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

P Shukla, D Gupta, P Gupta, S Singh Bisht, M Pant, S Gupta, N Husain, V Joshi. *Myxofibrosarcoma of Right Fronto-Temporal Region With Intracranial Extension: A case report*. The Internet Journal of Oncology. 2008 Volume 6 Number 1.

Abstract

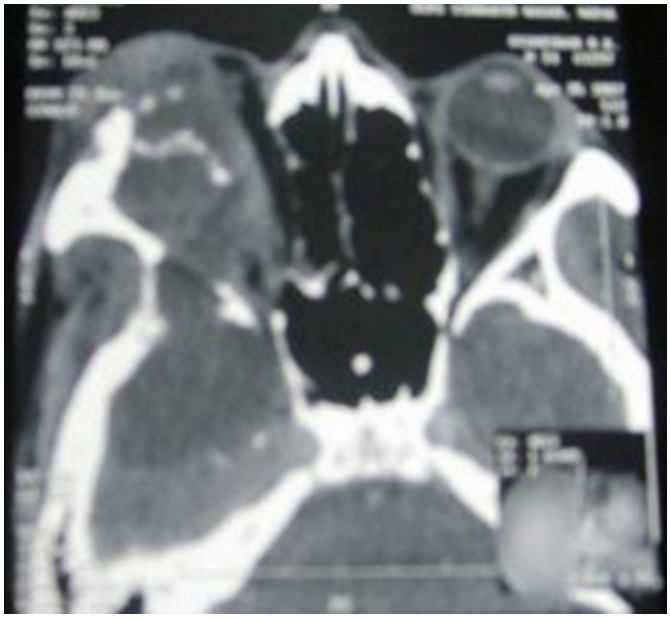
Myxofibrosarcoma, is a myxoid variant of Malignant Fibrous Histiocytoma (MFH) first described by Weiss & Enzinger (1) with a better prognosis. MFH rarely occurs in the skull, with only 17 cases documented in literature (2). The tumor frequently erodes the outer table of the skull and spreads in the extracranial 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).

Figure 1

Figure 1: CT scan showing tumor extension to involve the frontal bone with orbital and cranial spread



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

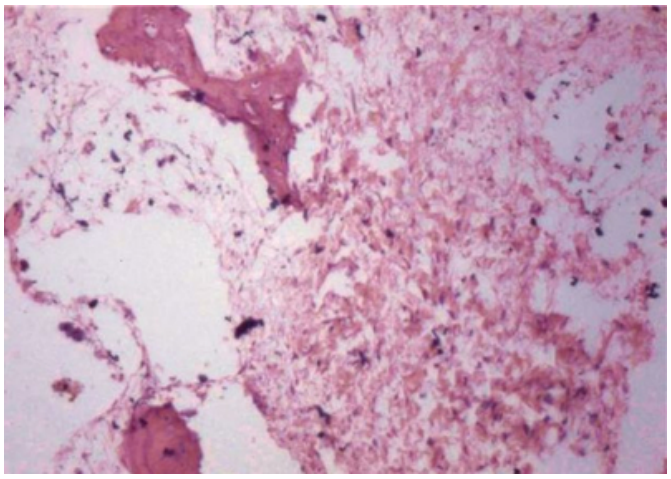
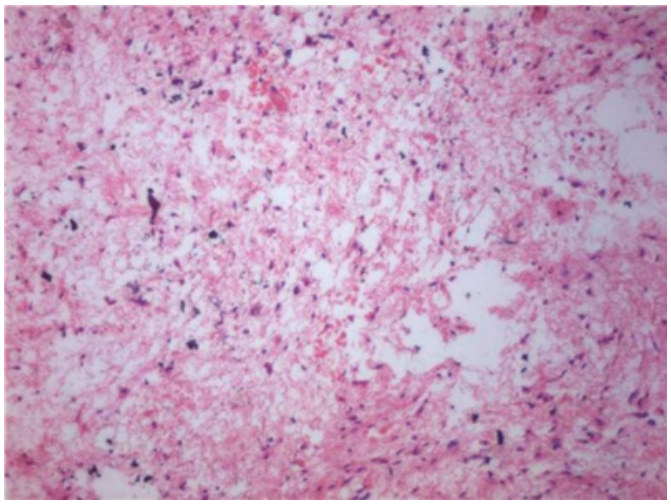


Figure 3



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 ([3,4,5]). 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 ([6,7,8]). 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 ([1,9]). 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 ([10]). 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 ([10,11]).

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. ([12]) 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]).

Figure 4

Table 1: Differential Diagnosis of Head and Neck Sarcomas

Clinical differential Diagnosis	Clinical distinguishing features	Radiological Features	Histological features
Osteosarcoma	Commonly are found in the mandible and maxilla	Osteolytic soft-tissue extensions. They may have calcification within the lesion, no periosteal reaction, and poorly defined margins. The typical (but not frequent) appearance is the sun-ray picture.	Malignant spindle cell stroma, which directly produces osteoid or immature bone. Histological types: osteoblastic, chondroblastic, or fibroplastic form.
Ewing's sarcoma	Young patients, usually involves the calvaria	Typical onion skin appearance with laminated periosteal changes. On CT scan is a dense mass surrounded by a hypodense area and hyperostosis.	Uniform, densely packed small cells with indistinct cytoplasmic borders and many mitotic figures
Chondrosarcoma	Usually involves the skull base	Characterized by lytic and sclerotic changes within poorly defined margins	Chondroid and immature cartilage deposition in areas of myxomatous change and cystic degeneration
Angiosarcoma	Usually involves the skull base, Cranial nerve deficits may be present	Destructive lesions with cortical erosion and reactive ossification. On CT scan, they show heterogeneous enhancement with focal necrosis	Irregular anatomising vascular channels lined by one or more layers of atypical endothelial cells and pericytes, which have an anaplastic immature appearance.
Chordoma	Soft-tissue masses usually seen in the nasopharyngeal area	CT scan shows a soft-tissue mass with extensive bone destruction.	Physaliphorous cells (large, vacuolated, mucus containing) with a lobular arrangement and abundant extracellular mucoid tissue.
Giant cell Tumor	Involve the sphenoid bone and commonly erode the sellar region	CT scan are hyperdense, contrast-enhancing masses.	Plump, spindle, or ovoid stroma cells together with uniformly dispersed, numerous, large, multinucleated giant cells.
Fibrous dysplasia with malignant change	Age 3-15 years 50% cranial cases have polyostotic form, frontal bone is involved more frequently. sion to a lytic lesion. Clinical findings of increasing pain and an enlarging soft-tissue mass suggest malignant change	Appears radiolucent on with ground-glass generalized hazy appearance of the bone, convex lesion both tables are intact and thinner. Malignant degeneration include a rapid increase in the size of the lesion and a change from a previously mineralized bony lesion to a lytic lesion	Normal bone formation is arrested at the woven stage; thus, lamellar bone is not formed. This results in an overgrowth of the fibrous tissue among woven bone, merges with fibrosarcoma - like proliferation
Fibromyxoid sarcoma	Rare soft tissue neoplasm characterized by a aggressive behavior.	Radiolucent lytic lesions with thinning and widening of the cortex. Intracranial extension has been reported	Mildly atypical fibroblastic cells swirling, whorled growth embedded within a myxoid matrix. Nuclear atypia and pleomorphism were minimal, and necrosis was not present.
Metastases	Multiple, small, nonmarginated lesions usually indicate metastatic disease	Osteoblastic, with sclerosis and thickening (eg, prostate, breast, bladder, hypernephroma) Osteoclastic, with bone destruction and lucency (eg, lung, uterus, GI tract, thyroid, melanoma, neuroblastoma)	Similar histologic features as their primary tumors

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 ([15]).

CORRESPONDENCE TO

Pragya Shukla, M.D. Department of Radiotherapy CSMMU, Lucknow Email: pragya_upadhyay@yahoo.co.in Phone number - +91- 510-2471553

References

1. Enzinger FM, Weiss SW. Soft Tissue Tumors. St. Louis: Mosby Year Book 1995;351–80
2. Matsuura S, Takagi T, Tan E-C, Mizuno S, Imagunbai N, Hasegawa R. Malignant fibrous histiocytoma of the occipital bone with intracranial extension: case report. Neurol Med Chir (Tokyo) 1991;31:219–222
3. Barney PL: Atypical fibrous histiocytoma (fibroxanthoma) of the temporal bone. Trans Am Acad Ophthalmol Otolaryngol 76: 1392-1393, 1972
4. Black SPW, Adelstein E, Levine C: Atypical fibrous histiocytoma in the skull of an infant. Case report. J Neurosurg 53: 556-559, 1980
5. Cook BR, Vries JK, Martinez AJ: Malignant fibrous histiocytoma of the clivus. Case report. Neurosurgery 20: 632-635, 1987
6. Dehner LP. Malignant fibrous histiocytoma. Nonspecific morphologic pattern, specific pathologic entity or both? Arch Pathol Lab Med 1988;112:236–7.
7. Fletcher CDM. Pleomorphic malignant fibrous histiocytomas: fact or fiction? A critical reappraisal based on 159 tumors diagnosed as pleomorphic sarcoma. Am J Surg Pathol 1992;16:213–28.
8. Hollowood K, Fletcher CDM. Malignant fibrous histocytoma: morphologic pattern or pathologic entity? Semin Diagn Pathol 1995;12:210–20.
9. Enjoji M, Hashimoto H, Tsuneyoshi M, Iwasaki H. Malignant fibrous histiocytoma: a clinicopathologic study of 130 cases. Acta Pathol Jpn 1980;30:727–41
10. Weiss SW, Enzinger FM. Myxoid variant of malignant fibrous histiocytoma. Cancer 1977; 39:1672–1685
11. Mentzel T, Calonje E, Wadden C, Camplejohn RS, Beham A, Smith MA, et al. Myxofibrosarcoma: clinicopathological analysis of 75 cases with emphasis on the low-grade variant. Am J Surg Pathol 1996;20:391–405.
12. Mahesh Kumar Neelala Anand. Fibrous Dysplasia. http://www.emedicine.com/RADIO/topic284.htm
13. Efstathios Papavassiliou, MD. Primary Malignant Skull Tumors. . http://www.emedicine.com/neuro/topic325.htm
14. Kagan Tun, Ozlem Ozen, Erkan Kaptanoglu, Oktay Gurcan, Ethem Beskonakli , Bulent Celasun. Primary intracranial low-grade fibromyxoid sarcoma (Evans tumor). Journal of Clinical Neuroscience, 2008; 15 (11) : 1298-1301
15. Evans HL. Low-grade fibromyxoid sarcoma: a report of 12 cases. Am J Surg Pathol 1993;17:595–600
16. Reiko Woodhams et.al. A Case of Malignant Fibrous Histiocytoma (MFH) in the Skull Bone Presenting Extensive Permeative Osteolysis. Radiation Medicine: 2002; 20: 141–145 .

Author Information

Pragya Shukla, M.D.
Department of Radiotherapy, King George's Medical University

Deepak Gupta, MD
Department of Radiotherapy, King George's Medical University

Pronati Gupta, M.D.
CSMMU Pathology SGPGI, King George's Medical University

Shyam Singh Bisht, MD
Department of Radiotherapy, King George's Medical University

Mohan Chand Pant, MD
Department of Radiotherapy, King George's Medical University

Seema Gupta, MD
Department of Radiotherapy, King George's Medical University

Nuzhat Husain, MD
Department of Pathology, King George's Medical University

Vindhya Joshi, MBBS
Department of Radiodiagnosis, King George's Medical University