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Teaching cases

Primary inflammatory malignant fibrous histiocytoma of the breast: A case report of an unusual variant and review of the literature



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ABSTRACT

Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma of adults, but its presence in the breast is rare. We report a case of primary inflammatory MFH in a 72-year-old Caucasian female with no previous medical history and no prior radiation exposure. She presented with a palpable mass that was suspicious for malignancy on mammography. Histologic evaluation of the core needle biopsy revealed sheets of large, pleomorphic neoplastic cells within a dense background of acute and chronic inflammatory cells. The neoplastic cells exhibited a moderate to abundant amount of finely vacuolated cytoplasm and atypical nuclei with vesicular nuclear chromatin and prominent nucleoli. Mitotic activity was readily identified, and foci of necrosis were noted. The neoplastic cells were immunoreactive with CD68, alpha 1-antitrypsin, alpha 1-antichymotrypsin, and vimentin. The diagnosis of MFH was rendered after thorough microscopic examination of the entire mass following mastectomy. MFH of the breast is a diagnosis of exclusion. The definitive treatment of MFH is surgical, either with wide local excision or total mastectomy. The roles of sentinel lymph node biopsy, axillary lymph node dissection, chemotherapy, and radiation have yet to be definitively clarified. The prognosis of MFH of the breast is generally poor.

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Introduction

Malignant fibrous histiocytoma (MFH) is considered to be the most common soft tissue sarcoma of middle and late adult life and characteristically affects the extremities and, less frequently, the retroperitoneum of elderly male patients [1–5]. However, its occurrence in the breast is extremely uncommon [3–11], especially in patients with no history of radiation for a prior breast carcinoma or in cases that do not arise in association with a malignant phyllodes tumor [5,6,12]. Indeed, primary sarcomas of the breast are rare and account for less than one percent of all malignant neoplasms of the breast [10,11,13]. As a result of the wide spectrum of histologic appearances, four subtypes of MFH have been described in descending order of frequency: pleomorphic MFH, myxofibrosarcoma, giant cell MFH, and inflammatory MFH [1]. The angiomatoid subtype has been reclassified as a fibrohistiocytic tumor of low malignant potential. To date, approximately 50-60 cases of primary MFH of the breast have been reported in the English literature, with

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most being pleomorphic MFH [2]. To our knowledge, ours is the first reported case of inflammatory MFH of the breast with a significant inflammatory component.

Case report

We report an exceedingly rare case of a 72-year-old Caucasian female with no known previous medical history and no prior exposure to radiation who presented with a 2-3 cm palpable mass in the central superior left breast at the 11–12 o'clock position. No skin involvement was noted and there was no evidence of any other extremity or truncal soft tissue masses. The remainder of the physical examination was unremarkable. Mammographic imaging showed a 2 cm nodular mass that was felt to be suspicious for malignancy. A subsequent ultrasound-guided core needle biopsy was performed, and histologic examination of the submitted tissue samples revealed sheets of loosely cohesive large, pleomorphic neoplastic cells within a prominent inflammatory background composed predominantly of neutrophils and eosinophils. The neoplastic cells exhibited a moderate to abundant amount of finely vacuolated cytoplasm and atypical nuclei with vesicular nuclear chromatin and prominent nucleoli. Mitotic activity was readily identified, and foci of necrosis were noted. A panel of properly

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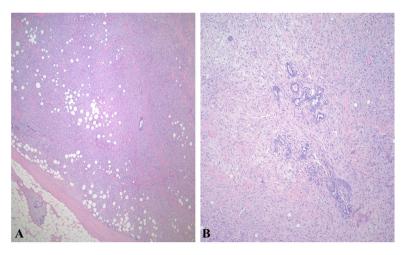


Fig. 1. (A) Sheets of atypical neoplastic cells resembling the pleomorphic areas seen in pleomorphic MFH lie adjacent to residual adipose tissue of the uninvolved breast parenchyma (H&E, original magnification $40\times$). (B) Entrapped benign-appearing mammary ductal epithelial elements are seen amid neoplastic spindled cells arranged in fascicles (H&E, original magnification $100\times$).

controlled immunohistochemical stains was performed. The neoplastic cells were immunoreactive with CD68, alpha 1-antitrypsin, alpha 1-antichymotrypsin, and vimentin. They were negative for cytokeratin AE1/AE3, cytokeratin 7, BRST-2, S100 protein, smooth muscle actin, desmin, myoglobin, CD30, and CD45. Although the histomorphologic features and immunohistochemical staining pattern were suggestive of a malignant fibrous histiocytoma (MFH), the needle core biopsy was diagnosed as malignant neoplasm with sarcomatoid features. A 3.0 cm \times 2.5 cm \times 2.0 cm tan-gray mass was identified in the upper outer quadrant in the ensuing unilateral mastectomy specimen. Microscopically, the neoplastic cells possessed morphologic characteristics similar to those as those seen on the core biopsy, with large, bizarre histiocytic cells admixed, atypical spindled cells, and a dense inflammatory infiltrate (Figs. 1-2). Foci of myxoid change were apparent but only accounted for approximately 10% of the tumor volume (Fig. 3). There was no evidence of angiolymphatic space invasion, and all surgical resection margins were uninvolved. A limited panel of immunohistochemical stains demonstrated results similar to those seen on the initial core biopsy (Fig. 4). A diagnosis of MFH was rendered after histologic evaluation of the entire mass. A sentinel lymph node biopsy was not performed at the time of surgery. A metastatic workup consisting of computed axial tomography (CAT) scans of the chest, abdomen, and pelvis was negative at the time of initial diagnosis, supporting the impression of a primary neoplasm arising in the breast.

A subsequent CAT scan performed approximately one month after the mastectomy demonstrated multiple minute nodules in the right and left lungs, all of which were consistent with metastatic disease. The patient was seen by Medical Oncology approximately two months after surgery, and no systemic therapy in the form of chemotherapy or radiation was recommended at that time.

Immunohistochemical staining procedure

All four micrometer thick sections were immunostained following the same procedure. The primary antibodies used in this case included CD68 (clone KP1, 1:100 dilution, Dako Systems, Inc., Denmark), alpha 1-antitrypsin (polyclonal, 1:1000 dilution, Dako Systems, Inc., Denmark), alpha 1-antichymotrypsin (polyclonal, pre-diluted, Cell Marque Corp., Rocklin, CA, USA), vimentin (clone 3B4, Ventana Medical Systems, Inc., Tucson, AZ, USA),

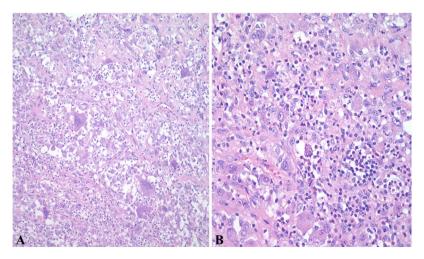


Fig. 2. (A) Sheets of loosely cohesive large, pleomorphic neoplastic cells within a prominent inflammatory background composed predominantly of neutrophils. Several giant cells with with multiple nuclei and deeply eosinophilic cytoplasm are noted (H&E, original magnification 200×). (B) The pleomorphic neoplastic cells exhibit a moderate to abundant amount of finely vacuolated cytoplasm and atypical nuclei with vesicular nuclear chromatin and prominent nucleoli. They lie within a dense inflammatory component made up of mostly neutrophils and eosinophils. A neoplastic xanthoma cell containing phagocytosed neutrophils is identified in the center of the field (H&E, original magnification 400×).

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