CASE REPORT

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Suprascapular malignant fibrous histiocytoma - A case report

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ABSTRACT

Malignant fibrous histiocytoma (MFH), the terminology implies that the origin of tumor cells is of fibroblastic and histiocytic cells. MFH was regarded as the single most common adult soft tissue sarcoma. MFH occurs most frequently on lower extremity, especially thigh, followed by upper extremity and retroperitoneum. We are presenting a case of MFH at right supra scapular region which is very rare site of presentation. After ruling out all the possible differential diagnosis and on the basis of microscopic examination as well as Immunohistochemistry (IHC) findings our diagnosis was of malignant fibrous histiocytoma (Undifferentiated high grade pleomorphic sarcoma). As the morphologic pattern seen with MFH is shared by a variety of poorly differentiated malignant neoplasms, careful sampling in conjunction with a targeted panel of immune stains is the mainstay of diagnosis.

Keywords: Malignant fibrous histiocytoma (MFH), Suprascapular region, Soft tissue sarcoma.

1. INTRODUCTION

Malignant fibrous histiocytoma, the terminology implies that the origin of tumor cells is of fibroblastic and histiocytic cells; however, the precise origin of MFH cells has been disputed and the concept of fibrohistiocytic differentiation has been challenged (Fletcher et al. 2001, Al-Agha et al. 2008, Nakayama et al. 2007, Murphey, 2007). Malignant fibrous histiocytoma (MFH) was first described in the early 1960s and became widely accepted as a specific soft-tissue sarcoma type in the 1970s (Fletcher et al. 2001, Kauffman et al. 1961, Ozzello et al. 1963, O'Brien et al. 1964). Subtypes of MFH includes storiform-pleomorphic, myxoid, giant cell and inflammatory. MFH was regarded as the single most common adult soft tissue sarcoma. The most common clinical presentation is an enlarging painless soft-tissue mass in the thigh and majority of tumors are intramuscular. We are presenting a case of MFH at right supra scapular region which is very rare site of presentation.

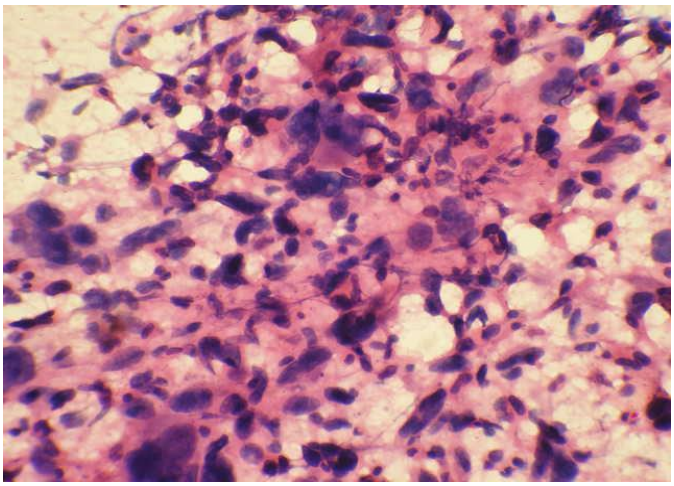

Figure 1

FNAC shows highly cellular smear with atypical spindle shaped cells and histiocytes with foamy cytoplasm

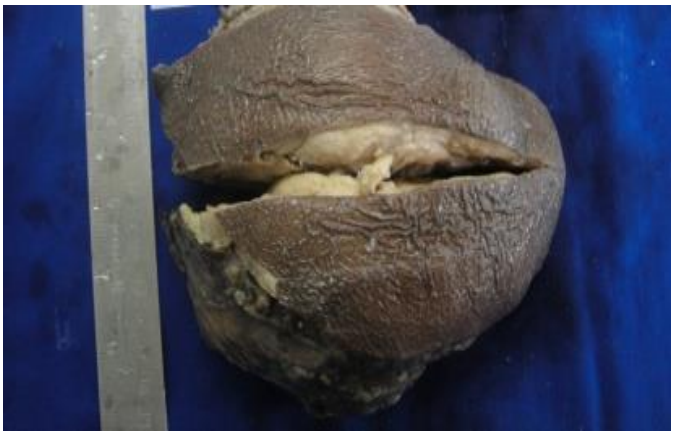

Figure 2

Large globular skin covered soft tissue mass with regular external surface

Figure 3

Cut surface shows multilobular greyish white areas admixed with areas of hemorrhage and necrosis

2. CASE REPORT

53 years old, male presented with history of rapidly growing painless swelling in the back for the past one month. On examination, lobulated swelling in the right supra scapular region measuring 12 X 8 cm, firm to hard in consistency was palpated. The surgeons decided to send the patient for the fine needle aspiration cytology (FNAC). FNAC smears showed highly cellular smear with atypical spindle shaped cells and histiocytes with foamy cytoplasm. Spindle cells showed marked pleomorphism and multinucleated tumor giant cells with bizarre nuclei were present. Mitotic figures were seen in necrotic background. Final impression was given of malignant spindle cell tumor (Figure 1).

Patient underwent wide excision of tumor mass and mass sent for histopathological examination. On Gross examination, there was a large globular soft tissue mass with attached elliptical skin flap which was measuring 14 X 11 X 7.5 cm with regular external surface (Figure 2). Cut surface showed multilobular grayish white areas admixed with areas of hemorrhage and necrosis (Figure 3). On microscopic examination, tumor consisted of spindle shaped fibroblast arranged in short fascicles and storiform pattern with plump to round shaped histiocytes showing marked pleomorphism were arranged haphazardly. There were numerous mitotic figures and many multinucleated giant cells with bizarre nuclei and areas of tumor necrosis were seen (Figure 4 & 5). Immunohistochemistry (IHC) was performed which showed invariably positive for vimentin, negative for most histiocytic marker such as CD1, some are positive for CD68, an antigen present in histiomonocytic cells, alpha -1 antitrypsin, alpha-1 antichymotrypsin were positive. After ruling out all possible differential diagnosis final impression was given as pleomorphic malignant fibrous histiocytoma [undifferentiated high-grade pleomorphic sarcoma (UHPS)].

3. DISCUSSION

Soft tissue sarcomas (STS) are a rare group of tumors, arising from mesenchymal tissue with heterogeneous differentiation. They account for 1 % of all malignancies (Fletcher et al. 2002, Ross et al.

1993). International incidence rates range between 1.8 and 5.0 cases per 100 000 per year (Ross et al. 1993, Storm, 1994, Schuurman et al. 1992, Levi et al. 1999, Gustafson, 1994, Toro et al. 2006). MFH alone have historically accounted for approximately 25 % of all patients accrued to sarcoma clinical trials (Lancet, 1997, Cochrane Database Syst Rev., 2000) but now account for no more than 5 % of adult soft tissue sarcomas (Randall et al. 2004). The tumor occurs most frequently on lower extremity, especially thigh, followed by upper extremity and retroperitoneum (Sharon W Weiss et al. 2008).

The concept of MFH has evolved since the upcoming of immunochemistry (Fletcher et al. 2002). Malignant fibrous histiocytoma (MFH) also known as undifferentiated high-grade pleomorphic sarcoma (UHPS) is possessed fibrohistiocytic morphology without definite true histiocytic differentiation. The results of the study from Sweden by Gustafson P, 1994, where the observed period ends in 1989, reflect the historical definition that the pleomorphic type of MFH has been regarded as the prototypical form of MFH and the most common histotype in adults. But in 2002, the World Health Organization (WHO) reappraised and modified the terminology and classification of MFH and its subtypes (Fletcher et al. 2002). Pleomorphic sarcoma is the alternate name advocated by the WHO to replace MFH, as it provides an accurate description of the tumor without implying the origin of the tumor cells (Al-Agha et al. 2008, Fletcher et al. 2002). The WHO nomenclature now designates storiform-pleomorphic MFH as "undifferentiated high-grade pleomorphic sarcoma," giant cell MFH as "undifferentiated pleomorphic sarcoma with giant cells," and inflammatory MFH as "undifferentiated pleomorphic sarcoma with prominent inflammation."

In our case the microscopic examination as well as IHC findings are suggestive of malignant fibrous histiocytoma (Undifferentiated high grade pleomorphic sarcoma). There were differential diagnoses of Pleomorphic Rhabdomyosarcoma, Fibrosarcoma, Pleomorphic Leiomyosarcoma and Pleomorphic malignant fibrous histiocytoma. Microscopic examination of Pleomorphic Rhabdomyosarcoma shows rhabdomyoblasts, cross striations, spindle to polygonal cells with deep eosinophilic cytoplasm and on IHC it shows positive myoglobin, MyoD1, myosin, myogenin. Fibrosarcoma composed of fibroblast with herringbone architecture and short fascicles, variable mitotic activity and no giant cells and IHC shows positivity for vimentin. In Pleomorphic Leiomyosarcoma, there will be intersecting fascicles and bundles of smooth muscle cells having pale eosinophilic fibrillar cytoplasm, osteoclastic type of multinucleated giant cells and IHC shows positive desmin, h-caldesmon and SMA. So after ruling out all the other possibilities and on the basis of IHC results we have given the diagnosis of undifferentiated high grade pleomorphic sarcoma. The same IHC findings were noted by Guo et al. that only vimentin was always expressed in MFH/ undifferentiated pleomorphic sarcoma, while some of the tumors were positive for myogenic antigen and CD68 (Guo et al. 2008). The vast majority of MFH are high grade lesions having a local recurrence rate ranging 19-31 %, a metastatic rate of 31-35 % and 5 years survival of 65-70 % (Murphey, 2007).

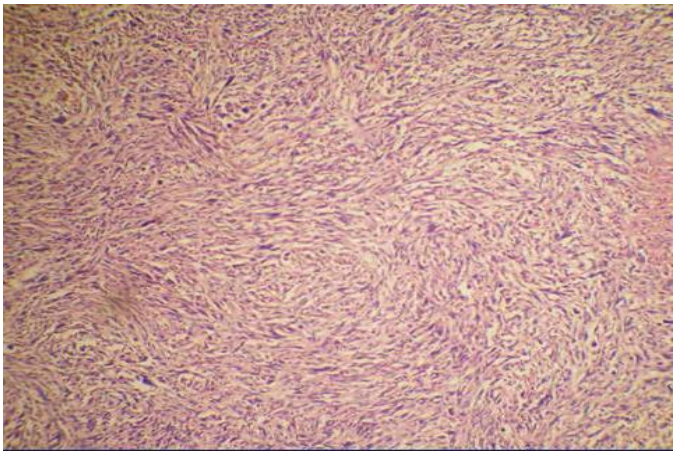

Figure 4

MFH tumors consist of spindle shaped fibroblast arranged in short fascicles and storiform pattern with plump to round shaped histiocytes

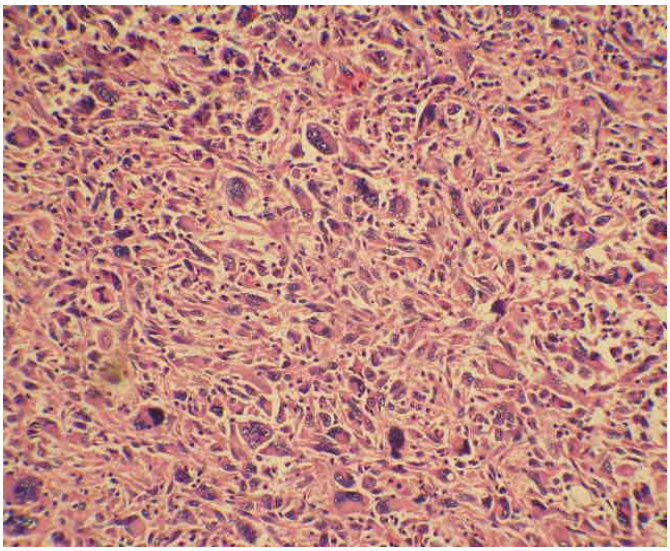

Figure 5

MFH Section showing marked pleomorphism with haphazard arrangement of tumor cells. There were numerous mitotic figures and many multinucleated giant cells with bizarre nuclei.

4. CONCLUSION

MFH is considered as diagnosis of exclusion for sarcomas that cannot be more precisely categorized as the morphologic pattern seen with MFH is shared by a variety of poorly differentiated malignant neoplasms. Furthermore, the prognosis also depends on the primary tumor grade and careful sampling in conjunction with a targeted panel of immunostains which is the mainstay of diagnosis.

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