CASE REPORT

Maxilla tuberosity malignant fibrous histiocytoma with giant fibroblastic cells: Case report and review of literature

K.A. Al-Salihi a,*, K.A. Al-Jashamy b, S. Ab Rahman a, A.R. Samsudin a

a School of Dental Sciences, University Sains Malaysia, 16150 HUSM, PPSG, Kubang Kerian, Kota Bharua, Kelantan, Malaysia
b School of Medical Sciences, University Sains Malaysia, 16150 HUSM, PPSG, Kubang Kerian, Kota Bharua, Kelantan, Malaysia

Received 26 September 2005; accepted 28 September 2005

Summary Malignant fibrous histiocytoma (MFH) of the maxilla is a rare neoplasm. A round 61 cases reported in the international literature since 1974. We present a rare case of primary MFH of the maxilla in the unusual location of maxilla in a 64-year-old man. The tumor was located in the left tuberosity of maxilla extending from the junction between soft and hard palate towards premolar area of edentulous ridge, and measured 7 cm × 6 cm. Histologically, it consisted of spindle-shaped, pleomorphic malignant cells in a storiform pattern associated with histioyte-like cells and giant cells. Mitotic figures were frequent Immunohistochemically, most of the tumor cells were strongly positive for vimentin, and negative with S-100 protein, cytokeratin, actin, desmin, HMB-45 and epithelial membrane antigen. Ultra structurally, the tumor have clearly shown spindle shaped fibroblastic and giant cells with well-known pleomorphic multi-segmented nuclei, prominent branching and often dilated rough endoplasmic reticulum (RER). Histopathological and ultra structural findings are consistent with high-grade MFH of the storiform/pleomorphic subtype. Four months later the patient came with residual/recurrent tumor that was confirmed histopathologically. The literature is briefly reviewed.

© 2005 Elsevier Ltd. All rights reserved.

KEYWORDS
Maxilla;
MFH;
Vimentin;
RER;
Multi-segmented nuclei

* Corresponding author. Tel.: +60 9607663749; fax: +60 9607642026.
E-mail address: elsalihi@yahoo.com (K.A. Al-Salihi).

1741-9409/S - see front matter © 2005 Elsevier Ltd. All rights reserved.
doi:10.1016/j.ooe.2005.09.014
Introduction

Arthur purdy stout, coined the term malignant fibrous histiocytoma, on the basis of tissue culture studies made by Margaret Murray, which purportedly showed that these pleomorphic fibroblastic tumors arose from tissue histiocytes capable of fibroblastic transformation "facultative fibroblasts". Enzinger and Weiss subsequently defined five subtypes of MFH as follows: (1) storiform-pleomorphic, (2) myxoid, (3) giant cell, (4) inflammatory, and (5) angiomatoid. MFH is the most common adult soft tissue sarcoma.1 Recently there are controversial entities regarding tumors of fibroblastic differentiation. The ubiquitous fibroblast is capable of a wide variety of morphologic and functional adaptation in relation to its body site as well as local physiologic and pathologic changes. The active collagen synthesizing fibroblast is a spindle shaped or plump epithelioid cell with prominent juxtanuclear Golgi apparatus and well developed branching rough endoplasmic reticulum (RER).2 There is longstanding disagreement as to whether the cell type composing MFH is a histiocyte or a fibroblast. Initially, it was proposed by Ozzello et al.3 that the cell was "facultative fibroblasts" (a histiocyte that could appear and function as a fibroblast). However, the majority of more recent histochemical, immunohistochemical, and ultrastructural studies of this tumor support the contention that MFH is a form of fibroblastic differentiation.3,4,5 Previously the diagnosis is made by a joint immunohistochemical and ultrastructural study.2

Malignant fibrous histiocytoma, the most common soft tissue sarcoma of adults has a variety of morphological appearance ranging from a markedly myxoid tumor to a hypercellular, often pleomorphic neoplasms. Frequently present are cell in a storiform arrangement.

Malignant fibrous histiocytomas can arise from soft tissue or bone. It’s most common in the soft tissues of the abdomen and extremities, with 23% occur in osseous sites. Although it can be found in the head and neck region, its occurrence is uncommon, accounting for 3–8.5% of the cases.6 Peak occurrence in persons aged 50–70 years. A slight male predominance is observed. It can occur everywhere, owing to its mesenchymal origin.5,9 The most common sites of occurrence in the head and neck are the sinonasal tract.10 soft tissues of the neck, craniofacial bones, and salivary glands. A literature search showed around 61 well-documented cases of malignant fibrous histiocytoma arising at maxilla, maxillary sinuses and zygoma (Table 1).11–40

We describe interesting case of unusual location at left tuberosity of maxilla tumors that had been diagnosed at a very late stage.

Report of the case

A 64-year-old Malay man presented with swelling at left tuberosity of maxilla, which was noticed 2 months prior to seeking treatment. Lately, however the swelling was found to be painful and easily bleed during eating. Upon examination, there was a huge exophytic growth measuring 7 cm × 6 cm over the left tuberosity of maxilla extending from the junction between soft and hard palate towards premolar area of edentulous ridge (Fig. 1). The growth extended laterally from the vestibule to the midline. Clinically, regional lymph nodes were not palpable. Radiographic examination showed aggressive enhances soft tissue mass at left infratemporal space posterior to left pterygoid plate of maxilla. Diffuse within the lateral pterygoid muscle. The mass occupied the left part of hard palate extending to premolar area in which involving the midline. A computed tomography scan (Plain and CECT) from base of skull to thoracic inlet revealed an ill defined soft tissue mass with heterogenous enhancement noted in the left palatine region with central non-enhancement suggestive of central necrosis. It measures 3.6 cm (AP) × 4.1 cm (W) × 5.0 cm (CC). Superiorly the mass extends till the nasopharynx (at C1 level) with obliteration of the right fossa of Rosen Muller. Inferiorly it extends to the level of tongue. Both parotid and submandibular glands are not involved (Fig. 2A and B). The patient underwent to left lower level partial hemimaxillectomy. A soft grayish to pinkish homogeneous tumor mass measuring 8 × 7 × 3 cm was removed and submitted for oral pathology laboratory. Unfortunately this patient had been defaulted the radiotherapy treatment, which was planned earlier as adjunct to the surgery (Fig. 3). He came back to clinic four months later with residual/recurrent tumor, which was confirmed histopathologically.

Materials and methods

Tumor mass was cut into small portions, some of these portions were fixed in 10% neutral buffered formalin, processed for light microscopy and stained with hematoxylin and eosin. For immunohistochemical staining 4 mm thick sections from selected paraffin block were stained with vimentin, S-100 protein, cytokeratin, actin, desmin, HMB45 and epithelial membrane antigen. For transmission electron microscopy study, a portion of solid mass was cut into 1 mm fragments, fixed in 2.5% gluteraldehyde and post-fixed in spurr resin. After examination of 1 μm survey ultra-thin sections, ultrathin sections were cut and stained with uranyl acetate followed by lead citrate. The specimen was examined by transmission electron microscope (TEM) (Zeiss, Germany).

Results

Microscopically, the mass contained interlacing pleomorphic mix of fibroblasts and multi-nucleated cells revealed bizarre nuclei, abundant eosinophilic cytoplasm, and abnormal frequent mitotic figures. Fibroblastic spindle cells arranged in fascicles and myxoid pattern. Necrosis and extensive cellular atypia were also seen. Many areas revealed spindle cells arranged in a storiform pattern. Histiocyte-like cells interspersed between the spindle cells (Fig. 4A and B).

Immunohistochemically, most of the tumor cells (80%) were strongly positive for vimentin (Fig. 5A and B) and negative with S-100 protein, cytokeratin, actin, desmin, HMB45 and epithelial membrane antigen.

Ultrastructural evaluation of the MFH revealed storiform–pleomorphic subtype. The section consisted of spin-
<table>
<thead>
<tr>
<th>No.</th>
<th>Author &amp; year</th>
<th>Case</th>
<th>Sex</th>
<th>Age (Year)</th>
<th>Site of lesion</th>
<th>Treat</th>
<th>Rec</th>
<th>Met</th>
<th>Follow up</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Watanabe (2005)</td>
<td>1 case Pr/MFH</td>
<td>M</td>
<td>83</td>
<td>Right maxilla</td>
<td>C+ Bounding</td>
<td>NM</td>
<td>NM</td>
<td>13 months</td>
<td>DC</td>
</tr>
<tr>
<td>2.</td>
<td>Sabesan et al. (2005)</td>
<td>15 cases Pr/MFH</td>
<td>NS</td>
<td>Mean age 43 (13–83)</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>17 months</td>
<td>AO</td>
</tr>
<tr>
<td>3.</td>
<td>Chan et al. (2004)</td>
<td>1 case Pr/MFH</td>
<td>F</td>
<td>44</td>
<td>Left maxillary molar region</td>
<td>S + R</td>
<td>–</td>
<td>–</td>
<td>8 months</td>
<td>AO</td>
</tr>
<tr>
<td>4.</td>
<td>Yamaguchi et al. (2003)</td>
<td>1. Pr/MFH</td>
<td>M</td>
<td>29</td>
<td>Max sinus</td>
<td>S + C</td>
<td>+</td>
<td>–</td>
<td>1 year</td>
<td>DC</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. Pr/MFH</td>
<td>M</td>
<td>56</td>
<td>Maxilla</td>
<td>S + C + R</td>
<td>–</td>
<td>–</td>
<td>11 years, 8 months</td>
<td>AO</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. Pr/MFH</td>
<td>M</td>
<td>57</td>
<td>Max sinus</td>
<td>S</td>
<td>+</td>
<td>L</td>
<td>9 months</td>
<td>DC</td>
</tr>
<tr>
<td></td>
<td></td>
<td>4. Pr/MFH</td>
<td>M</td>
<td>63</td>
<td>Max sinus</td>
<td>S + C + R</td>
<td>–</td>
<td>–</td>
<td>5 years</td>
<td>AO</td>
</tr>
<tr>
<td>5.</td>
<td>Sato et al. (2001)</td>
<td>1. Pr/MFH</td>
<td>F</td>
<td>48</td>
<td>Left maxillary premolar regions</td>
<td>S</td>
<td>+</td>
<td>+/Lung +/Skin bone lung</td>
<td>2 years</td>
<td>DC</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. Pr/MFH</td>
<td>F</td>
<td>47</td>
<td>Right maxillary premolar regions</td>
<td>S + C</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6.</td>
<td>Mardinger et al. (2001)</td>
<td>1 case Pr/MFH/high grade</td>
<td>F</td>
<td>32</td>
<td>Post-maxillary and alveolar ridge</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>77 months</td>
<td>AO</td>
</tr>
<tr>
<td>7.</td>
<td>Pandy et al. (2000)</td>
<td>1 case Pr/MFH</td>
<td>M</td>
<td>54</td>
<td>Zygoma + Maxilla</td>
<td>S(WE) + R</td>
<td>R after 18 months</td>
<td>NM</td>
<td>8 months</td>
<td>DC</td>
</tr>
<tr>
<td>8.</td>
<td>Amante (1997)</td>
<td>1 case Pr/MFH</td>
<td>F</td>
<td>40</td>
<td>Maxilla</td>
<td>S + C</td>
<td>NM</td>
<td>NM</td>
<td>10 months</td>
<td>DC</td>
</tr>
<tr>
<td>9.</td>
<td>Li et al. (1997)</td>
<td>1 case/after radiation</td>
<td>M</td>
<td>47</td>
<td>Maxillary sinus</td>
<td>S + R</td>
<td>+</td>
<td>NM</td>
<td>AC/Deteriorate condition AC/Very poor after 17 month survive</td>
<td></td>
</tr>
<tr>
<td>10.</td>
<td>Lin et al. (1994)</td>
<td>2 cases/after radiation</td>
<td>NM</td>
<td>NM</td>
<td>Maxilla</td>
<td>NM</td>
<td>NM</td>
<td>17 months</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11.</td>
<td>Besly et al. (1993)</td>
<td>2 case s/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>12.</td>
<td>Wang (1993)</td>
<td>1 case/Pr/MFH</td>
<td>F</td>
<td>19</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>13.</td>
<td>Paume et al. (1993)</td>
<td>1 case/an anaplastic sarcomatous zone of the malignant fibrous histiocytoma (MFH) type</td>
<td>F</td>
<td>19</td>
<td>Maxilla</td>
<td>S</td>
<td>R</td>
<td>L</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>14.</td>
<td>Namyslowski et al. (1993)</td>
<td>1 case/Pr/MFH</td>
<td>F</td>
<td>71</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>15.</td>
<td>Takahashi and Sato (1991)</td>
<td>1 case/Pr/MFH</td>
<td>F</td>
<td>45</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>16.</td>
<td>Ireland et al. (1988)</td>
<td>2 cases/Pr/MFH after radiation</td>
<td>NM</td>
<td>NM</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>17.</td>
<td>Min (1988)</td>
<td>1 case/Pr/MFH (article in Korean language)</td>
<td>F</td>
<td>71</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>18.</td>
<td>Fan et al. (1986)</td>
<td>7 cases/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>Maxilla</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>19.</td>
<td>Tagawa et al. (1986)</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>Right maxilla</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
<tr>
<td>20.</td>
<td>Block et al. (1986)</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>Maxillary sinus</td>
<td>S</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td></td>
</tr>
</tbody>
</table>
| 21. | Karcher et al. (1986) | 1 case/Pr/MFH | NM | NM | Maxilla | S | NM | NM | NM | (continued on next page)
dle shaped and giant fibroblastic cells with markedly pleomorphic multi-segmented nuclei, well developed branching rough endoplasmic reticulum contained a finely granular substance and more electron dense material. Primarily peripheral bundles of actin microfilaments with interspersed fusiform densities were also seen. Subplasmalemmal attachment plaques, focal basal lamina-like material, and micropinocytotic vesicles were present to variable degrees in these cells. Fibroblasts transformed into a contractile myofibroblast were well identified by exposed peripheral arrays of actin microfilaments. The intermediate filaments were found in a plenty amount and observed in the cytoplasm of spindle fibroblast-like cells joined by rudimentary cell junction (Fig. 6A–D).

Discussion

"As reflected in the World Health Organization classification of soft tissue tumors, so called malignant fibrous histiocytoma can no longer be regarded as a definable entity, and is now viewed as a synonym for undifferentiated pleomorphic sarcoma. Malignant Fibrous histiocytoma is the most common soft tissue sarcoma, first described by Ozzelo et al. and O'Brien and Stout. The storiform-pleomorphic type is the most common and is a highly cellular tumor, which can range from well differentiated to anaplastic. MFH affects individuals later in life and occurs more often in men, with an approximate 2:1 male:female ratio.

Maxillary MFH is very rare. It was first reported in 1974. The review of the literature produced 61 well-documented cases of maxillary malignant fibrous histiocytoma (Table 1) ranging in age from 12 to 83 years, with a median age of 44.7 years. Most patients 54/61 (88.5%) showed clinical signs of primary maxillary MFH. MFH can also arise in the site of previous radiation. However, radiation induced MFH of the head and neck is exceedingly rare. A literature search showed only 7/61 cases (11.4%) of post-radiation maxilla MFH. The sinonasal tract has been reported to be the commonest site (30%) of tumor involvement in the

Table 1

<table>
<thead>
<tr>
<th>No.</th>
<th>Author &amp; year Case</th>
<th>Site of lesion</th>
<th>Age (Year)</th>
<th>Sex</th>
<th>Site of treatment</th>
<th>Rec.</th>
<th>Treat</th>
<th>Met</th>
<th>Follow-up</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>20.</td>
<td>Shikimori and Oka (1985)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M &amp; F</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>1 year</td>
<td>DC</td>
</tr>
<tr>
<td>21.</td>
<td>Abdul-Karim et al. (1985)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M &amp; F</td>
<td>1 case/Pr/MFH and 1 case/post-irradiation</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>3–13 months</td>
<td>NM</td>
</tr>
<tr>
<td>22.</td>
<td>Hayter et al. (1985)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>12 months</td>
<td>NM</td>
</tr>
<tr>
<td>23.</td>
<td>Nishizawa et al. (1985)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M</td>
<td>1 case/Pr/MFH/after radiation</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>3 months</td>
<td>NM</td>
</tr>
<tr>
<td>24.</td>
<td>Kessler et al. (1981)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>3 months</td>
<td>NM</td>
</tr>
<tr>
<td>25.</td>
<td>Sonobe et al. (1980)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>3 months</td>
<td>NM</td>
</tr>
<tr>
<td>26.</td>
<td>Sidhu et al. (1978)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>3 months</td>
<td>NM</td>
</tr>
<tr>
<td>27.</td>
<td>Slootweg and Muller (1977)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>3 months</td>
<td>NM</td>
</tr>
<tr>
<td>28.</td>
<td>Spector and Ogura (1974)</td>
<td>Maxilla</td>
<td>12 to 75 years</td>
<td>M</td>
<td>1 case/Pr/MFH</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>3 months</td>
<td>NM</td>
</tr>
</tbody>
</table>

Figure 1 Photograph shows the clinical appearance of a huge exophytic growth measuring 7 cm × 6 cm over the left tuberosity of maxilla extending from the junction between soft and hard palate towards premolar area of edentulous ridge.
head and neck region. But Sabesan et al. found that maxilla is the most common site (15/54 cases). This literature review of Maxilla MFH revealed that 41/61 cases (67.2%) of the lesions were located in maxilla while, only 5/61 cases (8.19%) in maxillary sinus, and only two cases (3.2%) reported in post-maxillary and alveolar ridge and in zygoma respectively.

Our patient showed painful, easily bleed swelling at left tuberosity of maxilla. The lesion appeared as primary huge exophytic growth measuring 7 cm × 6 cm over the left tuberosity of maxilla extending from the junction between soft and hard palate towards premolar area of edentulous ridge. The growth extended laterally from the vestibule to the midline. This case presented uncommon location has not reported previously in literature.

Few reports have dealt with the radiographic findings of MFH. Reported radiographic findings were irregular bone margin, a moth-eaten appearance, erosion of cortex, pathological fracture, and tooth root resorption. Although, these radiographical findings are not specific to MFH, and are usually also observed in squamous cell carcinoma, the most common malignant tumor of the head and neck. Radiographical evaluation of two cases MFH affecting maxillary alveolar bone had been described by Sato et al. They represented in details the radiographical findings, which have seldom been described in previous reports. They reported the following findings, the presence of fairly well demarcated bone destruction in the intraoral radiograph, the relatively smooth surface, uniform density, or no necrotic area of the tumor. In computed tomography images, tumor showed clear separation of the tumor from surrounding soft tissues bone scintigraphs reflecting the periosteal reaction to tumor invasion and lymphoscintigraphy of the metastatic lymph nodes. Generally MFH of the maxilla dose not differ from tumors arising in flat bones in other parts of the skeleton. In our case, CT scan revealed an ill defined soft tissue mass with heterogeneous enhancement noted in the left palatine region with central non-enhancement suggestive of central necrosis. These findings are different from the finding described previously.

Most maxilla MFH exhibited a broad range of histological patterns. Information regarding histological type was available in cases reviewed from literature. Storiform-pleomorphic was the prominent type in most cases. However, only one case

Figure 2  (A) and (B): computed tomography revealed an ill defined soft tissue mass with heterogeneous enhancement noted in the left palatine region with central non-enhancement suggestive of central necrosis.

Figure 3  Photograph shows surgical location 10 days post-operation.

Figure 4  Photomicrographs reveal histological features of malignant fibrous histiocytoma: (A) low power magnification of histopathology showing a markedly cellular spindle-cell neoplasm with a diffuse growth pattern reveals multiple disseminated nodular cellular masses (×100) and (B) high-power magnification showing nuclear pleomorphism and mitotic activity. Note the histiocyte (arrow) with an indented nucleus and abundant cytoplasm (×200).
showed Pr/MFH/High grade. Our patient showed typical a storiform pattern, contained interlacing a mix of pleomorphic fibroblasts and multi-nucleated cells arranged in fascicles and myxoid pattern. The cells revealed abnormal high mitotic index. We suggested high grade of storiform MFH. Nevertheless, the term MFH is widely used in routine pathological practice for pleomorphic soft tissue sarcoma without specific line of differentiation. MFH is included in the category of fibrohistiocytic tumors. The presence of a large number of histiocyte-like or histiocytic cells in the pleomorphic sarcoma raised the question of biphasic (fibroblastic–myofibroblastic and histiocytic) differentiation.

The trend for some pathologists to diagnose malignant fibrous histiocytoma (MFH) less frequently than others may result from different diagnostic criteria for MFH among pathologists, reflecting the concept of MFH as a common morphological manifestation of a variety of poorly differentiated sarcomas, and the diagnosis of MFH through a process of exclusion. It was concluded that a re-evaluation of the diagnostic criteria was essential for so called MFH. "Ultrastructural observation of neoplastic cells suggests that the expression of smooth muscle markers in so called malignant fibrous histiocytoma is a result of myofibroblastic differentiation". Smooth muscle markers, such as smooth muscle actin (SMA) and occasionally even desmin, have

Figure 5  Photomicrographs of immunohistochemically, most of the tumor cells were strongly positive for vimentin: (A) low power magnification (×200) and (B) high-power magnification (×400).

Figure 6  Transmission electron microscopy photomicrographs, revealed storiform and pleomorphic MFH: (A) large fibroblastic tumor cell with prominent periphery filaments with pleomorphic nucleus, (B) histiocyte revealed multi-segmented nucleus and dilated RER, (C) giant cell tumor revealed significant accumulation of vimentin intermediate filaments and (D) histiocytes revealed giant nucleus with RER containing a finely granular substance and a more electron dense material.
been demonstrated immunohistochemically in a subset of "MFH". In this situation, the distinction between leiomyosarcoma and "MFH" becomes more difficult. Ultrastructural observation of neoplastic cells suggests that the expression of smooth muscle markers in "MFH" is a result of myofibroblastic differentiation.57–59 There are limited data on the frequency and degree of positively for smooth muscle markers in a large number of "MFHs".

A cases reported in literature demonstrated that most cases were positive for vimentin and only few cases were positive for cytokeratin and desmin. In our case tumor was negative for epithelial markers, protein S-100 (a marker of melanoma and schwann cell tumor) and desmin (smooth muscle markers). But it was strong positive for vimentin (mesenchymal markers). More recent histochemical, immunohistochemical, and ultra structural studies of MFH support the contention that MFH is a form of fibrosarcoma and the tumor is very likely over diagnosed. This is because the more pleomorphic the tumor the more difficult is to distinguish from other types of sarcomas, such as pleomorphic leiomyosarcoma, pleomorphic liposarcoma and dedifferentiated liposarcoma. Distinction among these pleomorphic soft tissue tumors is best achieved by a joint immunohistochemical and ultrastructural study.2 Commonly, ultra structural findings of MFH have demonstrated that tumor cells were composed of fibroblast-like cells with abundant RER, histiocyte-like cells with numerous lysosomes, and transitional cells with characteristics of both the fibroblast-like and histiocyte-like cell.50 Our present electron microscopic observations compatible with typical ultra structural findings reported previously. It revealed spindly or stellate fibroblast with long thin cytoplasmic processes as a cellular microfilaments with interspersed fusiform densities. In response to inflammation and injury, the fibroblast often transforms into contractile myofibroblast with peripheral arrays of actin microfilaments and/or a fibrohistocytes with prominent lysosome that is often mistaken for a true histocytes. Although the histocytes in MFH are though by some to be neoplastic, the origin of both histiocyte-like cells and multi-nucleated giant cells in MFH has been the subject of much debate.51

Most of the cases 54/61 (88.5%) reported in literature review were treated surgically. In 4/61 (6.5%) cases, surgery was followed by post-operative radiotherapy and in other 4/61 (6.5%) cases was followed by post-operative chemotherapy. While in 3/61 (4.9%) cases radiotherapy and chemotherapy had been used. Only one case was treated with chemotherapy and boun ding, which resulted in a large reduction tumor size without surgery.12 The proportion of local recurrence rate of MFH after initial local excision ranges between 16% and 52%,62,53 The presence of positive surgical margins after definitive treatment is the single most important factor relating to local recurrence.52 According to Barnes and Kanbour,64 80% of patients with local recurrences after incomplete surgical treatment subsequently die from disease. Recurrence is related to size, depth of invasion, and microscopically positive surgical margins. From the review of literature we found 9/61 (14.7%) recurrent cases and 3/61 (4.9%) non-recurrent cases while other cases, they did not mentioned about the recurrence. In 6/61 (9.8%) cases of maxilla MFH reported metastasized distantly to lung, skin, local, bone, pleurae, pancreas, kidneys and bone marrows,5,23,33,37 Only 6/61 (9.8%) cases survived with out disease while all other case died with disease or lived with deteriorates condition. In our case the patient came 4 months later with recurrence lesion confirmed histopathologically.

In conclusion, we believe that MFH of the maxilla may have poorer prognosis than those tumors in the other parts of the maxillofacial skeleton. Size, depth, histopathologic features, immunohistochemistry and ultrastructural features, surgical margin status and adjuvant radiotherapy or chemotherapy are the most important predictors of outcome.

References

2. Erlandson RA, Woodruff JM. Role of electron microscopy in the evaluation of soft tissue neoplasms, with emphasis on spindle cell and pleomorphic tumors. Human pathology 1998;29(12).


