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Case Report

Primary breast sarcoma in a male patient

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ABSTRACT

Primary sarcoma of the breast is an extremely rare tumor in men. Furthermore, only a few cases of undifferentiated/unclassified sarcoma of the male breast have been reported in the literature. We report a case of primary undifferentiated sarcoma occurring in the male breast. A 66-year-old male patient presented with a rapidly growing ulcerative left breast lump. A large partially circumscribed, dense mass was noted on mammography which involved the overlying skin with contour bulge. Sonographic evaluation revealed a large hypervascular heteroechoic mass showing areas of internal necrosis. An initial core-needle biopsy suggested malignant phyllodes tumor, but final histopathology revealed a high-grade primary sarcoma with immunohistochemistry suggesting uncertain differentiation. A review of the literature is presented with focus on imaging and treatment of these unusual neoplasms.

Keywords: Male, Breast, Sarcoma, Primary, Undifferentiated

INTRODUCTION

Primary breast sarcomas (PBSs) are rare tumors originating from the mesenchymal tissue of the breast.[1] Undifferentiated/unclassified sarcomas have been defined as a group of sarcomas that lack an identifiable line of differentiation and cannot be classified. These have been included in the category of tumors of uncertain differentiation in the 2020 WHO classification of soft tissue sarcomas.[2] They usually occur in middle-aged women and are very rarely reported in men. To the best of our knowledge, only eight cases of primary undifferentiated breast sarcoma have been reported in males in literature till the year 2023.[3] We present a case of primary undifferentiated breast sarcoma in a 66-year-old man with a review of the literature and discuss the imaging and treatment of these unusual neoplasms.

CASE REPORT

We present a case of a 66-year-old male patient with a left breast lump that was progressively increasing in size. On examination, an ulcerative, firm reddish nodule was noted in the left outer breast at 3'o clock position [Figure 1] with mild reddish discoloration of surrounding skin.

On mammography [Figure 2], a large, partially circumscribed, dense mass was noted in the outer half of the left breast with indistinct posterior margins, involving the overlying skin with contour bulge (BIRADS 4c). Ultrasonography [Figure 3] revealed a large, heteroechoic mass with significant internal vascularity and areas of internal necrosis. No evidence of axillary lymphadenopathy was noted.

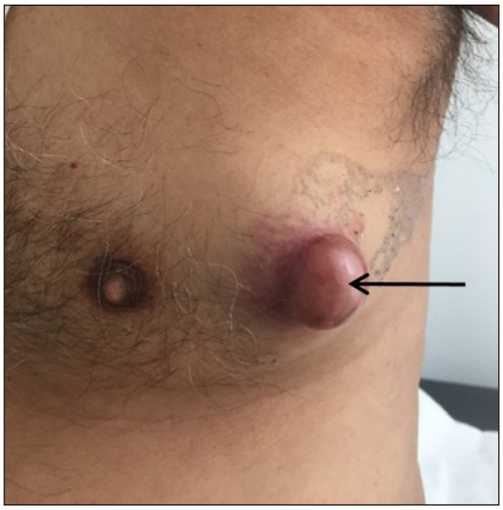

Figure 1: On clinical examination, a reddish nodule (black arrow) was noted in the left outer breast at 3 o'clock position with mild reddish discoloration of surrounding skin.

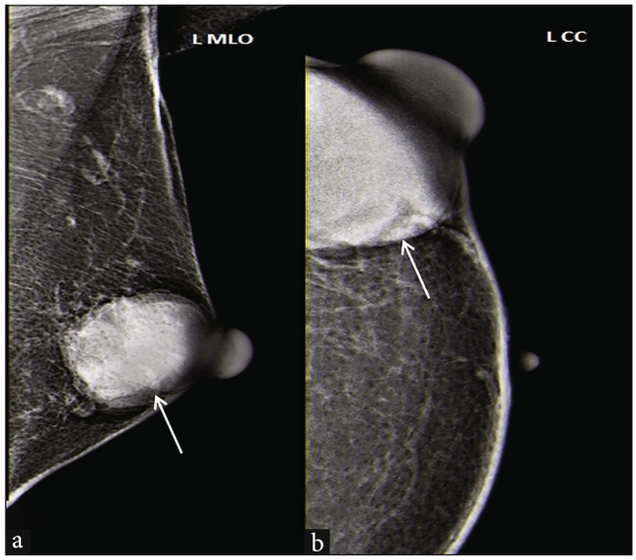

Figure 2: (a-b) Mammogram revealed a large partially circumscribed, dense mass (white arrows) in the outer half of the left breast with indistinct posterior margins and involvement of overlying skin with focal bulge. LMLO: Left mediolateral oblique; LCC: Left craniocaudal.

Since the imaging findings were suspicious for malignancy, ultrasonography-guided core-needle biopsy was performed which suggested a malignant phyllodes tumor.

Contrast-enhanced computed tomography (CT) of the chest and abdomen revealed no evidence of metastasis. Left breast mastectomy was performed. Macroscopically, a central lobulated tumor was seen bulging over the skin. Microscopic examination [Figure 4] revealed a neoplasm with infiltrative borders comprising spindle cells arranged in sheets and fascicles with moderate eosinophilic cytoplasm and moderate pleomorphic nuclei, showing brisk mitosis with necrosis. As no evidence of entrapped epithelial components was noted after an extensive histological examination, the malignant phyllodes tumor was ruled out. Immunohistochemistry (IHC) [Figure 5] revealed weak focal positivity for smooth muscle actin. It was negative for cytokeratins, desmin, S-100, caldesmon, CD31, CD34, and P63. Hence, a final diagnosis of high-grade primary sarcoma of breast was made, best favoring an unclassified/undifferentiated sarcoma.

DISCUSSION

PBSs are exceedingly rare, representing fewer than 0.1% of all malignant tumors of the breast.[1] PBSs arise from the the mesenchymal tissue of the mammary gland and include liposarcoma, fibrosarcoma, leiomyosarcoma, sarcomas with bone and cartilage, and sarcomas of uncertain differentiation. Approximately 10–20% of soft tissue sarcomas lack an identifiable line of differentiation and cannot be classified, termed undifferentiated/unclassified sarcomas.[2] PBSs usually occur in older females (with a median age of 56), and patients usually present with a unilateral painless lump. They can rapidly grow into large masses, but the commonly reported median tumor size is 4 cm.[4] These tumors are locally aggressive with a tendency for recurrence. Pectoral muscle invasion, skin changes, and nipple retraction are rare. Metastasis from breast sarcoma commonly spreads hematogenously and typically to the lungs, bones, and liver.

Breast sarcomas have a nonspecific imaging appearance and can mimic benign entities or relatively well-circumscribed malignant lesions like malignant phyllodes tumors and metaplastic carcinomas. Mammographic findings most commonly include mass with well-circumscribed, microlobulated, or indistinct margins, usually without any spiculations, microcalcifications, or axillary lymphadenopathy, as in our case.[5,6] However, the tumor can still depict histologic invasion despite having well-circumscribed borders grossly and radiologically. On the other hand, invasive breast carcinomas usually have stellate or ill-defined borders on imaging and show extensive infiltration histologically.[7] Furthermore, axillary lymphadenopathy is frequently seen in carcinomas including metaplastic carcinoma. The second most common pattern is architectural distortion which is a feature more typically seen in breast carcinoma.[8] Among sarcomas, this finding is more frequently associated with angiosarcomas which may be attributed to their different growth pattern. Unlike most breast sarcomas, which grow as rapidly expanding mass lesions with a pushing border, leading to a radiological appearance of circumscribed or

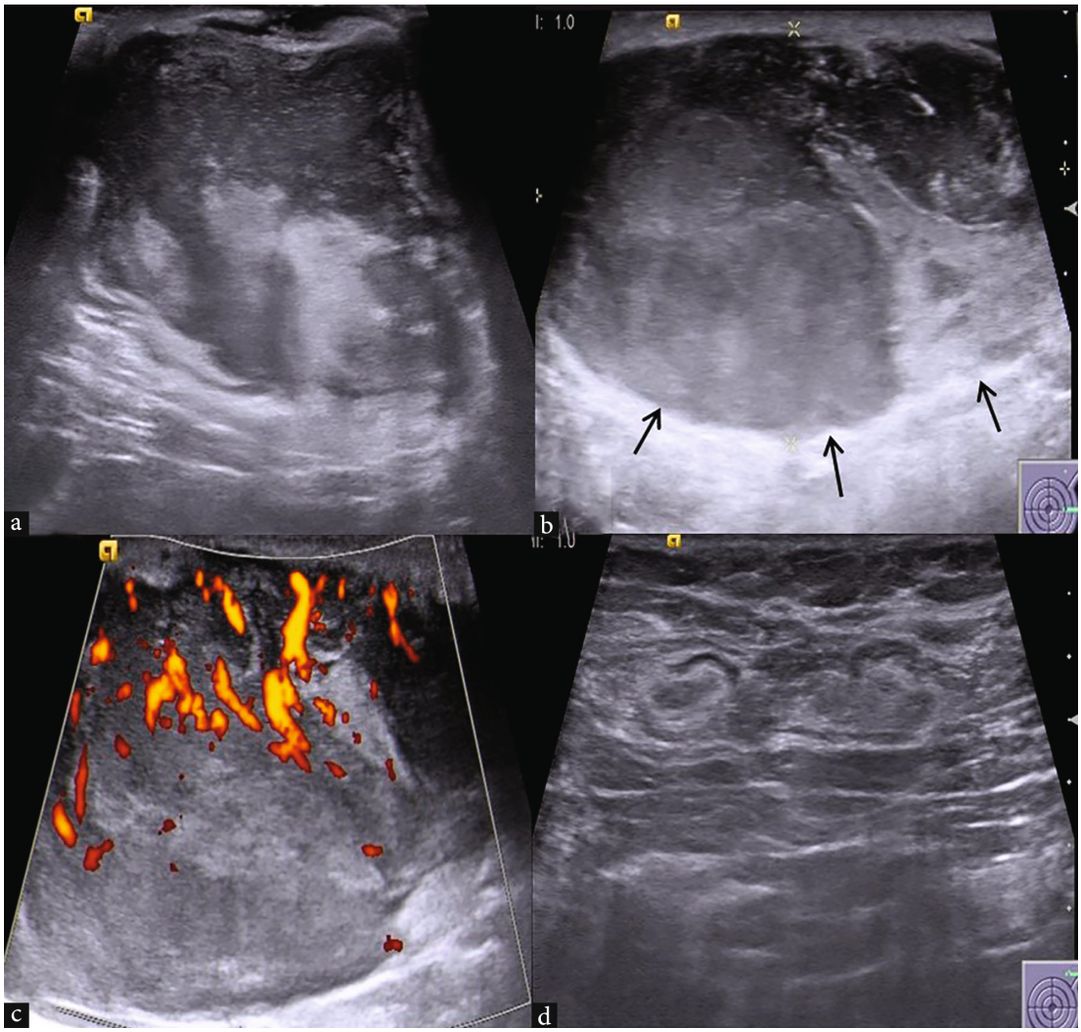

Figure 3: (a-b) The grey scale sonogram of the left breast at 3 o'clock position showed a large heteroechoic mass (black arrows) with areas of internal necrosis. (c) The Doppler sonogram evaluation showed internal hypervascularity within the mass. (d) No lymphadenopathy was seen in the ipsilateral axilla.

indistinct margins, angiosarcomas typically present as poorly circumscribed hemorrhagic masses containing fewer cellular elements scattered around the central tumor area.[8] Focal asymmetry and negative mammograms have also been reported.[6] A retrospective review has also reported osteoid calcification in 21% of the cases in contrast to microcalcifications seen in invasive ductal carcinomas.[7]

On ultrasound, breast sarcoma usually appears as hypoechoic, highly vascular masses with microlobulated or indistinct margins without posterior shadowing.[9] Hyperechoic masses have been also described.[5] In our study, the lesion showed mixed echogenicity with internal necrosis and significant vascularity. It is often challenging to differentiate between phyllodes tumors and PBSs using mammography and ultrasound.

Although we did not use magnetic resonance imaging (MRI) in this case as it would not have impacted the management plan, it can be helpful for further characterization and evaluating the extent of the tumor. Breast sarcomas appear as lobulated T1 hypointense and T2, short tau inversion recovery (STIR) hyperintense masses with variable enhancement and internal necrosis on contrast-enhanced MRI. Features such as internal septations and hyperintense slit-like fluid spaces on T2-weighted images, which have been described in phyllodes tumors, were not observed in breast sarcomas in the study by Smith *et al.*[7] However, histopathological differentiation is often necessary. The absence of entrapped epithelial components in our case precluded the diagnosis of the phyllodes tumor. Furthermore, a lack of reactivity for cytokeratin and many other markers on an extensive IHC panel excluded epithelial cell origin differentials such as metaplastic carcinoma.

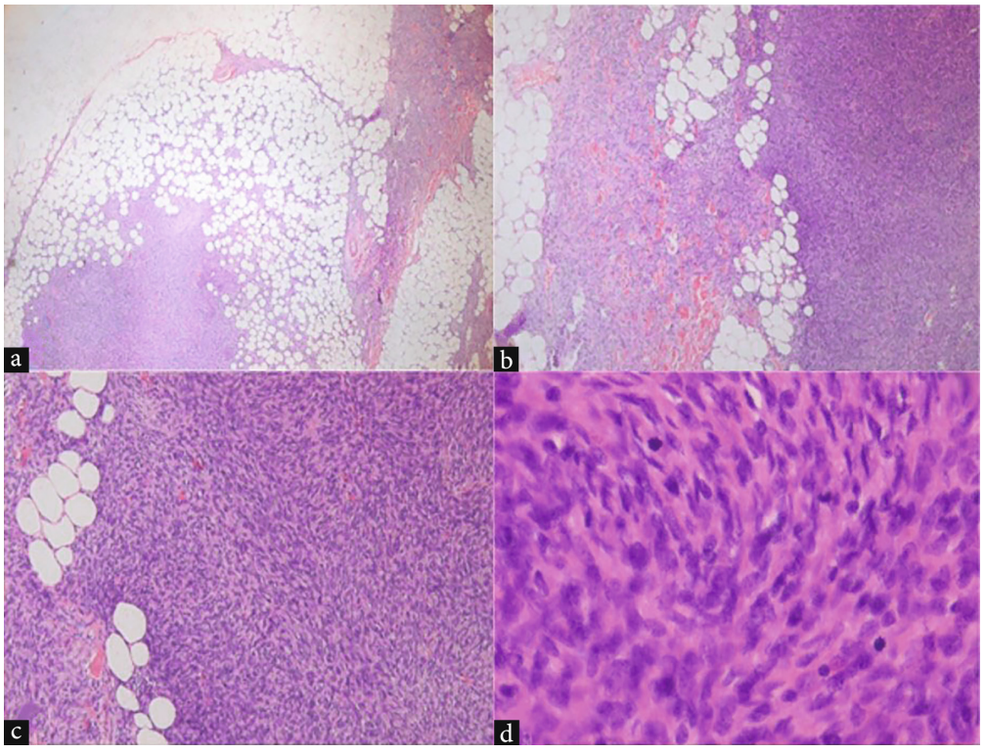

Figure 4: Hematoxylin and eosin (H & E)-stained sections showed (a-b) a neoplasm with infiltrative borders (c) comprising of spindle cells arranged in sheets and fascicles with (d) moderate eosinophilic cytoplasm and moderate pleomorphic nuclei. Brisk mitotic activity was noted. Foci of necrosis were seen.

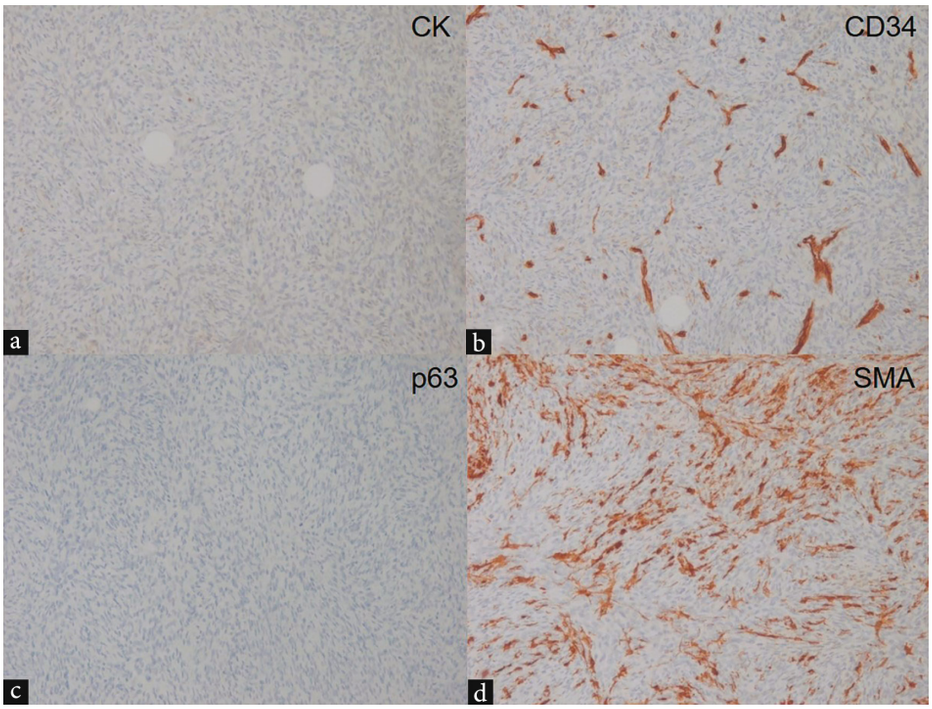

Figure 5: (a-d) Immunohistochemistry revealed weak focal positivity for smooth muscle actin (SMA). It was negative for cytokeratins (CKs), CD34, and P63. It was also negative for caldesmon, S-100, and CD31 (not shown). CD: Cluster of differentiation.

The most reliable prognostic criterion on imaging is the tumor size, with tumors smaller than 5 cm associated with a better prognosis.[10] The primary treatment involves surgical excision with a recommended resection margin of 1 cm.[11,12] Achieving an adequate resection margin is the most crucial factor for long-term survival.[13] Lymph node dissection is not needed unless involved. Adjuvant radiation has been shown to improve outcome in high-grade tumors and is recommended for tumors larger than 5 cm. Response to chemotherapy is variable, but neoadjuvant as well as adjuvant chemotherapy should be considered in high-risk cases.[14] The overall 5-year survival rate ranges from 49% to 67%.

CONCLUSION

PBSs are extremely rare in men. They can appear benign on clinical examination and imaging, especially when small, but rapid growth should alert the clinician to the possibility of an underlying sarcoma. On imaging, these can mimic phyllodes or metaplastic carcinoma, but the diagnosis can be confirmed on histopathology. Surgery with an adequate resection margin is the mainstay of treatment followed by adjuvant radiation in larger tumors, with the role of chemotherapy being controversial.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that they have used artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript or image creations.

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