Myxofibrosarcoma of Right Fronto-Temporal Region With Intracranial Extension: A case report

P Shukla, D Gupta, P Gupta, S Singh Bisht, M Pant, S Gupta, N Husain, V Joshi

Citation

Abstract
Myxofibrosarcoma, is a myxoid variant of Malignant Fibrous Histiocytoma (MFH) first described by Weiss & Enzinger (1) with a better prognosis. MFH rarely occurs in the skull, with only 17 cases documented in literature (2). The tumor frequently erodes the outer table of the skull and spreads in the extracranial space. Intradural and brain invasion are very rare. We report a case of myxofibrosarcoma originating in the right fronto-temporal region with subsequent intracranial extension.

CASE REPORT
A 27 year old male presented with decreased vision in the right eye lasting for the past two years, swelling and bulging of the right eye, and severe pain for the past five months. He had a history of trauma to the right frontal area about seven months prior to the consultation. On examination, there was proptosis in the right eye. The conjunctiva was dry and swollen and there was corneal opacity. Neurological examination showed absence of vision in the right eye. The radiological examination of the chest was normal. A CT scan showed a heterogeneous enhancing space occupying lesion with calcification in the right frontal bone, right temporal bone, greater wing of sphenoid, medial wall of the right orbit infiltrating the right lateral rectus, superior rectus, and superior oblique, destroying the medial wall of the right ethmoid sinus and ethmoid septae. Involvement of the soft tissue extending to the upper and lower eyelids and infiltration of the right eye wall was also present. Inferiorly, extension in the intraconal compartment of the right orbit was seen. Superiorly intracranial extension was present (fig 1).

The differential diagnosis was a primary skull tumors such as an osteosarcoma, a Ewing sarcoma, and a chondrosarcoma, or a metastatic lesion. The possibility of MFH was not considered before surgery. The patient’s age and the absence of a primary tumor on full staging work-up made the possibility of a metastatic tumor less likely. An open biopsy of the lesion disclosed MFH. Right craniotomy with extensive, en bloc resection of the temporal region infiltrated by tumor and enucleation of the right eye was
performed.

On pathological examination, the cut surface grossly showed myxoid areas with small solid white areas. Microscopically, the section showed an infiltrating malignant neoplasm with both spindle cell and bony areas. The tumor was predominantly made of cells with loose pale background with scattered spindle cells showing slender nuclei with focal nuclear pleomorphism. Interspersed foci of increased cellularity were present with compactly arranged fascicles of spindle cells showing hyperchromatic nuclei and moderate amount of bipolar cytoplasm. Fair number of mitoses were evident in the cellular areas. Foci of necrosis and haemorrhage with lymphatic infiltration were also present. Vascular proliferation was evident (fig 2 a and b).

Figure 2
Figure 2: (a) H & E stained section showing a spindle cell neoplasm with hypocellular myxoid areas and (b) tumor invading bony lamellae 125 x digital magnification

The patient received two cycles of adjuvant chemotherapy with ifosfamide and doxorubicin, followed by 5600 cGy in 28 fractions followed by four more cycles of the same chemotherapy. The patient has completed the treatment and is presently in follow up.

DISCUSSION

Myxofibrosarcoma, also known as the myxoid variant of MFH, is one of the most common sarcomas in the extremities of elderly people and occurs more frequently in the dermal and subcutaneous tissues than in deeper soft tissues. It is relatively rare in the head and neck region. The maxilla and the mandible are the most frequently involved sites. MFH originating in the cranial bone rarely invades the brain tissues. Our case presented with a large defect in the right frontotemporal bone intracranial extension.

In recent years, the concept of MFH has come under discussion. This tumor is now believed to show fibroblastic rather than histiocytic differentiation. MFH is divided into four subtypes based on histological appearance; storiform–pleomorphic, myxoid, giant cell, and inflammatory. The myxoid type, also known as a myxofibrosarcoma, is the second most common type comprising 20% of all MFH. The myxofibrosarcoma is thought to be a variant, featuring less cellularity and a rich matrix; it also tends to be associated with a better prognosis than its more cellular counterparts. The rate of metastases of myxofibrosarcomas decrease as their myxoid components increases. The myxomatous change in these tumors does not merely represent degenerative change but is regarded as a form of differentiation areas in which the tumor cells multiply more slowly and produce an abundant mucoid matrix thus explaining the better prognosis.

MFH has been reported in irradiated areas or traumatized bone, in bone infarction, or in association with Paget disease and fibrous dysplasia. Mentzel et al. analyzed clinicopathological features of 75 cases of myxofibrosarcoma which had myxoid areas in at least 10% of the whole tumor. These lesions varied in appearance from a hypocellular, mainly myxoid and purely low-grade spindle cell to high-grade, pleomorphic (MFH-like) neoplasms with multinucleated giant cells, high mitotic activity and areas of necrosis. There was local recurrence in 33 of 60 (54%) cases between 2 and 72 months after surgery independently of the histological grade. Thirteen patients had metastases and 13 tumor-related deaths occurred. Most metastasizing lesions were high-grade.
Mentzel et al. (12) classified myxofibrosarcoma into two groups: superficial (dermal/subcutaneous) and deep (intramuscular/mainly subfascial). The first group tends to infiltrate while the second group tends to form a single discrete mass. The first group is characterized by lesions composed of gelatinous or firmer nodules that often spread extensively in a longitudinal manner. Sometimes these tumors extend to the deep fascia, losing their nodular appearance and demonstrating a more infiltrative growth pattern. Differential diagnoses in our case included various sarcomas of the skull as well as metastatic lesions. Clinical, radiological and histological distinguishing features are listed in Table 1 (13, 14, 15, 16).

<table>
<thead>
<tr>
<th>Clinical differential Diagnosis</th>
<th>Clinical distinguishing features</th>
<th>Radiological features</th>
<th>Histological features</th>
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<tbody>
<tr>
<td>Osteosarcoma</td>
<td>Commonly are found in the mandible and maxilla</td>
<td>Osteolytic soft-tissue exhausions. They may have calcifications within the lesion, no perilesional reaction, and poorly defined margins. Tumor (but not frequent) appearance is the runny picture.</td>
<td>Malignant spindle cell stroma, which directly produces osteoid or immature bone. Histological type: osteoblastic, chondroblastic, or fibroblastic form.</td>
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<td>Chondrosarcoma</td>
<td>Usually involves the skull base</td>
<td>Characterized by lytic and sclerotic changes within poorly defined margins.</td>
<td>Cartilaginous and immature cartilage deposition in areas of myxomatous change and cystic degeneration.</td>
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<tr>
<td>Angiosarcoma</td>
<td>Usually involves the head and neck region</td>
<td>Irregularly marginating vascular channels lined by one or more layers of atypical endothelial cells and pericytes, which have an anaplastic immature appearance.</td>
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<tr>
<td>Chondroma</td>
<td>Soft tissue masses usually seen in the subcutaneous area</td>
<td>CT scan shows a soft tissue mass with extensive bone destruction.</td>
<td>Physaliphorous cells (large, vacuolated, mucin containing) with a tubular arrangement and abundant extracellular mucinous capsule.</td>
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<tr>
<td>Giant cell Tumor</td>
<td>Involves the paranasal sinuses and commonly erodes the skull base</td>
<td>CT scan are hyperdense, contrast-enhancing masses.</td>
<td>Plump, spindle, or ovoid stromal cells together with uniformly dispersed, mononuclear, multinucleated giant cells.</td>
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<td>Fibrous dysplasia with malignant change</td>
<td>Age 2-15 years 50% present 90% present bone formation is involved at the woven stage (2 years), then cranial bone is involved more frequently, then to the skull. Clinical findings of increasing pain and an enlarging soft tissue mass suggest malignant change.</td>
<td>Appears radiolucent on ground glass generalized hazy appearance of the bone, convex lesion 2-10 mm in width. Destructive lesions with cortical erosion and reactive overgrowth. On CT scan, they show heterogeneous enhancement with focal necrosis.</td>
<td>Normal bone formation is involved at the woven stage, then cranial bone is involved more frequently, then to the skull. Clinical findings of increasing pain and an enlarging soft tissue mass suggest malignant change.</td>
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<td>Fibromyxoid sarcoma</td>
<td>Rare soft tissue neoplasm characterized by an aggressive behavior.</td>
<td>Radiolucent lytic lesions with thinning and widening of the cortex. Intramedullary extension has been reported.</td>
<td>Mildly atypical fibroblastic cells overlying, walled off growth embedded within a myxoid matrix. Nuclear atypia and pleomorphism were minimal, and necrosis was not present.</td>
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<td>Metastases</td>
<td>Multiple, small, non-encapsulated lesions usually indicate metastatic disease</td>
<td>Osteoblastic, with sclerosis and thickening (eg, prostate, breast, bladder, hypernephroma). Osteolytic, with bone destruction and lucency (eg, lung, uterus, GI tract, thyroid, melanoma, neuroblastoma).</td>
<td>Similar histologic features as their primary tumors.</td>
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Recurrent myxofibrosarcoma have usually higher histological grades. The higher-grade neoplasms tend to metastasize. Primary low and intermediate grade lesions can
be treated by wide resection. Higher grade lesions and recurrences may be treated by radiation or chemotherapy, however due to the small numbers of cases in the head and neck region this modality has not been well established. In a significant number of patients, local recurrence or metastases develop within 2 years (13).

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References
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