MYXOID VARIANT OF MALIGNANT FIBROUS Histiocytoma: A CASE REPORT

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ABSTRACT: The Myxoid variant of malignant fibrous histiocytoma (MFH) is proposed as a separate subgroup of MFH because of its distinctive appearance and better prognosis compared with the storiform-pleomorphic type. A 56 year old female patient presented with a recurrent painful swelling in the lateral aspect of right thigh since 2 months. Histopathological examination revealed MFH of myxoid subtype. Excision with wide margins is the treatment of first choice.

Key words: Malignant fibrous histiocytoma, Myxoid, Storiform-pleomorphic, Recurrence

INTRODUCTION
MFH is a common soft tissue tumor in adults occurring in the 5th-7th decades of life and accounts for 10.5%-21.6% of all soft tissue malignant neoplasms [7, 8]. It is a high grade and aggressive sarcoma. Its histological origin is uncertain and its diagnosis is chiefly based on histopathological examination of routine H&E stained sections [2]. No ultrastructural features and immunohistochemical markers are ultimately specific for MFH. The term MFH was first introduced in 1963 to refer to a group of soft tissue tumors characterized by storiform or cartwheel like growth pattern. Although the tumors initially described by Ozzello et al. and later by O’Brein and Stout had a predominantly fibroblastic appearance. It was postulated that they were derived from histiocytes that could assume the appearance and function of fibroblasts [2]. Out of four variants of MFH, myxoid variant is the rare subtype of MFH category and it is recognized as myxofibrosarcoma [7]. This case report presents a patient with myxoid type of MFH of thigh region.

CASE REPORT
A 56 year old female presented in September 2012 to our department with a painful recurrent swelling in the lateral aspect of right thigh since 2 months. Past history of similar type of swelling at the same site was present, which was operated one year ago and was reported as fibromyxoma. On clinical examination, 9x9 cms soft swelling was present in the subcutaneous plane. The tumor was associated with tenderness without local rise of temperature. FNAC was done. Smears were moderately cellular comprising of pleomorphic spindle cells singly scattered against a myxoid background suggesting a myxoid mesenchymal tumor. Excisional biopsy was performed and the specimen sent for histopathological examination. Grossly, the tumor was 9x9x5 cms in size, globular. Cut section reveal circumscribed multinodular tumor with solid and cystic areas (figure-1). Cystic areas were filled with jelly like material. No fixity to overlying skin or underlying fascia.
Histologic examination showed a circumscribed multi nodular tumor composed of oval to plump spindle cells arranged in fascicles and diffusely in a prominent myxoid background. Moderate nuclear pleomorphism with few multinucleated giant cells were seen. Tumor showed prominent vasculature with occasional curved arcing of vessels. There were no areas of necrosis, no lipoblasts or cells with striations. The combination of pleomorphic spindle cells in myxoid stroma accounting for >50% of tumor suggested a diagnosis of Malignant Fibrous Histiocytoma of myxoid type. Immunohistochemistry staining was not done as there are no specific markers to confirm MFH. The patient recovered well and there were no symptoms of recurrence or metastasis till date.

Figure: 1: Gross specimen depicting the gelatinous appearance of solid and cystic areas.

RESULTS AND DISCUSSION
MFH manifests a broad range of histologic appearances and for this reason it is divided into four histologic subtypes. Storiform-pleomorphic, giant cell, inflammatory and myxoid MFH. MFH is a tumor of late adult life and clinical features of various subtypes are similar. The tumor seems most frequently on the lower extremity especially thigh, followed by lower extremity and retroperitonium. Out of all variants of MFH, storiform –pleomorphic variant is the most common type with a highly aggressive behavior, whereas myxoid variant of MFH is a rare subtype with a better prognosis.

The consistent feature in storiform-pleomorphic subtype is high nuclear grade with numerous atypical mitotic figures. Extensive necrosis with cystic degeneration and hemorrhage are other common features. Giant cell MFH is composed of undifferentiated pleomorphic cells with associated stromal osteclast giant cells.

Inflammatory MFH is now reserved for an undifferentiated high grade pleomorphic sarcoma with prominent infiltrate in addition to histiocyte, eosinophil, xanthoma cells. Most cases arise in retroperitonium.

The myxoid variant of MFH was proposed as a separate subgroup by Enzinger et al [2] in 1976 because of its distinctive appearance and better prognosis compared with the storiform-pleomorphic type. By definition, this tumor is composed of cells that qualitatively resemble those of an ordinary MFH, implying that the cells have a moderate or marked degree of nuclear atypia. In addition, at least one half of the tumor should be characterized by an abundant myxoid background [2]. Myxoid MFH has a predilection for patients older than 50 years [1]. The presented case was 56 years old. Most cases present in the dermis and subcutis. Only about 1/3rd occur in the deep soft tissue [1]. It usually arises in the limb girdles and limbs. Our case presented as swelling in the thigh region in a subcutaneous plane. This tumor recurs in almost 2/3rd’s of cases but metastasizes in only about 1/4th. The indolent course of the tumor is furthermore underscored by the longer interval between the time of diagnosis and metastasis. Although in part, the better prognosis is due to the superficial location, it has been shown by multivariate analysis that the myxoid subtype is by itself an independent prognostic factor [2].
The significance of myxoid change is not entirely clear. It is unlikely that it is a degenerative feature because other features of degeneration are lacking. Viewed simplistically, it might be regarded as an area in which the cells multiply more slowly but produce abundant myxoid matrix as a form of differentiation. This would explain the better prognosis most myxoid tumors have, compared to their cellular counterparts [2].

There has been a trend to popularize use of the term “Myxofibrosarcoma”, originally used during the 1970s instead of Myxoid MFH because of the belief that the lesions are fibroblastic rather than fibrohistiocytic. According to the approach adopted by Enzinger et al, highly myxoid fibroblastic/ fibrohistiocytic sarcomas (50% myxoid) can be divided into two groups depending on the level of atypia. Those with minimal atypia were referred to as ‘myxofibrosarcoma’ grade I, where as those with significant atypia are designated as myxoid MFH grade II-III [2].

The most important aspect of differential diagnosis is the clear distinction of this lesion from benign myxoid lesions such as nodular fasciitis, myxoma and malignant myxoid liposarcoma. These benign tumors lack extensive vasculature, bizarre cells seen in myxoid MFH. Although myxoid liposarcoma resembles this tumor grossly, it consists of a more uniform population of small spindle cells embedded in a clear matrix with a delicate plexiform vasculature. Bizarre cells are absent and mitotic figures are infrequent. Lipoblasts are usually present [5]. Despite the better prognosis, the myxoid form of MFH should be treated by wide local excision or amputation [8, 2].

Survival correlates with histologic grade but overall 5 year survival is 60-70% [2, 4]. The lowest grade lesions have no capacity to metastasize but may become higher grade and hence acquire metastatic potential in a recurrence. The higher grade lesions have an unusual but distinctive tendency to demonstrate lymph node metastasis, in addition to anticipated pulmonary and osseous spread [5].

![Figure: 2 Cyst wall showing highly pleomorphic cells- X400](image)

![Figure: 3 Broad myxoid zones abut sharply on cellular areas- X100.](image)
CONCLUSION
Wide local excision is the treatment of choice. Although the tumor has a better prognosis as compared to other histological variants, recurrences are common if not excised with wide margins. Awareness of this histological variant and adequate surgical clearance is essential in preventing recurrence.

REFERENCES