Report of a Malignant Fibrous Histiocytoma in a young patient: A fast growing lump

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ABSTRACT: Malignant fibrous histiocytoma (MFH), a type of sarcoma, is a malignant neoplasm of uncertain origin that arises both in soft tissue and bone. We present a case of MFH in a 18-year-old female patient with swelling on the left maxillary posterior region. This paper demonstrates the malignant character of this neoplasm. CT scans revealed a clouding lesion in the left maxillary premolar and molar region. Histopathological examination showed numerous short fascicles of plump spindle cells arranged in a storiform pattern, admixed with areas of pleomorphic giant cells. Treatment plan consisted of resection of the tumor with maxillary osteotomy and chemotherapy.

Keywords: Malignant fibrous histiocytoma, giant cell, maxilla.

INTRODUCTION

Malignant fibrous histiocytoma is the most commonly diagnosed subtype of soft tissue sarcoma, and was first reported in 1964 by O'Brien and Stout [1]. Malignant fibrous histiocytoma (MFH) arises mainly in the deep soft tissues of the extremities (70%) and retroperitoneum (16%). Occasionally it happens in the inguinal region [2] and rarely involves the spermatic cord and was first described at this site by Cole et al in 1972 [3]. The MFH has variable morphologic features and frequently shows transitions from areas having a highly ordered storiform pattern to less differentiated areas having a pleomorphic appearance. The rate of local recurrence and metastasis is 44% and 42%, respectively. Most frequently sites of metastasis were lung (82%) and lymph nodes (32%) [4].

The most common complaint is an expanding mass that may or may not be painful or ulcerated. Tumors of the nasal cavity and Para nasal sinuses produce obstructive symptoms [5].

Several histopathology subtypes have been described. Multiple subtypes of MFH, including storiform-pleomorphic, myxoid, giant cell, and inflammatory variants, were defined.

The term malignant fibrous histiocytoma implies that the tumor cells are of fibroblastic and histiolytic origin; however, the precise origin of MFH cells has been disputed and the concept of fibrohistiocytic differentiation has been challenged [6]. Furthermore, the morphologic pattern seen with pleomorphic MFH is shared by a variety of poorly differentiated malignant neoplasms.

The storiform-pleomorphic type is the most common. This pattern is characterized by short fascicles of plump spindle cells arranged in a storiform pattern, admixed with areas of pleomorphic giant cells [5].

Initial treatment consists of surgery, surgery and radiotherapy, radiotherapy, and chemotherapy [6]. MFH is usually treated by radical surgical resection. Approximately 40% of patients have local recurrences [5].

Case report

A 18-year-old female patient presented to private office, Qom, Iran, on August 21, 2013, with a compliant of sudden swelling on the left maxillary posterior region. On extra oral examination, the swelling was tender and expansion of the buccal and palatal aspects was apparent. [Figure 1].

Chief complaint of the patient was a firm lump, which first appeared 2 weeks ago. The review of system did not reveal any systemic disorders. According to clinical examination and considering the fast growth of the lesion a malignant lesion was suspected. Malignant mesenchymal tumors were considered in the clinical differential diagnosis of the lesion. To determine the extent of the lesion, CT scan was ordered for the patient, since ultimately the patient had to go under surgery (because of the little influence of FNA results on clinical management). The CT showed the presence of mild bone destruction and clouding lesion in the left maxillary...
premolar and molar region. The cortical bone was substantially expanded. There was no radiographic evidence of maxillary sinus involvement [Figure 2].

After incisional biopsy histopathological examination showed numerous short fascicles of plump spindle cells arranged in a storiform pattern [Figure3], admixed with areas of pleomorphic giant cells [Figure4]. In the more cellular areas, the cells were arranged in short fascicles with focal storiform patterns and pleomorphic cells with hyper chromatic nuclei and a mean of two mitoses per 10 HPF were observed [Figure5]. Curvilinear capillaries were present in the fibrous septa. Angiocentrically distributed aggregates of lymphocytes and plasma cells were seen and no areas of necrosis were noted.

Immunohistochemically, the tumor cells were intensely positive for vimentin and focally positive for CD68 and completely negative for Desmin. [Figure6]. With consideration of immunohistochemical staining results leiomyosarcoma, rhabdomyosarcoma, liposarcoma have been ruled out. The final diagnosis of malignant Fibrous Histiocytoma was made.

A neoadjuvant chemotherapy protocol of epirubicin 60 mg/day on days 1 and 2 and ifosfamide 1.8 gr/day on days 1 through 5 was carried out every three weeks for the patient.

After chemotherapy, maxillary osteotomy from mesial aspect of the second incisor to distal aspect of the first molar was performed. Following surgery, the patient was referred to oncology department for postoperative chemotherapy and radiotherapy.

Six months after surgery, the patient had no sign and symptoms and recurrence.

**DISCUSSION**

Malignant fibrous histiocytoma (MFH) was first described in 1964 under the name malignant fibrous xanthoma. Since that time several major variants have been identified and it has become the most commonly diagnosed of all the sarcomas of adults. Guo et al assessed 33 previously diagnosed MFH cases based on the WHO classification to identify whether some of the MFH diagnoses would have the diagnosis changed. Among the 33 cases, 17 cases (51.5%) of MFH had their diagnoses changed, including 5 leiomyosarcomas, 3 malignant peripheral nerve sheath tumors, 1 fibrosarcoma, 1 inflammatory myofibrosarcoma, 1 giant cell tumor, and 1 angiomatoid fibrous histiocytoma. The remaining 16 cases (48.5%) were reconfirmed as MFH/undifferentiated pleomorphic sarcoma. Only vimentin was always expressed in MFH/undifferentiated pleomorphic sarcoma, while some of the tumors were positive for myogenic antigen and CD68 [8]. Oral and maxillofacial sites are seldom involved, however, and the tumor occurs primarily in the soft tissues of the extremities and retro peritoneum. Most undifferentiated high-grade pleomorphic sarcomas and undifferentiated pleomorphic sarcomas with giant cells occur in the deep soft tissues of the extremities or trunk.

The MFH occurs primarily in adults, especially those 50-70 years of age, but rare cases have been described in children [9, 10]. Regardless of the histopathology subtype, men are affected almost twice as frequently as women but in our case it occurred in an 18-year-old female patient.

Within the maxillofacial region the most common complaint is a moderately firm mucosal mass expanding slowly or moderately fast, with or without pain and surface ulceration. The irregular nodular lesion is typically encapsulated and attached to surrounding tissues and adjacent structures. In our case, the swelling was tender and expansion of the buccal and palatal aspects was apparent [11].

On histopathology MFH has a wide spectrum of cellular and tissue alterations. The cellular differentiation and density vary markedly, even within the same tumor [11].

MFH manifests a broad range of histologic appearances with four sub-types described: Storiform-pleomorphic, Myxoid, Giant cell, Inflammatory. Of these, the storiform-pleomorphic is the most common type, accounting for up to 70% of most cases. The myxoid variant is the second most common accounting for approximately 20% of cases.

Unlike the other sub-types of MFH, the myxoid form tends to be less aggressive and as a result is associated with a better prognosis. Giant cell and inflammatory types are rare. Inflammatory MFH tends to occur in the retro peritoneum [4].

The classic histopathologic features, however, include at least mild cellular and nuclear pleomorphic, an admixture of fibroblastic and histolytic elements [13], and focal areas with a storiform or cartwheel pattern of streaming spindle cells. This classic pattern is the one most frequently encountered in head and neck sites and is often referred to as the storiform-pleomorphic MFH[14,15]. Most lesion cells are spindled fibroblast-like cells which tend to be arranged in short woven fascicles or bundles with scattered areas showing a storiform pattern where fascicles intertwine[12]. In our case, histopathology revealed more cellular areas which were arranged in short fascicles with focal storiform patterns.

The spindle cells in the MFH may be long and thin with minimal atypical, but there are usually areas with plump spindle cells containing enlarged, hyper chromatic and irregular nuclei [16]. Varying numbers of rounded, polygonal and irregularly shaped histiocyte-like cells may dominate some areas of the lesion, often
with very pleomorphic, multinucleated giant cells interspersed [17]. In our case, in the more cellular areas, pleomorphic cells with hyperchromatic nuclei were seen.

The histolytic cells have either abundant eosinophilia cytoplasm or pale foamy cytoplasm, and cell membranes are not easily visualized. Areas with histiocytic predominance usually have a haphazard structural appearance [18].

Chronic inflammatory cells are often scattered sparsely throughout the tumor, including foamy histiocytes, lymphocytes and plasma cells [19]. In our case angiocentrically distributed aggregates of lymphocytes and plasma cells.

Mitotic activity varies widely and is directly related to the degree of cellular pleomorphism [20]. In our case a mean of two mitoses per 10 HPF were observed.

The giant cell variant of MFH is characterized by large numbers of osteoclast-like multinucleated giant cells scattered throughout a cellular stromal of histiocytoid and fibroblastic cells arranged in storiform nodules separated by fibrous septa [21]. Stromal cells usually demonstrate moderate nuclear pleomorphism with occasional bizarre, hyperchromatic nuclei. A large proportion of cases show osteoid, even calcified bone, at the periphery [22, 15]. In our case, histopathology demonstrated numerous short fascicles of plump spindle cells arranged in a storiform pattern, admixed with areas of pleomorphic giant cells.

While blood vessels are usually inconspicuous. Some lesions present with numerous dilated, branching vessels [23]. In our case curvilinear capillaries were present in the fibrous septa.

The neoplastic cells were diffused positive for vimentin and histiocytic marker CD68 and focally positive in spindle cells for neuroectodermic marker (S100) and smooth muscle actin (SMA). In other authors' work, S100 and SMA were negative [24]. In our case, MFH did not have myofibroblastic features and completely had been negative for Desmin.

Differential diagnosis of MFH includes: Leiomyosarcoma, Rhabdomyosarcoma, Liposarcoma, angiosarcoma, hemangiopericytoma [25].

The malignant fibrous histiocytoma is an aggressive tumor that is usually treated by radical surgical resection. Surgery with block resection of tumor is the first choice of treatment. Surgical technique varies, depending on the size and localization of the tumor [26, 27].

The prognosis depends on tumor differentiation, vascular invasion, size and on the existence or not of metastasis [29, 30].

42% of the lesions develop metastases, usually within 2 years of the initial diagnosis [30, 31]. The survival rate for patients with oral tumors appears to be worse than for those with tumors at other body sites [32]. In our case, 6 month after surgery, the patient had no sign and symptoms and recurrence.

![Figure 1](image1.png) Figure 1. The lesion had expanded the buccal and palatal aspects.
Figure 2. The CT showed the presence of mild bone destruction and clouding lesion in the left maxillary premolar and molar region.

Figure 3. Numerous short fascicles of plump spindle cells arranged in a storiform pattern (H&E ×100).

Figure 4. Areas of pleomorphic giant cells (H&E ×100).
CONCLUSION

MFH is found in the extremities, less common sites include the head and neck. The giant cell variant of malignant fibrous histiocytoma is determined to be a rare case in the oral cavity. We believe that current case represents a true sarcoma of the oral cavity, currently best classified as a malignant fibrous histiocytoma, giant cell type. This tumor should be distinguished from the epithelial type of osteoclastic giant cell tumor of the oral cavity.

Patients may benefit from pre-operative chemotherapy before surgery. Chemotherapy may reduce the tumor bulk and increases the chance of a limb sparing procedure.

REFERENCES