

Diffusely Spreading, Superficial Malignant Fibrous Histiocytoma: Case Report with Cyto-and Histomorphologic Features

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Citation

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Abstract

Malignant fibrous histiocytoma (MFH) is a high grade soft tissue tumor occurring normally in the late adult life. Fine needle aspiration (FNA) cytology of MFH shows characteristic features of high grade pleomorphic sarcoma.

We report a case of diffusely spreading, superficial, large subcutaneous MFH in the back of a 35 year old Mexican male. FNA cytology showed a mixed population of malignant cells from small round to large polygonal or spindled cells with scant to moderate volumes of cytoplasm. Numerous bizarre tumor giant cells were present with one or more large hyperchromatic nuclei in a background of necrosis. This case is unusual because of young age of the patient and large diffuse plaque like tumor presentation.

FNA cytologic features of MFH are characteristic, but not diagnostic. In cases of atypical presentation, other high grade neoplasms are needed to be excluded by immunohistochemistry, electronic microscopy and cytogenetic studies.

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a relatively common malignant soft tissue tumor. It mostly occurs in the population of age between 50-70 years and often arises in deep soft tissue of the proximal extremities or retroperitoneum. Although there were a few case reports of fine needle aspiration cytology on the classic presentation of MFH or atypical fibroxanthoma (^{1,2,3,4,5}), we are reporting an unusual case of superficially spreading plaque like subcutaneous MFH in the back of a 35 year old Hispanic male with typical cytologic features and histologic patterns confirmed by ancillary ultrastructural and cytogenetic studies.

CASE REPORT

A 35-year-old man presented with burning pain on his back for several months. On examination, a dark purplish, indurated, plaque-like lesion was noted on his back in a band-like formation that spreaded from his left axilla to the right. The lesion was approximately 30X25 cm (Fig. 1).

Figure 1

Figure 1: A 35-year-old male present with dark purplish, indurated, plaque-like skin lesion.



It was not tender on palpation. It was focally ulcerated with variegated color and purulent discharge. He also had bilateral axillaries lymphadenopathy. All other organ systems of routine laboratory tests were negative for any significant findings. The computerized tomography scan with/without contrast showed absence of visceral metastasis. However, both liver and spleen were slightly enlarged. The initial clinical impression was cutaneous lymphoma or other

similar malignancies.

PATHOLOGIC FINDINGS

Fine needle aspiration (FNA) of the lesion obtained scant bloody fluid. Diff-Quik and Papanicolaou stain (Pap stain) slides demonstrated scattered mixed populations of pleomorphic tumor cells in a background of blood and necrotic debris. The neoplastic cells were present singly or in small groups. They were variable in size and shape with enlarged, hyperchromatic nuclei. Multinucleated giant cells, prominent nucleoli, and mitotic activity were readily identified. The cytoplasm of the neoplastic cells was finely vesicular to microvacuolated and showed variable shapes ranging from strap cells to round, polygonal and spindle shapes (Fig. 2, 3, 4).

Figure 2

Figure 2: FNA findings revealing pleomorphic neoplastic cells singly and in small groups (Diff Quick stain, x200).

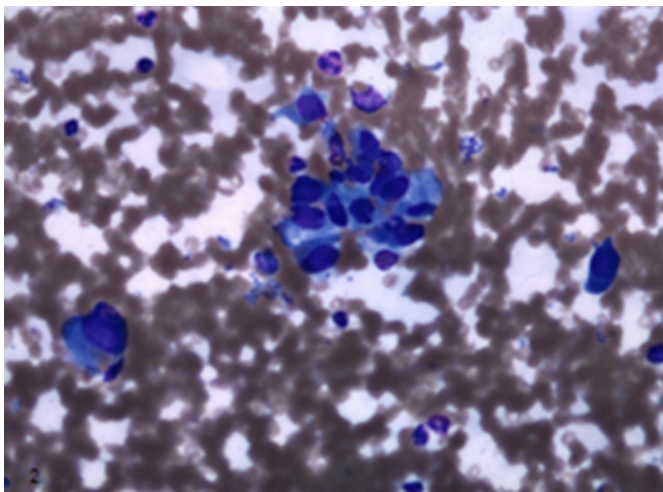


Figure 3

Figure 3: Pleomorphic neoplastic cells with binucleation (Diff Quick stain, X400).

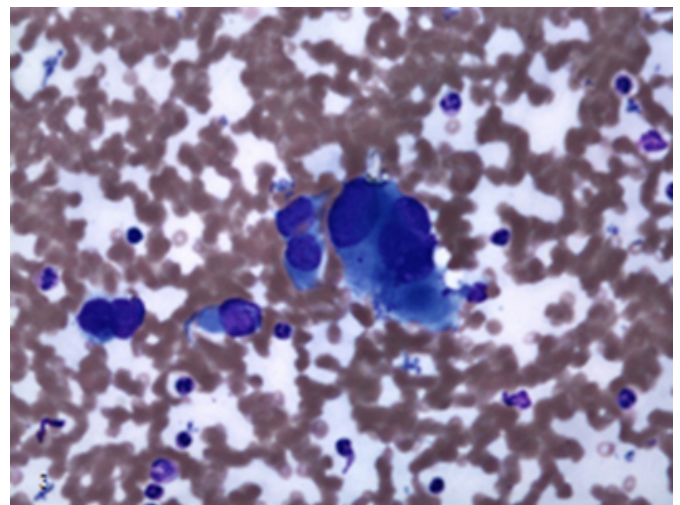
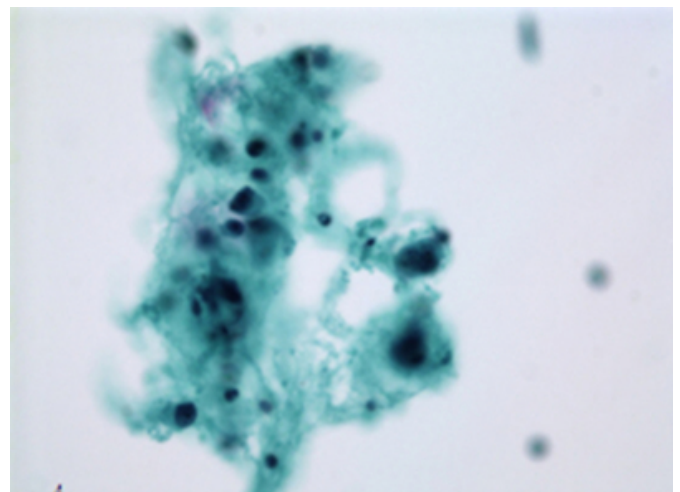


Figure 4

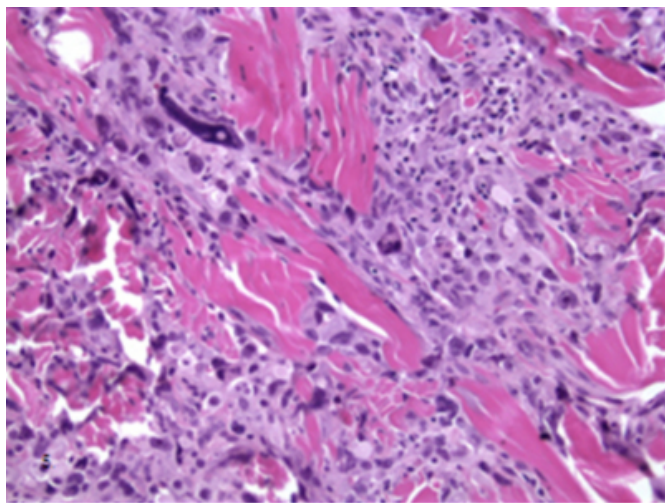
Figure 4: FNA demonstrating large bizarre neoplastic cells with histiocytic features (Papanicolaou stain, X200).



The follow-up tissue biopsy of the tumor showed a diffusely tumor infiltrating, composed of large, faintly spindle-shaped neoplastic cells, which in some areas appeared to be arranged in a vague, storiform pattern. There was considerable nuclear pleomorphism, marked hyperchromaticity and many of the nuclei contained large macronucleoli. The mitotic rate was brisk and there were numerous apoptotic cells. These neoplastic cells insinuated between the collagen fibers and infiltrated the adipose tissue and the deep adnexal structure (Fig. 5).

Figure 5

Figure 5: The neoplastic cells infiltrating the deep adnexal structure and subcutaneous tissue (H&E stain, X400).



Immunohistochemical stains showed the tumor cells positive for vimentin; and histiocytic markers CD68 and Factor XIIIa. Melanoma markers (S100, HMB45, Melan A, and MITF), carcinoma markers (cytokeratin AE1/3, EMA, CK7, CK20), lymphoma markers (CD45, CD20, CD3, CD4, CD43, CD5, CD30, ALK, and CD56) and markers for specific sarcomas (SMA, CD34, CD117, and desmin) were all negative. Lysozyme stain was negative. The tumor exhibited very high proliferation index (Ki67- more than 90%). The above immunoprofile was considered consistent with the diagnosis of a high grade sarcoma, favoring malignant fibrous histiocytoma.

Ultrastructurally, the neoplastic cells were remarkable for absence of any diagnostic features. The cells were large, roughly polygonal, with markedly enlarged, regular and angulated nuclei, many of which have very large nucleoli. The cytoplasm of these cells showed a relative paucity of intercellular organelles. There was a scattering of the rough endoplasmic reticulum, Golgi apparatus and various vacuoles. There were some interdigitations between the cells and an occasional tight junction was noted and no desmosomes were seen. No melanosomes or premelanosomes were noted. An occasional non-specific electron dense membrane-bound granule was noted. No intermediate or micro filament bundles were noted and no dense bodies were noted.

Cytogenetic analysis of the tumor cells with fluorescence in situ hybridization (FISH) method demonstrated numerous chromosomal abnormalities, but no unique abnormalities were found. Final diagnosis of MFH was rendered following

exclusion of other possible neoplasms.

DISCUSSION

Malignant fibrohistiocytic tumors can present in a cutaneous form, atypical fibroxanthoma, and a deeply situated form, MFH. Both of them share almost identical histopathologic features, and frequently occur in elderly. Atypical fibroxanthoma usually occurs on actinic-damaged skin such as head and neck, particularly the nose, cheek, and ear. They are generally present as a solitary nodule or bleeding ulcer on average less than 2 cm in diameter and without any distinctive borders. Characteristically atypical fibroxanthoma does not extensively involve the subcutis, nor does it invade the deeper structures such as fascia or muscle (6,7,8,9,10). In contrast the MFH can be deep seated with a broad range of histopathologic appearances (storiform-pleomorphic, myxoid, giant cell, and inflammatory). Despite the nature and classification of MFH have been a matter of controversy among the experts, MFH rarely present as infiltrating, giant, plaque-like cutaneous mass.

FNA cytology of MFH shows typical neoplastic giant cells, spindle cells, and round cells with/without myxoid, fibrous, inflammatory, and necrotic background. There are no single diagnostic criteria exclusive for MFH (12, 13). Presence of neoplastic multi-nucleated giant cells or large histiocyte-like cells with foamy or vacuolated cytoplasm is characteristic, but not specific for MFH pleomorphic/storiform type. On the other hand, presence of fibroblast-like cells with mild to moderate nuclear atypia in a myxoid background can be very difficult for the diagnosis of MFH myxoid type. Without significant malignant features such as marked nuclear pleomorphism, multinucleated tumor giant cells bizarre nuclei, irregular nuclear membrane, and coarse chromatin, a conservative diagnosis such as spindle cell neoplasm should be rendered. Although cytopathologic presentation of MFH is diverse and heterogeneous due to histomorphologic variety of MFH, identification of high grade pleomorphic sarcomas without phenotype differentiation is feasible by FNA. The immunostains and ultrastructural studies are helpful in excluding other malignancies.

Since MFH is a diagnosis of exclusion, extensive immunostain markers for malignant melanomas, spindle cell carcinomas, and leiomyosarcomas were employed on multiple biopsy specimens with ultrastructural and cytogenetic studies. The nature of undifferentiated high grade sarcomas was undisputedly established.

Large cutaneous MFH in trunk of young adult is extremely rare. Atypical cytologic presentation in FNA allows us to establish the diagnosis based upon appropriate clinical information and ancillary studies. It is imperative to rule out other malignancies such as other high grade sarcomas, malignant melanomas, spindle cell carcinomas, or cutaneous T cell lymphomas.

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