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Case Report
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A Rare Case of Spindle Cell Sarcoma Presenting as a Recurrent Breast Cyst

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Abstract

Primary sarcomas of the breast are rare neoplasms which comprise between 0.2%-1.0% of all breast malignancies. They often carry a poor prognosis with mortality rates greater than 40%. In addition, these malignancies can be difficult to diagnose in a timely fashion as they can mimic other benign breast pathologies such as cysts.

Here we present a case of a primary spindle cell carcinoma of the breast in a 53 year old female with fibrocystic breast disease which presented as a recurrent large breast cyst. Although she underwent a cyst aspiration and a FNA biopsy of her breast lesion during outpatient visits the results of both were negative for malignancy. Following suspicious mammography findings the patient underwent a breast cyst resection. A diagnosis of a high grade spindle cell sarcoma was made after immunohistochemical analysis showed a tumor in the wall of the cyst staining positive with Vimentin, Smooth Muscle Antigen (SMA) and Ki-67 as well as focal positive staining with S100, CD 68, p63. Despite undergoing a completion mastectomy and resection of the pectoralis major the patient's sarcoma recurred one month later and she subsequently refused further treatment.

We have illustrated a rare case of primary spindle cell sarcoma of the breast which presented as a recurrent breast cyst that had a negative cytology as well as a negative FNA. Clinicians should be aware of such atypical presentations as delays in diagnosis can contribute to the mortality rates of this rare but aggressive malignancy.

Introduction

A sarcoma is a cancer that originates from mesenchymal cells and can originate from almost anywhere in the body. Primary breast sarcomas are extremely rare and account for 0.2%-1.0% of all breast cancers [1]. They can arise as primary lesions, after radiation therapy [2,3], or in the setting of lymphedema of the arm or breast [4]. The majority of breast sarcomas appear in the fourth and fifth decade of life. Mean age is roughly 40 years [5]. Coming to a definitive diagnosis of sarcomas can be quite difficult and immunohistochemical techniques are helpful in confirming this pathology. Here we report a case of spindle cell sarcoma which presented a recurrent cyst and eluded initial biopsy attempts at diagnosis until there was advanced local disease.

Case Report

A 53 year old African-American woman with a life long history of cystic breasts related to menstrual cycle and a family history significant for breast cancer in 4 sisters presented in May 2013 to clinic due to pain and enlargement of the left breast which she first noted in April. On physical exam she was found to have a large breast cyst in the left breast from which approximately 500cc of sero-sanguinous fluid was aspirated and sent to cytology which did not show evidence of malignancy. She continued to follow up in clinic for multiple cyst aspirations. She also underwent breast imaging in July that included breast ultrasound (US) and mammogram. The mammogram results showed a new large 12cm mass in the left breast that was not present in prior mammograms with portions of its border that were poorly defined the over it was noted to be thickened consistent with an inflammatory process, overall BIRADS-0 (Figure 1). The ultrasound revealed multiple small cysts in both breasts as well as a large 9.8cm x 6.9cm x 10cm cyst (Figure 2) in the upper half of the left breast with low level internal echoes and a thickened, irregular, hyper vascular wall (Figure 3a and b). Subsequently Fine Needle Biopsy (FNA) and core needle biopsies were performed. Her FNA results were negative for malignancy and her core biopsies showed sclerosing ad enosis, fibrocystic changes and micro calcifications but no malignancy.

When the patient returned in September 2013, on physical exam residents noticed a large mass

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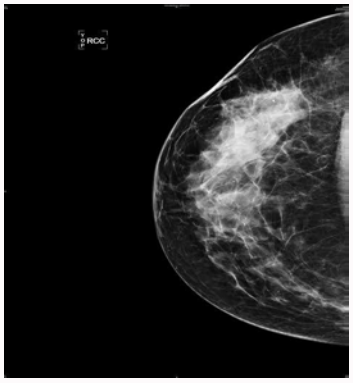

Figure 1: Left breast mammogram showing a large 12cm mass, that was not present on prior imaging.

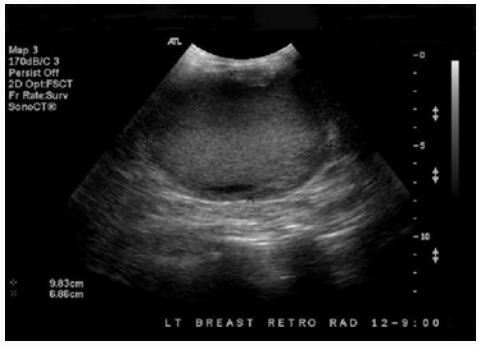

Figure 2: Ultrasound of left breast showing large 9.8cm x 6.9cm x 10cm cyst.

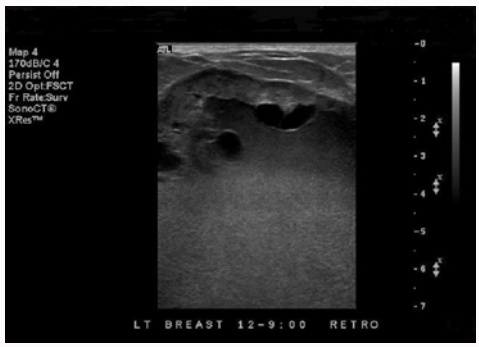

Figure 3a: Closer ultrasound of left breast cyst wall showing irregular border.

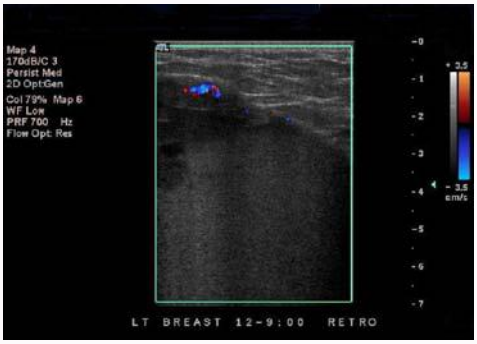

Figure 3b: Doppler ultrasound showing hyper vascular cyst wall.

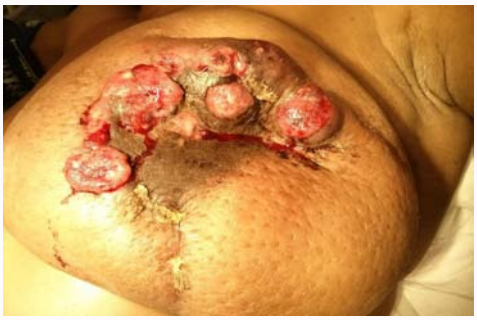

Figure 4a: Left breast prior to mastectomy.

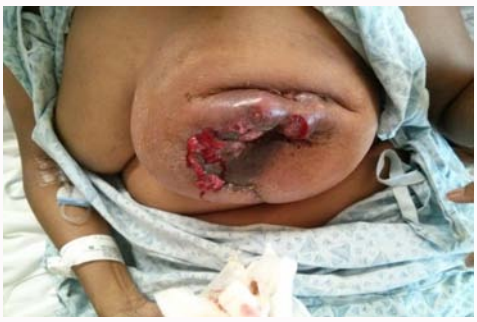

Figure 4b: Left breast prior to mastectomy.

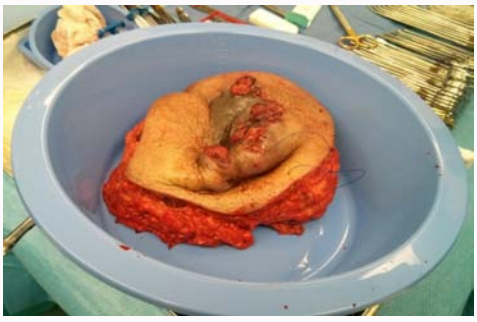

Figure 5: Left breast post mastectomy specimen.

in the left breast that was associated with pain, nipple retraction, erythematous changes, and purple discoloration along with peau d'orange skin which was hot to touch (Figure 4a and b). A lumpectomy and cyst excision was performed on September 16, 2013 in Brookdale Hospital and the initial frozen sections only showed spindle cells. Permanent sections were sent to Johns Hopkins for analysis. The Immuno-histochemistry study revealed positive staining with Vimentin, smooth muscle antigen (SMA), Ki-67, focal positive staining with S100, CD 68, p63. There was negative staining with Actin, Desmin, AE-1/AE-3, CAM5, GCDFP-15, ER, Mammaglobin, GATA-3, CK903, CK5, 6, HER2/NEU, Melan A and HMB46. These established histological features of a malignant spindle cell tumor as well as the immunohistochemical results led to the diagnosis of invasive malignant spindle cell neoplasm of high grade with scattered associated osteoclast like giant cells. Subsequently, staging Computed Tomography (CT) scans of the thorax, abdomen and brain were negative for metastases. The patient underwent completion mastectomy of the left breast and excision of pectoralis major muscle in November 2013 (Figure 5); axillary lymph nodes were not excised. Routine hematological and biochemical examinations, Ca15–3, and CEA were found to be normal. Her final pathology report is consistent with a high grade spindle and epithelioid neoplasm. The tumor was seen in multiple foci measuring from 0.5cm to 5cm in

greatest dimension and the margins were clear.

Discussion

The most common breast malignancy by far is ductal or lobular carcinoma with breast sarcomas making less than 1% of all breast malignancies [5]. Sarcomas may be primary or metastatic and they derived from mesenchymal cells but constitute many histopathologically distinct neoplasms. Some of the more frequently encountered breast sarcomas include angiosarcoma, malignant fibrous histiocytoma, liposarcoma, fibro sarcoma, clear cell sarcoma and neurogenic sarcoma among others [6]. Of these, angiosarcoma is the most common and is often associated with prior radiation and lymphedema [7]. Cyst sarcoma phylloids is another distinct clinicopathologic entity which is not a true stromal sarcoma as it retains benign epithelial components which true sarcomas lack [6,8]. Small published case series studies have reported 5 year survival rates for true breast sarcomas between 40%-66% [6,8-10] which is somewhat less than the 60%-80% 5 year survival rates reported for malignant cyst sarcoma phylloids [11-13].

Usually breast sarcomas present similarly to sarcomas on the rest of the body as a gradually increasing mass, typically without pain or overlying skin changes [10]. This presentation is variable however, such as in the case outlined here where the sarcoma presented as a large recurrent cyst, rather than a solid mass. Despite usually being sporadic, sarcomas have been associated with prior radiation exposure and chronic lymphedema [2-4]. Diagnosis can be made by core needle biopsy or open biopsy with immunohistochemistry staining for desmin, vimentin, smooth muscle antigen, keratin, and leukocyte common antigen, CD34, HMB45, EMA, and S-100 [7]. Prognosis has been cited to be most often correlated with tumor size and grade [9,10].

Treatment generally entails either low wide local excision with negative margins or simple mastectomy depending on the size and location of the tumor. Margin of surgical resection has been shown to be the most important prognostic indicator of adequate surgical treatment with a hazard ratio of a positive margin reported to be 7.9 [10]. Routine axillary lymph node dissection is not indicated [6] because sarcomas metastasize haematogenous and rarely travel to lymph nodes [7]. Use of radiation therapy and chemotherapy is controversial as there are no clearly defined guidelines for their use, however, some studies advocate using both for all high grade sarcomas [10].

Conclusion

This case report serves to show the presentation, workup and treatment of a rare primary breast spindle cell sarcoma. Breast sarcomas are rare but highly lethal cancers which need to be caught early to ensure prompt treatment and adequate surgical resection to minimize patient mortality. Unfortunately, as this case illustrates, sarcomas are often difficult diagnoses to make in the breast as findings are often non-specific and can be initially confused with benign conditions. This is further complicated by the fact that multiple biopsies including FNA, core needle, cytology and even operative frozen sections may fail to diagnose a breast sarcoma. Clinicians need to be aware that a high index of suspicion based on history and physical findings of the patient may be all that is available to make this diagnosis in its early stages.

Authors Declaration

The author have no conflict of interest.

NB This case report was presented as a poster in the South Eastern Surgical Conference in February 2015.

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