prognosis of patients with solitary oral primary MFH may be good.

This tumor usually occurs as a painless mass with progressive growth that habitually has a history of less than six months. MFH typically arises in soft tissues, especially in the extremities and the trunk.[6] It is relatively rare in the head and neck region, where the most commonly affected sites are the sinonasal tract, craniofacial bones, larynx, and the soft tissues of the neck.[6] In patients with maxillary sinus tumors, the most frequent symptom at the onset is swelling of the cheek, followed by nasal obstruction, nasal discharge, and epistaxis. These features were in accordance with our case reported here.

Most maxillary sinus tumors are squamous cell carcinomas, which radiographically show an obscure tumor margin, a necrotic area and an infiltrating growth into the surrounding soft tissues.[7] In our case, however, the findings of the computed tomography (CT) scans did not favor a diagnosis of squamous cell carcinoma. The relatively smooth surface, the uniform density, lack of a necrotic area, and clear demarcation from surrounding soft tissues in the CT images may lead to a misdiagnosis of benign tumors or low-grade malignant tumors.[7]

Although MFH has been accepted as a distinct clinicopathologic entity, the exact histogenesis of this sarcoma remains uncertain. The majority of investigators have favoured primitive cells such as fibroblasts and histiocyte-like cells as the origin of this tumours.[8] In general, the tumor contains both fibroblast like and histiocyte like cells in varying proportions, with spindled and rounded cells exhibiting a storiform arrangement. It is classified, based on appearance and predominant cell population, into five different subtypes: Storiform-pleomorphic, Myxoid, Giant-cell, Inflammatory and Angiomatoid.

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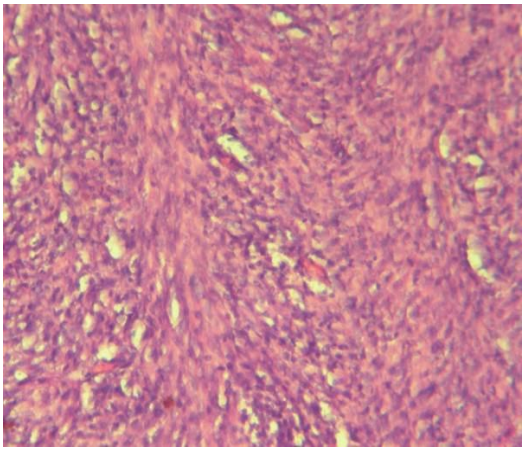

Figure 1: A highly cellular lesion showing sweeping and streaming fascicles of dark spindle shaped cells and pale round and ovoid cells in the background.

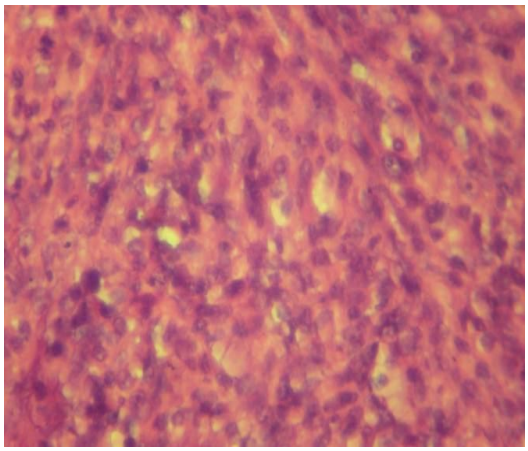

Figure 2: Pleomorphic spindle shaped fibroblasts with features of hyperchromatism, altered nuclear cytoplasmic ratio and vesicular nuclei. Large atypical pleomorphic histiocytes are seen in the background.

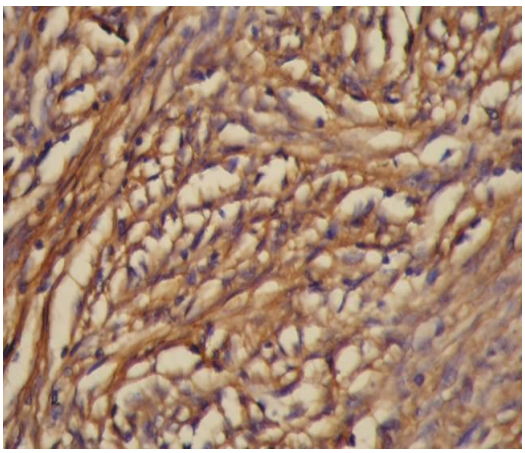

Figure 3: Tumor showing positivity for vimentin.

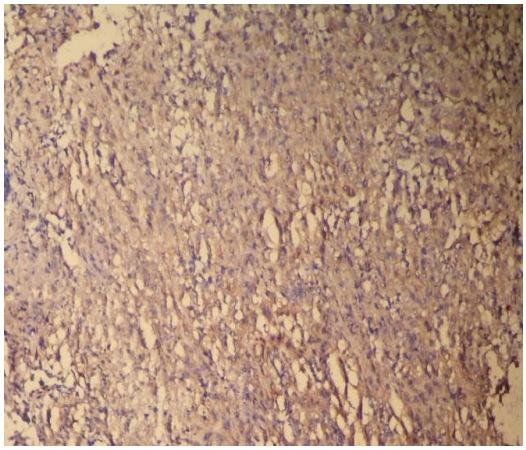

Figure 4: Positivity for alpha-1 antitrypsin.

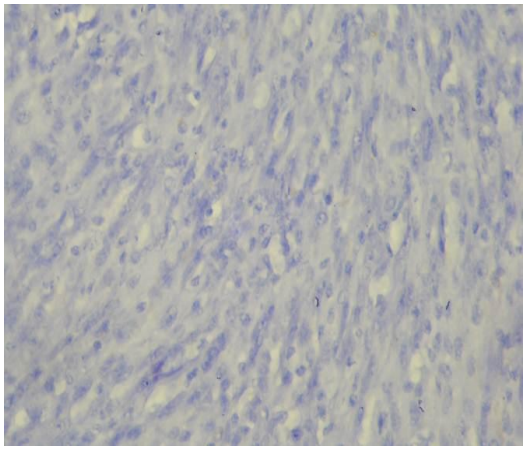

Figure 5: Negativity for S-100 protein.

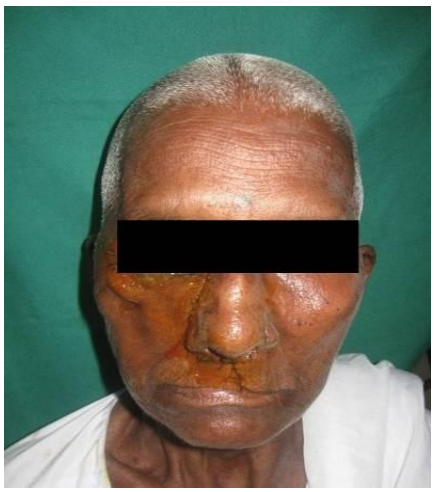

Figure 6: Postoperative photograph.

The pleomophic-storiform type is the most common and is characterized as containing groups of spindle-shape neoplastic cells in a storiform pattern around blood vessels, in addition to histiocyte-like round cells that frequently exhibit a certain degree of pleomorphism. This type tends to be poorly

differentiated and more frequently high grade.[9] In the myxoid variant, mesenchymal cells are presenting a myxoid stroma rich in mucopolysaccharides. The myxoid variant also exhibits a certain degree of cellular pleomorphism, but much less than the previous subtype. It has fewer tendencies to metastasize at a distance and thus has a better prognosis. In the giant-cell subtype, what is peculiar is the presence of a mixture of malignant and benign multinucleate cells and in particular, osteoclastic type giant cells associated with stromal cells that resemble mononuclear histiocytes with variable degrees of anaplasia.[9] The inflammatory variant is characterized by the presence of abundant neutrophils, plasma cells, lymphocytes and occasionally foam cells in poorly collagenized stroma. The angiomatoid type predominates in subjects under the age of 40 years. This type usually is low-grade, with areas of bleeding and hemosiderin deposits, together with a pleomorphic fibrohistiocytic population and high mitotic activity.[9] The present case pertained to the storiform variant.

The diagnosis of MFH is based upon the pathologic features characterized by an admixture of fibroblastic and histiocytic-like cells in a storiform pattern. However, the histologic diagnosis of MFH sometimes is difficult. It should be immunohistochemically differentiated from spindle cell carcinoma, pleomorphic rhabdomyosarcoma, leiomyosarcoma, malignant lymphoma, fibrosarcoma, osteosarcoma, angiosarcoma, pleomorphic liposarcoma and melanoma. Fibrous histiocytoma is typically immunoreactive for vimentin.[10] This was true in our case which showed immunoreactivity for vimentin confirming its mesenchymal origin. Other IHC markers were used, positivity for alpha-1 antitrypsin was noted and negativity for S100 protein was also observed.

Radiation-induced sarcoma of the head and neck (RISHN) is a long-term complication of radiotherapy. In 10 patients with features of RISHN, MFH was the most common pathologic diagnosis. The period of latency between initial radiation therapy and diagnosis of RISHN ranged from 9 - 45 years with a median of 17 years.[11] Wanebo et al[12] in a study of 214 head and neck sarcomas found that the mean survival of MFH at 5 years was 72%, which is higher than the survival for other, more aggressive types of sarcoma, such as angiosarcoma, osteosarcoma and rhabdomyosarcoma.

The advent of more advanced reconstructive techniques, including free tissue transfer, has made more aggressive surgical resection of these tumors possible. The 5-year survival rate ranges from 35-60%. 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 factors include histologic subtype, method of surgical treatment, anatomic site and depth of the primary tumor.[13] Recurrence is usually due to inadequate surgical margins and lack of radiotherapy.

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. For that reason, the primary treatment of this type of tumor is resection as extensive as possible. In case of tumors in which adequate margins are not possible due to their location or large size, postoperative irradiation may improve control of the disease. The mainstays of treatment for MFH are complete surgical excision most often supplemented with adjuvant radiation therapy. Chemotherapy is reserved for patients at the highest risk of disease recurrence or patients that already have recurrence. Favorable prognostic factors that correspond to superior survival include small tumor size, low grade, extremity location, superficial location, and localized disease. We have reported a case of malignant fibrous histiocytoma of the maxillary sinus which is rare, with recurrence.

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