Primary Low Grade Spindle Cell Sarcoma Of The Breast

Anne Wilkinson[1], Radhika Mhatre[2], Anjali Patrikar[3]
[1] Associate Professor, [2] Resident, [3] Associate Professor, Pathology Department, NKP Salve Institute Of Medical Sciences And Research Centre, Hingna Nagpur - 440019, Maharashtra, India

ABSTRACT

Spindle cell sarcomas of the breast are very rare accounting for 0.2 to 1 % of all breast malignancies. Sarcomas of the breast differ from the invasive epithelial cancers in their presentation, behavior, management, staging and prognosis. Hence it is important to be aware of these rare lesions. This article describes a case of primary low grade spindle cell sarcoma of the breast in a 70 year old female patient.

Key words: Breast, Spindle cell sarcoma, primary

Corresponding author address: Anne Wilkinson, Associate Professor, Pathology Department, Nkp Salve Institute Of Medical Sciences And Research Centre, Hingna Nagpur - 440019,Maharashtra,India. **M:** 9823269369, **E-Mail:** anne_cerry@yahoo.co.in

Conflict of interest: No

Case report is Original: YES

Whether case report publishes any where? NO

INTRODUCTION

Breast sarcomas are a very rare form of breast cancer, accounting for 0.2 to 1% of all breast malignancies, arising from the mesenchymal tissue of the breast.[1] They can be primary or secondary tumors. Etiology of primary breast sarcoma is often unknown. Secondary sarcomas can develop after radiation therapy or chronic lymphedema. Breast sarcoma is mostly diagnosed in patients who are in their fifth or sixth decade of life. Menopausal status may influence the development of breast carcinoma. The tumour grows more rapidly than the epithelial breast carcinomas and present as unilateral, well defined, mobile and painless lump. Metastases from breast sarcoma commonly spread haematogenously, to the lungs, bones and liver. Lymph node metastases are rare. Breast skin and the nipple areola complex are rarely involved.[2]

CASE REPORT

A 70 years old female presented to the out patient department with a complaint of lump in left breast which was gradually increasing in size since 20 years. She had no personal or family history of breast cancer and no history of exposure to radiation. On physical examination a mass of 20 x 17 cm was palpated involving the entire breast. It was hard and non mobile. No axillary lymph nodes were palpated. Fine needle aspiration cytology of the lump revealed round to oval cells with prominent nucleoli with scanty cytoplasm and mild anisonucleosis (Figure 1). Cytological diagnosis of malignancy was given. The patient then

underwent a simple mastectomy. On gross examination the cut surface of the tumour was solid grayish white in appearance (Figure 2). Histopathology revealed oval to spindled pleomorphic cells arranged in fascicles and lobules separated by dense collagen bands (Figure 3). Few mitotic figures were seen. Ductal epithelium was not seen. Provisional diagnosis of spindle cell tumour was given with the differentials of Fibrosarcoma, Malignant peripheral nerve sheath tumour and Myofibroblastoma. On Immunohistochemistry the neoplastic cells only expressed Vimentin and were negative for EMA, pan CK, S-100 protein, Calponin, CD34, CD31, p63, Desmin and CK7. The final diagnosis of Primary Low grade Spindle cell sarcoma of the breast was given. The patient was then referred to the oncologist for further follow up.

Figure 1: Photomicrograph of FNAC stained with MGG showing round to oval pleomorphic cells.

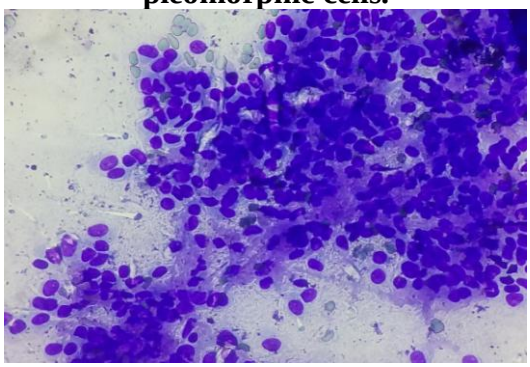


Figure 2: Simple mastectomy of the breast showing the cut surface of the tumour which is solid grayish white.

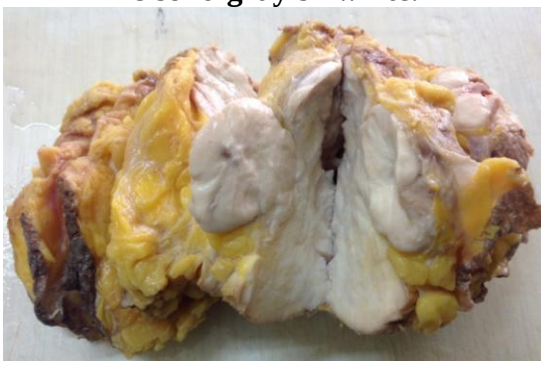
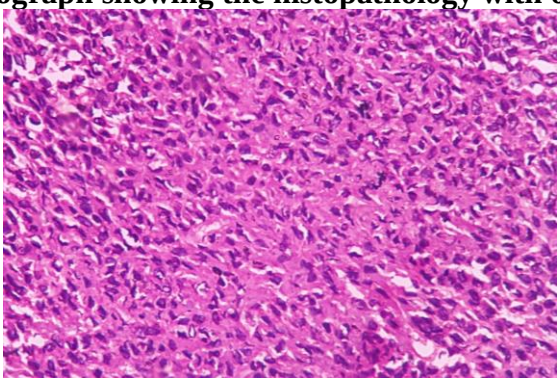


Figure 3: Photomicrograph showing the histopathology with oval to spindled cells.



DISCUSSION

Sarcomas can occur anywhere in the body but are most commonly seen in the extremities or intra-abdominal sites.[1] Breast sarcomas are rarely seen. Primary sarcoma of the breast is an extremely rare lesion than sarcomas arising secondary to radiotherapy.

Mammography in breast sarcomas is non specific and demonstrates a non speculated dense mass without microcalcifications.[3] Ultrasound is better than mammography in the diagnosis of breast sarcomas and appears as hyperechoic with no shadowing.[4] MRI demonstrates low signal intensity on T1-weighted images, and higher signal intensity on T2-weighted images.[5]

Staging and treatment of breast sarcomas differ from other types of breast cancer. Lymph node status is not as important in staging breast sarcomas as in other kinds of breast cancer. When sarcomas spread, they typically do not travel through the lymphatic system. Even in the case of large breast sarcomas, the lymph nodes are usually negative for cancer, and axillary node dissection is usually not required.

The most commonly used staging system for breast sarcomas is the American Joint Committee on Cancer staging system (Table 1) for soft tissue sarcoma.[6] Histological grade, tumour size, nodal involvement and distant metastasis contribute to this system.

Table 1: Staging system for soft tissue sarcomas of the breast

Stage	Histological grade	Size	Location (relative to fascia)	Systemic/metastatic disease present
IA	Low	< 5 cm	Superficial or deep	No
IB	Low	> 5 cm	Superficial	No
IIA	Low	> 5 cm	Deep	No
IIB	High	< 5 cm	Superficial or deep	No
IIC	High	> 5 cm	Superficial	No
III	High	> 5 cm	Deep	No
IV	Any	Any	Any	Yes

Our case was of low histological grade with size more than 5 cm and located superficial to the fascia, without any metastasis. Hence it was stage IB. She had no history of exposure to radiation or any previous breast pathology. Hence it was a case of primary low grade spindle cell sarcoma of the breast.

Treatment of breast sarcoma depends on the histological type, degree of differentiation, tumor size and advancement of disease. Wide local excision without axillary dissection yields good local control. The role of radiation and chemotherapy in the treatment of breast sarcoma is unclear. Some authors suggest that radiation is efficacious after surgical resection particularly with microscopically involved margins however this was not supported by other studies.[1]

Differential diagnosis of spindle cell lesions in breast include:- fibromatosis, nodular fasciitits, fibrosarcoma, inflammatory myofibroblastic tumour, myofibroblastoma, malignant

fibrous histiocytoma, leiomyoma, leiomyosarcoma, nerve sheath tumour, spindle cell carcinoma, phyllodes tumour, solitary fibrous tumour, hemangiopericytoma, dermatofibrosarcoma protuberance, malignant melanoma as well as metastatic tumours with spindle cell morphology. Immunohistochemistry may be useful to rule out other tumors such as non-mesenchymal malignant tumors or sarcomas with a specific line of differentiation. Desmin, vimentin, smooth muscle actin, keratin, leukocyte common antigen, CD34, HMB45, EMA, and S-100 should be analyzed.[7,8]

In primary breast sarcoma prognostic factors are adequate surgical excision, tumor grade and tumor diameter. Tumor grade is reported to be the single most important pathologic prognostic factor in soft tissue sarcomas. Mutation of p53 and a high Ki-67 proliferation index are associated with poor prognosis. 5-year survival rates for patients with breast sarcoma range from 40 to 91%.[1]

CONCLUSION

Majority of the invasive breast neoplasms are epithelial tumors while mesenchymal breast tumors are very rare. It is important that this lesion is recognized as a separate entity due to the difference in management and behavior of the breast sarcomas from the more common epithelial carcinomas.

REFERENCES

1. Cil T, Altintas A, Pasa S, Buyukbayram H, Isikdogan A. Primary spindle cell sarcoma of the breast. Breast care 2008; 3:197-99
2. Al-Benna S, Poggemann K, Steinau H, Steinstraesser L. Diagnosis and management of primary breast sarcoma. Breast cancer res treat 2010; 122:619-26
3. Elson BC et al. Fibrosarcoma of the breast: mammographic findings in five cases. Am J Roentgenol 1992; 158:993–95
4. Pencavel TD, Hayes A. Breast sarcoma - a review of diagnosis and management. Int J Surg 2009; 7:20–23
5. Yang WT et al. Mammary angiosarcomas: imaging findings in 24 patients. Radiology 2007; 242:725–34
6. Greene FL, Fritz AG, Balch CM. AJCC cancer staging manual. Soft tissue sarcoma. Springer-Verlag, New York 2002
7. Abd El All HS. Breast spindle cell tumours: About eight cases. Diagnostic Pathology 2006; 1:13
8. Wei S, Hameed O. Spindle-cell lesions of the breast with emphasis on the differential diagnosis. The open breast cancer journal 2010; 2:95-100