

## Case Report

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# Malignant Fibrous Histiocytoma of the Maxillary Sinus Presenting as Toothache

Malignant fibrous histiocytoma (MFH) is a high-grade and aggressive sarcoma. It is relatively rare in the head and neck region. Its diagnosis is based on immunohistochemical stains. Wide excision followed by postoperative radiotherapy is believed to be the treatment of choice for MFH. In October 2001, a case of MFH in the maxillary sinus, which presented as a toothache at the beginning, was successfully diagnosed and treated. Using the external approach, the tumor mass was completely removed, and postoperative radiotherapy was subsequently performed. Seventeen months after the surgery, the patient was clinically well without any evidence of local recurrence or distant metastasis.

### Key Words

immunohistochemical stain;  
malignant fibrous histiocytoma;  
maxillary sinus

**M**alignant fibrous histiocytoma (MFH) is a high-grade and aggressive sarcoma.<sup>1-3</sup> Its diagnosis is based on immunohistochemical stains.<sup>3</sup> Wide excision followed by postoperative radiotherapy is believed to be the treatment of choice for MFH.<sup>1-3</sup> The case we report here is one of MFH arising from the left maxillary sinus. Persistent toothache or non-healing extraction sites should warn the clinician to note that there might be a serious underlying condition, such as a tumor.

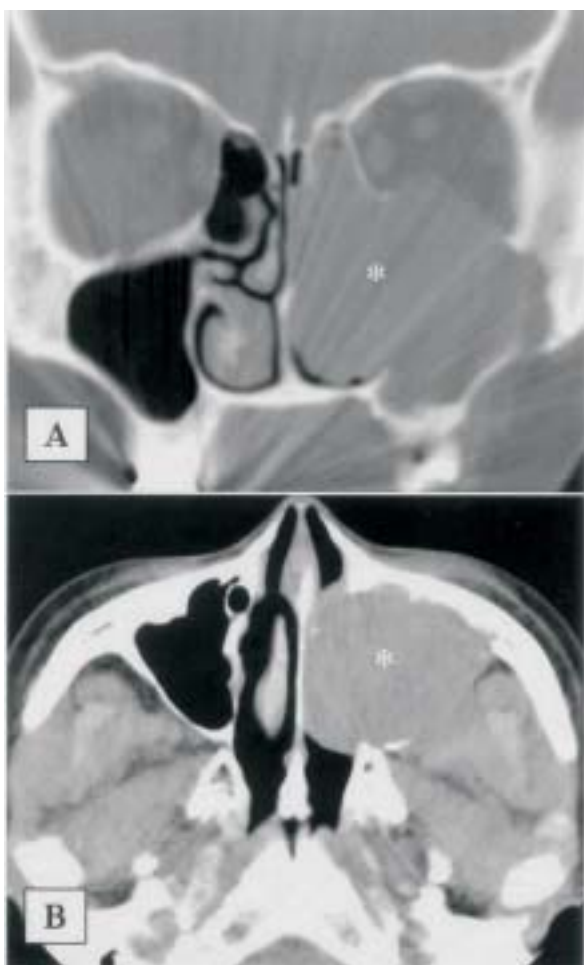
### CASE REPORT

In October 2001, a 44-year-old woman presented with a two-month history of a painful swelling over the left maxillary molar region following extraction of the left upper third molar tooth for a progressive toothache. A clinical sinuscope examination revealed bulging of the medial wall of the left maxillary sinus near the septum. There were neither regional lymphadenopathies nor limitation of eye movement. Computed tomographic (CT) scans demonstrated gross bony destruction of the medial, lat-

eral, posterior and superior walls of the maxillary sinus, and an enhanced soft tissue mass that occupied the entire left maxillary sinus and extended to the left ethmoid sinus, nasal cavity and orbital floor. The tumor mass had a relatively smooth surface without necrotic areas and was easy to distinguish from the surrounding soft tissues (Fig. 1). A low-grade malignancy arising from the left maxillary sinus was highly suspected. Using the mid-face degloving approach, we removed the tumor mass completely, without orbital exenteration. Microscopically, the mass contained spindle cells arranged in a storiform pattern. Histiocyte-like cells interspersed between the spindle cells and mitotic activity were easily noted (Figs. 2A and 2B). Immunohistochemical stains (Fig. 3) confirmed the diagnosis of malignant fibrous histiocytoma; they were focally positive for alpha-1 antitrypsin, CD117 (KIT), CD68 (KP1) and HHF-35 (muscle-specific actin), but negative for CD34 (QBEND/10), keratin and S-100 protein. The patient recovered well and received postoperative radiotherapy with a total dose of 6000 cGy. Seventeen months after surgery, this patient was well clinically without any evidence of local recurrence or distant metastasis.

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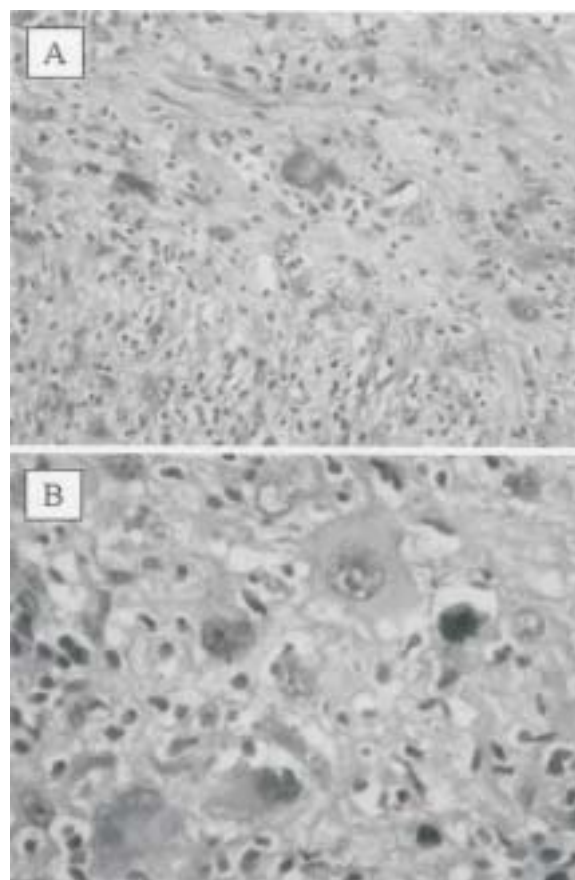
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**Fig. 1.** CT scans of the paranasal sinuses showing bony destruction of the medial, lateral, posterior, and superior walls of the maxillary sinus, and a relatively smooth, homogeneous, enhanced soft tissue mass (asterisk) that occupied the entire left maxillary sinus and extended to the left ethmoid sinus, nasal cavity, and orbital floor. (A) coronal view under soft tissue window; (B) axial view under bone window.

## DISCUSSION

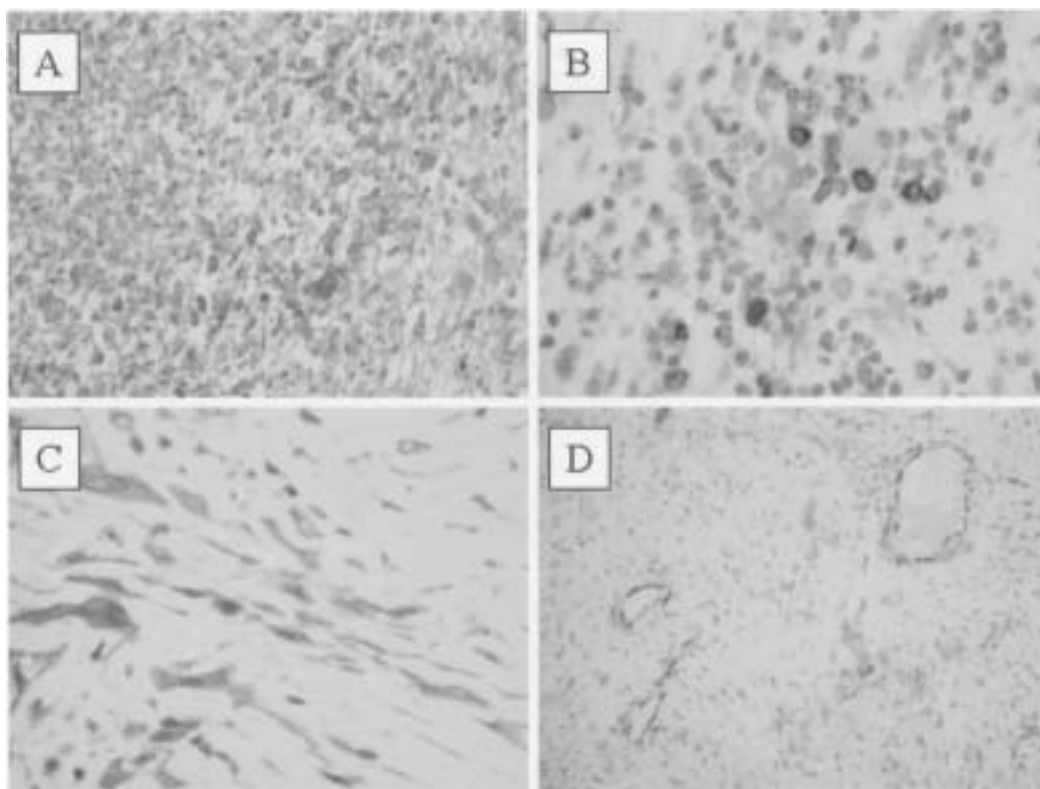
Malignant fibrous histiocytoma (MFH) is a high-grade, pleomorphic and aggressive soft tissue sarcoma, which was first described by O'Brien and Stout in 1964.<sup>4</sup> The majority of its investigators have suspected primitive mesenchymal cells, such as fibroblasts and histiocyte-like cells, to be the origin of these tumors. Microscopically, MFH can be classified into 5 types: storiform-pleomorphic, giant cell, inflammatory, angiomatoid and myxoid.<sup>1-3</sup> Storiform-pleomorphic and myxoid variants are the most common types. This case was proven as a storiform type. MFH is usually detected in patients be-



**Fig. 2.** Histopathology of the tumor. (A) spindle cells with a storiform pattern and an admixture of histiocyte-like cells with prominent pleomorphism, hyperchromatism and frequent mitotic figures (H & E,  $\times 150$ ); (B) bizarre multinucleated giant cells shown in higher magnification (H & E,  $\times 300$ ).

tween the ages of 50 and 70 years. An exception is the angiomatoid variant that usually affects individuals who are younger than 20 years old.<sup>1-3</sup> There is a higher incidence of MFH for males. No etiological factors have been identified for MFH, but radiation exposure could play an important role.<sup>5,6</sup>

MFH typically arises in soft tissues, especially in the extremities and the trunk.<sup>1-3</sup> 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.<sup>1-3</sup> 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.<sup>7</sup> Although toothache is less frequently present in cases with maxillary sinus tumors, persistent toothache or non-healing extraction sites, as in our case, should warn the clinician that



**Fig. 3.** Immunohistochemical stains. (A) positive for alpha-1 antitrypsin; (B) positive for CD68 (KP1); (C) positive for CD117 (KIT); (D) weak positive HHF-35 (muscle-specific actin).

there might be a serous underlying condition, such as a tumor.<sup>7</sup>

MFH occurring in the maxillary sinus is very rare. To date, there have been approximately 23 cases reported.<sup>1,2,8-11</sup> They were 13 men and 10 women, ranging in age from 10 to 79 years, with a median age of 47.7 years. Thirteen lesions were classified as the storiform pattern, 2 as storiform-pleomorphic, 2 as myxoid, 1 as pleomorphic and 5 not reported. Their initial clinical presentations suggested the typical features of the maxillary sinus disease: swelling of the cheek (14 of 23), facial pain (12 of 23), and nasal obstruction or discharge (7 of 23). Only 3 cases (13%) presented with a toothache. The relatively rare symptoms included infraorbital nerve paresthesia, visual disturbance, epistaxis, proptosis, delayed healing of an extraction wound and difficult chewing. Treatment included surgical ablation followed by postoperative radiation therapy (6 of 23), surgical ablation only (4 of 23), surgical ablation combined with radiation therapy and chemotherapy (6 of 23), preoperative radiation therapy followed by surgical ablation (3 of 23), radiation therapy

only (2 of 23) and radiation therapy combined with chemotherapy (2 of 23). Distant metastases, occurring in 7 patients, included 3 to the lung, 1 to the mandible, 1 to the lumbar spine, 1 to the cerebral cortex and 1 to both the calvarium and the lumbar spine. The follow-up periods ranged from 3 to 54 months. Only 8 patients (34.8%) remained free of disease more than 1 year after diagnosis. Moreover, 1 patient died immediately 3 months after diagnosis because of local recurrence and distant metastases. Due to the lack of documented cases, it is difficult to evaluate survival rates. This case is an additional 1; even though the tumor presented as an unusual “toothache” symptom, it was successfully diagnosed and treated with adequate surgical exposure and resection followed by full dose of radiation therapy. Till now, this patient still remains healthy and free of disease.

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.<sup>11</sup> In our case, however, the findings of the 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.<sup>11</sup>

The diagnosis of MFH is based upon the pathologic features characterized by an admixture of fibroblastic and histiocytic-like cells in a storiform pattern.<sup>1-3</sup> 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.<sup>1-3</sup> Fibrous histiocytoma is typically immunoreactive for vimentin (V9), and sometimes for smooth muscle actin (HHF35) or alpha-1 antitrypsin, but not for desmin, keratin, epithelial membrane antigen, S-100 protein, factor VIII-related antigen, CD34, nor carcino-embryonic antigen, supporting the hypothesis that the tumor cells are of mesenchymal origin. CD68 (KP1) is a monoclonal antibody to a lysosomal component and is considered to be highly specific for histiocytes; however, the application of anti-CD68 in MFH has revealed conflicting results.<sup>3</sup> CD117 (KIT) is a transmembrane, tyrosine kinase growth factor receptor that is expressed on numerous diverse fetal and adult cells, including hematopoietic cells, mast cells, melanocytes, germ cells and the interstitial cells of Cajal.<sup>12</sup>

Surgical excision of MFH in the head and neck region remains the definitive treatment.<sup>1-11</sup> The mid-face degloving approach, undertaken in our case, might be anticipated given its excellent exposure of the nasal cavity, the middle-third of the face, and the central skull base combined with its outstanding cosmetic results.<sup>13</sup> Prophylactic neck dissection is not recommended because the percentage of cases with occult metastasis is low.<sup>1-11</sup> Based upon the difficulties of adequate resection and the high incidence of local recurrence, postoperative radiotherapy is advocated. The primary tumor factors associated with a worse prognosis are necrosis, a high mitotic count, histiologically undifferentiated tumors, and blood vessel invasion.<sup>2,3</sup> 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.<sup>2,3</sup>

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