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Case Report

Myxoid Type of Malignant Fibrous Histiocytoma of the Maxillary Sinus: A Case Report

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Abstract

Introduction:
Myxofibrosarcoma was originally described as the myxoid variant of Malignant Fibrous Histiocytoma (MFH), a high-grade and aggressive sarcoma, which is very uncommon in the head and neck region, with about 100 cases reported up to now. MFH occurring in the maxillary sinus is so rare that only 23 cases have been reported. We hereby report a case of myxofibrosarcoma in the maxillary sinus.

Case Report:
The case was a 54-year-old male with symptoms of toothache in the right posterior maxillary teeth, a swelling adjacent to maxillary molar region and symptoms of chronic maxillary sinusitis. In clinical examination, the teeth were sensitive to percussion and palpation, but no caries and restoration was detected on his molar teeth. He was suffering from local pain and tenderness over his midface and mild fever, fatigue and some nonspecific vague pain. CT scan showed a mass lesion involving right nasal cavity, maxillary and ethmoidal sinuses. A low-grade malignancy arising from the right maxillary sinus was highly suspected. A surgery was done to remove the mass. The histological and immunohistochemical studies proved the diagnosis of myxoid variant of Malignant Fibrous Histiocytoma. Therefore radiotherapy and chemotherapy was started for the patient but six months later the symptoms returned and CT scan showed a right maxillary and ethmoidal mass that extended to base of the skull. Maxillectomy and ethmoidectomy were performed for the patient, but 2 months later he died because of the extension of the tumor, which confirmed the necessity of early diagnosis.

Conclusion:
Amplified radical surgery is the first choice of treatment. The second surgery has special value to the recurrent patients. Radiotherapy alone or chemotherapy alone is not effective to MFH of head and neck region

Keywords:
Immunohistochemical, Malignant fibrous histiocytoma, Paranasal sinuses

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Introduction

Malignant fibrous histiocytoma (MFH) is a common soft tissue tumor in adults occurring in the 5th to 7th decades of life and accounts for 10.5% to 21.6% of all soft tissue malignant neoplasms (1). It is a high-grade and aggressive sarcomatous neoplastic disease with complex pathological structure. Its histological origin is uncertain (2,3,4) and its diagnosis is based on immunohistochemical staining (5). It is very rare in the head and neck region, with about 100 cases reported up to now (6). Moreover, it is one of the least common tumors occurring in maxillary sinus. In addition, myxoid variant is the rare subtype of MFH category and is recognized as myxofibrosarcoma (5).

This case report presents a patient with myxoid type of Malignant Fibrous Histiocytoma of the maxillary sinus region presenting as a rapidly growing tumor.

Case Report

A 54-year-old male was admitted in May 2005, who was suffering from severe toothache in right posterior maxillary teeth and aggravated symptoms of chronic maxillary sinusitis of two to three months duration before his admission. On clinical examination a swelling was seen in right maxillary molar region, the teeth were sensitive to percussion and palpation, but no caries and restoration was detected on his molar teeth and no pathosis could be `local pain and tenderness over his midface and mild fever, fatigue and some nonspecific vague pain. In physical examination the patient was pale and anemic. His CT scan and MRI showed a large soft tissue mass in right maxillary sinus protruded to the right nasal cavity and ethmoidal sinus. The destruction of medial wall of right maxillary sinus and extension to nasopharynx was seen as well (Fig 1). A surgery was done to remove the mass. The mass size was 7×5 cm, cream color, globular, with firm and slippery (mucoid) consistency, fixed to the underlying bone, destructive behavior with the same involvement as previously showed in CT scan.

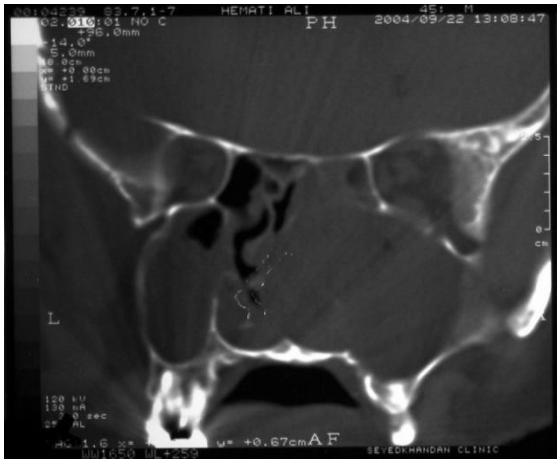

Fig 1: CT scans of the paranasal sinuses showing a large soft tissue density in right maxillary sinus protrude to right nasal cavity and destruction of medial wall of right maxillary sinus and extention to nasopharynx is seen.

The tumor was excised with a rim of uninvolved bone all around. It was very vascular and infiltrating to the adjacent structures. Microscopic examination showed the tumor to be poorly encapsulated. There were prominent plump spindle cells in a myxoid background with a focal storiform pattern. Marked nuclear pleomorphism and atypia with a few multinucleated giant cells were seen. There were areas of necrosis were present with no lipoblasts or cells with striations. Inflammatory cells such as lymphocytes and neutrophils were present focally in a myxoid stroma. The combination of pleomorphism and spindled cells with focal storiform pattern in myxoid stroma suggested a diagnosis of malignant fibrous histiocytoma of myxoid type or "myxofibrosarcom" (Fig 2).

Immunohistochemical staining was positive for vimentin, CD68 (Fig 3,4), and negative for S100, desmin, cytokeratin, CD34, smooth muscle actin.

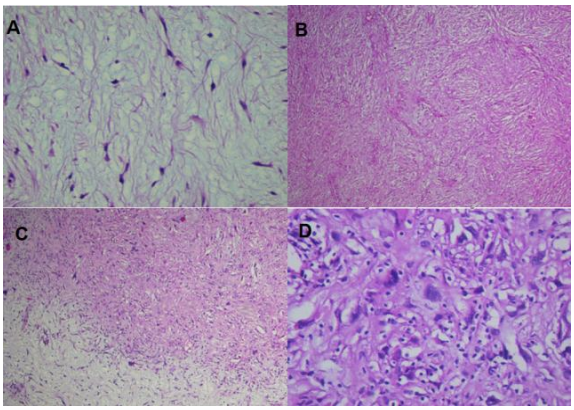

Fig 2: MFH A: Mixoid back ground (H&E ×400), B: Fascicular pattern (H&E ×100), C: Fascicular pattern and myxoid pattern (H&E ×100), D: Marked pleomorphism (H&E ×400)

The patient recovered well and received postoperative radiotherapy with a total dose of 6000 cGy and chemotherapy with Doxorubicin and Dacarbazin for 3 cycles. Inspite of the fulfilled treatment, six months later the symptoms returned and CT scan showed a right maxillary and ethmoidal mass that extended to the base of the skull .The maxilectomy and ethmoidectomy were performed for the patient, the size of tumor was 3×2×1/5 cm. The pathological diagnosis was myxoid MFH, extending to the base of skull and was very progressive. In spite of the fulfilled treatment for the patient and the performed surgery, he died 2 months later. This may be indicative of the aggressiveness of this tumor and suggests the fact that early diagnosis is essential.

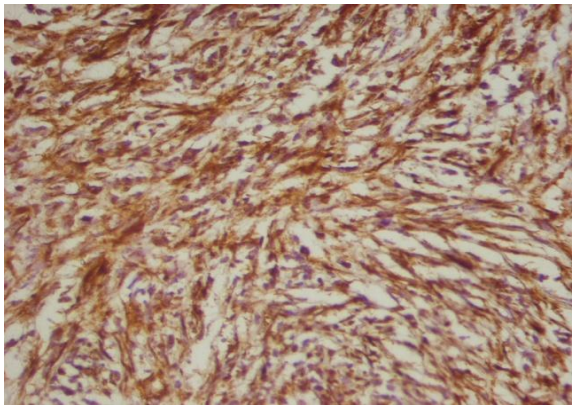

Fig 3: Immunohistochemxical positive reactivity for vimentin × 100

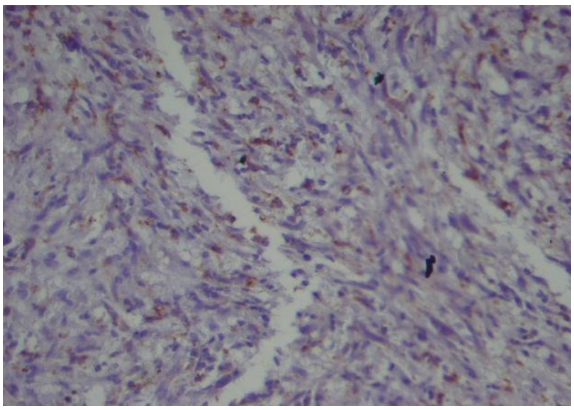

Fig 4: Positive immunoreactivity for CD68 ×400

Discussion

Malignant Fibrous Histiocytoma, in locations except head and neck, is a common soft tissue sarcoma in adults, and the most common sites are the retroperitoneum and the deep soft tissue of the extremities. Only less than 3% arise in the head and neck region (6). MFH generally arises during the fifth to seventh decades of life. The presented case was in the 5th one.

In the head and neck, the most common sites are the sinonasal tract, larynx, and soft tissues of the neck (7). Tumors of the sinonasal tract present symptoms of nasal obstruction and epistaxis, whereas tumors of the larynx cause hoarseness and dysphagia (7,8). The presented case was also in the maxillary, ethmoidal sinus, one of the most common sites, and suffering from signs of sinusitis and nasal obstructions. The sensitivity of posterior maxillary teeth to percussion and palpation was due to tumor extension in maxillary sinus and sinusitis symptoms.

MFH has five histological sub types: storiform- pleomorphic, myxoid, giant cell, angiomatoid and inflammatory; however, only the first two variant have been reported to arise in the head and neck. Storiform-pleomorphic is the most prevalent histological subtype.

Myxoid MFH or myxofibrosarcoma is a variant of MFH that most commonly occurs in the extremities of elderly

individuals, but also may arise in the head and neck region (6). The mean age at diagnosis is 60 years. The presented case was a myxoid subtype of MFH in the head region and the patient age was 57, which is almost the same as what was mentioned in previous articles. These tumors may be superficial or deep, and gross inspection reveals a gelatinous mass with well-defined or infiltrative borders. Microscopically, hypo- to moderately cellular nodules of spindle- and stellate-shaped cells in a myxoid background are separated by fibrous septae. The cell cytoplasm is wispy and ill defined and the nuclei irregularly shaped and are hyper chromatic. Some cells may contain cytoplasmic vacuoles that scallop the nuclei causing these cells to resemble lipoblasts. The vacuoles, however, contain nonsecreted myxoid matrix and are not optically clear as in true lipoblasts. Tumor cells tend to condense around elongated capillaries that course throughout the stroma. High grade lesions contain more cellular areas that resemble standard fibrosarcoma or MFH with fascicular growth pattern, abundant mitoses and necrosis (8). In this case there were prominent plump spindle cells with focal storiform pattern and nuclear pleomorphism and atypia as well. Despite the fact that presence of the cells resembling lipoblasts is a common findng in myxoid subtype, there was no lipoblasts or cells with striations.

Immunohistochemical stains performed on MFH show positivity for vimentin, and histiocyte associated markers like CD68. Both of these immunohistochemical markers were positive for the presented case, which confirmed the diagnosis. Myofibroblastic differentiation also may be demonstrated by staining with actin and desmin; and keratin staining has been described in 25 percents of cases (3). This case was immunohistochemically negative for desmin and actin. The differential diagnosis is the same as other pleomorphic sarcomas, principally liposarcoma, malignant peripheral nerve sheet tumor (MPNST) and leiomyosarcoma. Although cells within an MFH may be vacuolated, true lipoblasts are not present. MPNSTs generally lack the pleomorphism and storiform growth pattern present in MFH, and immunohistochemical positivity for S-100 protein or lEU 7 favors the diagnosis of MPNST over MFH.

Lytic lesion with areas of calcification while tomograms reveal subtle permeative radiolucent changes extending beyond the poorly demarcated central area of radiolucency tumor (7).

MFH occurring in the maxillary sinus is very rare (there have been approximately 23 cases reported) (5). There 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.

The diagnosis of MFH is based upon the pathologic features characterized by an admixture of fibroblastic and histiocytic-like cells in a storiform pattern (1,5,9). However, the histological 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 (1,5). 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 (10,11,12).

Radiographic findings of malignant fibrous histiocytoma (MFH) are helpful for differentiation of MFH from other malignant tumors of the head and neck, although MFH is a rare disease and there are no radiographic findings that would indicate a specific diagnosis of MFH (5).

The treatment for MFH has been described as a combination of radical excision, radiotherapy and chemotherapy. Amplified radical surgery is the first choice of treatment. The second surgery has special value to the recurrent patients. Radiotherapy alone or chemotherapy alone is not effective to MFH of head and neck region (2).

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