



SPINDLE CELL SARCOMA OF BREAST : A CASE REPORT

Muktanjalee Deka

Associate professor, Department of Pathology , Gauhati Medical College and Hospital

Bini Borah*

Post graduate trainee, Department of Pathology , Gauhati Medical College and Hospital *Corresponding Author

ABSTRACT Primary spindle cell sarcoma is a rare neoplasm constituting 0.2-1% of all mammary malignancies comprising of <5% of all soft tissue tumors and spindle cell sarcomas being one of the most common subtypes . Here we report a case of 38 years old female with swelling in the left breast for a period of 2 years . FNAC was performed and was diagnosed as malignant spindle cell lesion with differential diagnosis of metaplastic carcinoma of breast and malignant phyllodes tumor . The patient underwent simple mastectomy and after both histopathological and immunohistochemical examination of the post-mastectomy specimen , the case was reported as intermediate grade spindle cell sarcoma of breast . Four months later patient was readmitted with recurrence of the tumor mass at the mastectomy site and the tumor was again diagnosed as intermediate grade spindle cell sarcoma as before . Primary spindle cell sarcoma often carries a poor prognosis . Combination of histopathological and immunohistochemical examination is mandatory for diagnosis .

KEYWORDS :**Introduction**

Primary sarcoma of breast is a rare neoplasm and constitute 0.2-1% of all mammary malignancies , comprising of <5% of all soft tissue tumors^{3,7}. It usually occurs in 4th to 5th decades of life and mean age is around 40 years⁷ . It shows female preponderance and male cases usually represent less than 5% of primary breast sarcomas⁷ . Spindle cell sarcoma is one of the most common subtypes⁶ .

Case Report

We present a case of 38 years old female, who presented with a swelling in the left breast for a period of 2 years, which was insidious in onset, gradually progressive in size with no associated pain, ulceration or nipple discharge along with no family history. Physical examination revealed a non tender, firm to hard palpable mass in the left breast. Subsequently FNAC was performed and on microscopic examination, cellular smears showed hyperchromatic pleomorphic spindle cells, both in clusters as well as singly dispersed in a background of abundant myxoid stroma. The case was diagnosed as malignant spindle cell lesion with the differential diagnosis of metaplastic carcinoma of breast and malignant phyllodes tumor. The patient underwent simple mastectomy and the specimen was sent to department of pathology, Gauhati Medical College and Hospital, for histopathological examination. On gross examination of the specimen, a tumor mass was detected measuring (20×15×12) cm³. Cut surface of the tumor mass is well circumscribed, solid, grayish white. Histopathological examination showed spindle to stellate cells with fascicular arrangement at places. Cells were hyperchromatic showing moderate pleomorphism and delicate cytoplasm. Delicate vasculature noted in the background. The tumor had infiltrative edge. Mitotic count was 10/10 HPF. Epithelial elements not seen. The following differential diagnosis were considered – 1) malignant phyllodes tumor 2) metaplastic spindle cell carcinoma 3) monophasic synovial sarcoma 4) myoepithelial carcinoma. Immunohistochemical examination was performed with tumor cells showing diffuse and strong vimentin positivity and EMA, desmin, ER, PR, Her2neu, S-100, calponin negativity. Immunostaining with Ck was done three times showing negative results. Both histopathological and immunohistochemical findings ruled out the above mentioned differentials. Therefore the tumor was reported as intermediate grade spindle cell sarcoma of breast.

Four months later, the patient was readmitted with recurrence of tumor mass at the mastectomy site. Revision mastectomy with excision of the mass was performed. On gross examination, a tumor mass measuring (8.5×5.5×5) cm³ was noted with cut surface of the mass being well circumscribed, solid, grayish white

Microscopic examination of the tumor was suggestive of the recurrence of intermediate grade spindle cell sarcoma of breast.



Fig-1 : A well circumscribed tumor mass, Solid, grayish white in colour



Fig-2: 100X: Spindle cells are arranged in clusters as well as singly dispersed in a background of myxoid stroma (MGG stain)

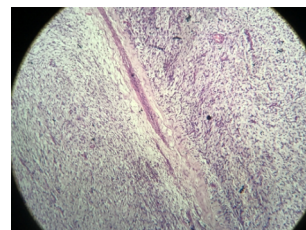


Fig-3: 100X: Cells are arranged in fascicular pattern (H & E stain)

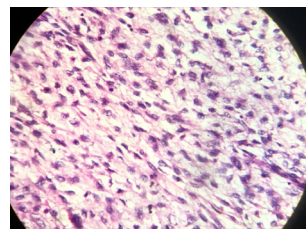


Fig – 4 : 400X : Hyperchromatic moderately pleomorphic spindle cells .Mitoses are seen in the background (H & E stain)

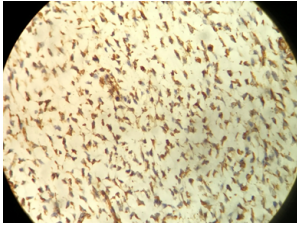


Fig-5: Shows strong vimentin positivity

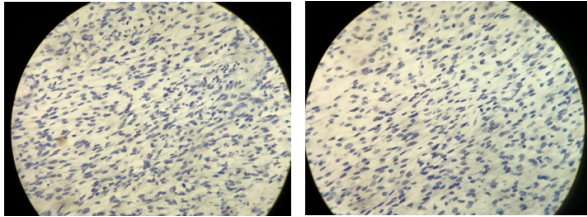


Fig – 6 : ER negativity

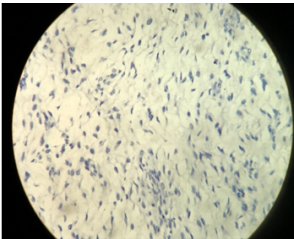


Fig – 7 : PR negativity

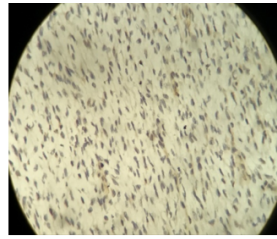


Fig – 8 : Her2neu negativity

Fig – 9 : Ck negativity

Discussion

Mammary sarcomas are heterogenous group of malignant neoplasms that arise from mammary stroma with spindle cell sarcoma being a descriptive subtype. Other descriptive subtypes are myxoid sarcomas, pleomorphic sarcomas and small round cell sarcomas³. They can arise as a primary lesion, or more commonly secondary to radiotherapy³. The use of radiotherapy increases the risk of development of angiosarcomas, pleomorphic undifferentiated carcinomas and other rare types of sarcomas within the field of irradiation¹⁰.

They usually present with gradually increasing mass typically without any pain or overlying skin changes⁸. As a group, mammary sarcomas vary greatly in size, ranging from less than 1 to 30 cm or more. In most studies, the mean and median sizes fall between 4 to 7 cm. The gross appearance of the tumor is influenced in part by the specific histological characteristics of the lesion, but the specimens typically consist of fleshy, moderately firm, pale tissue with varying amounts of haemorrhage and necrosis. Most sarcomas appear well circumscribed grossly, even if the border is invasive histologically¹⁰.

Breast sarcomas are often refractory to diagnosis by conventional triple assessment. Mammographic findings in sarcoma are non specific. Depending on the type, it may appear as a nonspiculated dense mass with indistinct borders to areas of asymmetry or even no mammographic abnormality at all¹¹. Ultrasonography usually reveals an irregular hypoechoic mass, with indistinct margins and posterior acoustic shadowing⁷. MRI in breast sarcomas can indicate malignancy. The tumors are lobulated and display rapid enhancement and so called “wash out” characteristics, all of which may be helpful in suggesting the diagnosis of malignancy¹¹. But overall, primary breast sarcomas have no pathognomonic imaging features³. On FNAC, features of breast sarcomas can mimic other spindle cell lesions like phyllodes tumor or metaplastic carcinoma. That is why histopathology is of utmost importance for diagnosis of sarcomas. But at the same time, even histopathologically, features of spindle cell sarcomas can overlap with other malignant neoplasms

like metaplastic spindle cell carcinoma, myoepithelial carcinoma, malignant phyllodes tumor etc. Also, morphology of some of the histopathological types of spindle cell sarcomas show extensive similarities with each other. These factors mandate the importance of immunohistochemistry. But it is to be noted that, sarcomas like monophasic synovial sarcomas can be difficult to differentiate from other spindle cell sarcomas based on histopathological and immunohistochemical profiles only. Therefore, molecular studies like break-apart FISH and RT-PCR of fusion transcripts have been employed for accurate diagnosis¹².

The mainstay of treatment is surgical, generally comprising of either wide local excision or simple mastectomy depending on the size and the location of the tumor. Routine axillary lymph node dissection is not indicated since sarcomas predominantly exhibit haematogenous spread⁹. Also, most studies show that, at the very least, adjuvant radiotherapy tends towards better survival, but the role of chemotherapy is less clear.

Primary spindle cell sarcomas often carry a poor prognosis with prognostic factor depending mostly on the adequate surgical excision, tumor grade and tumor diameter. Studies show that tumor size >5 cm, often carries a poor prognosis². The reported 5 year survival rate for patients with breast sarcomas range from 40 to 91%³.

Conclusion

Primary spindle cell sarcoma is a rare neoplasm, that needed to be diagnosed early for prompt treatment and adequate surgical resection for patient survival. The combination of histopathological, immunohistochemical and in some cases, molecular studies is mandatory for diagnosis.

REFERENCES

- Adem C, Reynolds C, Ingle JN, Nascimento AG. Primary breast sarcoma : Clinicopathologic series from the mayo clinic and review of the literature. *Br J Cancer* . 2004;91(2):237-47.
- Al Benna, S., Poggemann, K., Steinar, HU. et al. *Breast Cancer Res Treat* (2010) 122: 619.
- Cil, Timucin et al. “Primary Spindle Cell Sarcoma of the Breast“. *breast care* 3.3(2008): 197-199.
- Dimosthenis Miliaras and Emmanouel Konstantinides, “Malignant Fibrous Histiocytoma of the Breast: A Case Report,” *Case Reports in pathology*, vol 2012, Article ID 579245, 3 pages, 2012.
- Eur J Radiol*. 2017 Mar; 88: 1-7. doi: 10.1016/j.ejrad.2016.12.020. epub 2016 Dec 21
- Gesakis, K., Tanos, G., Onyekwelu, O., Gaitis, A., Gudur, L., & Agarwal, A. (2014). Primary Spindle Cell Sarcoma of the Breast Masquerading as Necrotizing Fasciitis. *Journal of Surgical Case Reports*, 2014.1(2014), rj096.
- Moore MP, Kinne DW. *Breast Sarcoma*. *Surg Clin North Am*. 1996; 76:383-392
- Pandey M, Mathew A, Abraham EK, Rajan B. Primary sarcoma of the breast. *J Sur Onc*. 2004; 121-125
- Pollard SG, Marks PV, Temple LN, Thompson HH. Breast sarcoma. A clinicopathological review of 25 cases. *Cancer* 1990;66:941-4.
- Rosen's *Breast Pathology*, fourth edition
- Timothy D. Pencavel, Andrew Hayes, Breast sarcoma – a review of diagnosis and management, *International journal of Surgery*, Volume 7, Issue 1, Pages 20-23, ISSN 1743-1991
- WHO classification of Tumours of Soft Tissue and Bone, 4th edition