Malignant peripheral nerve sheath tumor mimicking breast cancer: a case report

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European Journal of Medical Case Reports
Volume 7(8):164–166
https://doi.org/10.24911/ejmcr/173-1677945890

ABSTRACT

Background: Malignant peripheral nerve sheath tumor (MPNST) is a rare variety of soft 10-tissue sarcoma of ectomesenchymal origin. It represents approximately 10% of all soft tissue sarcomas. Though these tumors due to their Schwann cell origin may occur anywhere near a nerve trunk, breast is a rare site of its occurrence. This is a rare case of MPNST arising in the breast and not associated with neurofibromatosis type 1.

Case Presentation: A 22-year-old female presented with a large painless breast lump for 5 months, which was clinically suspected to be a Carcinoma. No confirmatory results were 18 obtained with fine needle aspiration biopsy , core biopsy, and magnetic resonance imaging. Wide local excision was done. Histopathology and immunohistochemistry confirmed MPNST. Furthermore, she underwent adjuvant radiation to her breast.

Conclusion: The unusual location and size of the tumor make our study noteworthy. Owing to the infrequent occurrence of the tumor, there are no definite guidelines for its management. We recommend wide local excision with postop radiotherapy as the optimum treatment for this condition.

Keywords: Malignant peripheral nerve sheath tumor MPNST, soft tissue sarcoma, breast, immunohistochemistry, wide local excision.

Received: 04 March 2023 **Accepted:** 25 August 2023 **Type of Article:** CASE REPORT **Specialty:** Oncology

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Background

Sarcomas are a rare type of breast malignancy and malignant peripheral nerve sheath tumors (MPNSTs) of the breast are even rarer. There are no specific clinical and radiological features for the diagnosis of this tumor and histological features are also reported to be nonspecific. Therefore, immunohistochemistry (IHC) is required for its diagnosis. A definitive treatment protocol is unavailable because of its rarity.

Case Presentation

Our patient, a 22-year-old female presented with a rapidly enlarging painless left breast lump for 5 months. There were no other breast-related symptoms. She had no previous significant surgical or medical history. Family history suggestive of any breast disease was negative on clinical examination; there was a 16 × 11 × 12 cm mass on the left upper outer quadrant. It was hard, nontender, fixed, and attached to the pectoralis major muscle with well-defined margins. There were no visible veins and the overlying skin was normal. Magnetic resonance imaging (MRI) revealed a breast mass with type-II kinetic curve characteristics. Fine needle aspiration biopsy (FNAC) suggested inflammatory changes, while core biopsy suggested a malignant spindle cell tumor. Wide local excision was planned to confirm the diagnosis and guide further management. The excised specimen consisted of a well-circumscribed, firm, solid mass measuring 16 × 10.5 × 12 cm. The cut section revealed a grayish-white myxoid mass without necrosis or calcification. Histopathology revealed a highly cellular spindle cell tumor with alternate hyper and hypocellular areas arranged in a fascicular pattern with nerve-like whorl formations. Brisk mitotic activity and hemangiopericytoma pattern were noted. MPNST was diagnosed. IHC revealed diffuse positivity for Vimentin and S-100 and 20%-30% for KI-67. The aforementioned findings confirmed MPNST. Clinical criteria were inadequate for diagnosis of neurofibromatosis 1. There were no café-au-lait spots/freckles. Mutational analysis was negative for p53, RB1, CDKN2A, and NF-1 associated loss of domain. The patient underwent radiotherapy and 5 years into follow up, no local recurrence or distant metastasis has occurred. Initially, a clinical examination for 3 months for 2 years, followed by 6 months for 5 years was done, supplemented by a yearly MRI for 5 years. The clinical examination consists of local site (both breast, axilla)

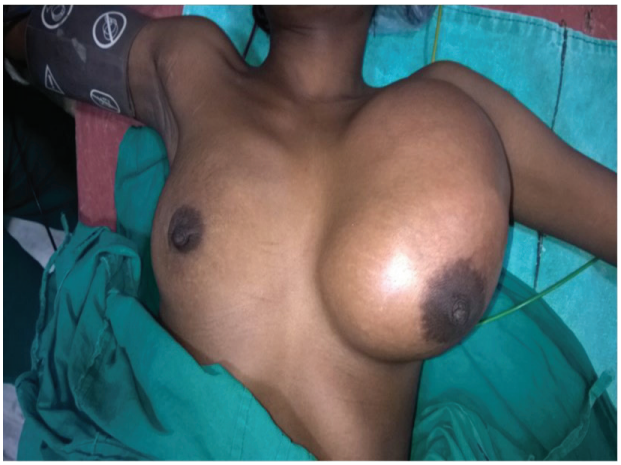

Figure 1. *Clinical photograph showing left breast mass.*

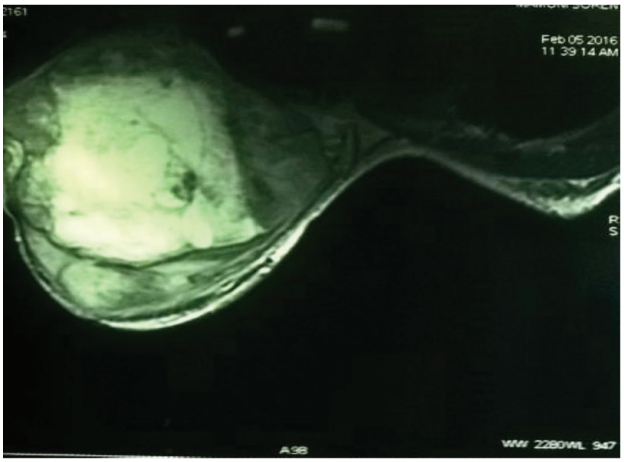

Figure 2. *MRI breast is done in the preoperative period.*

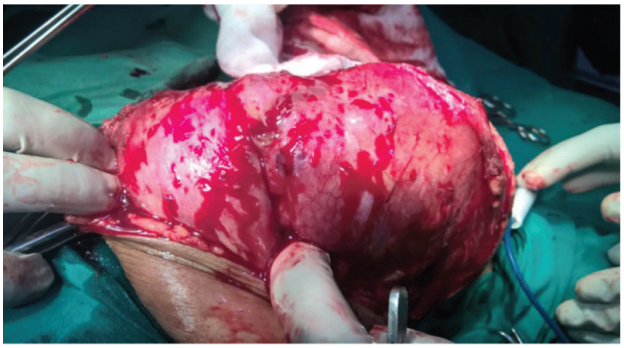

Figure 3. *Intraoperative photograph of the mass; separated from skin.*

examination, skin inspection for neurofibroma, and ocular examination for lisch nodules/optic nerve pathology. She is doing well now.

Discussion

MPNST is the preferred term for tumors originating from peripheral nerves or from cells associated with the nerve sheath. It is a rare variety of soft tissue sarcoma of ectomesenchymal origin [1,2]. One-fourth to half of the cases reported are associated with neurofibromatosis

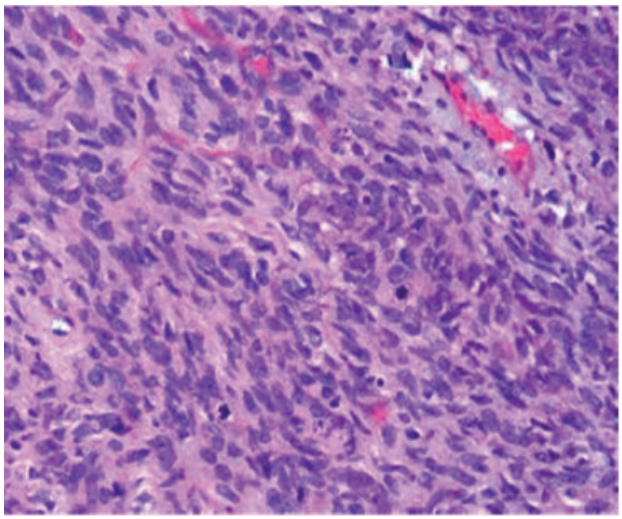

Figure 4. *Histopathology showing highly cellular spindle-like tumor with nerve arranged in whorls. IHC showed positive immunostaining for S-100, Vimentin, and Nestin.*

1 syndrome. The most common sites of occurrence of MPNSTs are proximal portions of the trunk (46%), upper and lower extremities 72 (34%), and head and neck region (19%) [3,4]. Though these tumors due to their Schwann cell origin may occur anywhere near a nerve trunk, breast is a rare site of its occurrence [5]. Limited cases have been reported in the literature. MPNSTs have to be set apart from other soft tissue sarcomas. MPNSTs of breast present as a hard lump which may clinically be confused with fibroadenosis or primary breast carcinoma. FNAC and core biopsy suggest a diagnosis of a soft tissue sarcoma concomitantly differentiating it from fibroadenosis and breast carcinoma. To tell apart, the various types of soft tissue sarcomas histopathologic examination is done. The absence of a leaf-like intra canalicular pattern rules out phyllodes tumor and the absence of a herringbone pattern excludes fibrosarcoma whereas the presence of spindle cells in fascicular 84 pattern with wavy nuclei suggests MPNST as a diagnosis in this case. IHC evaluation is the confirmatory tool that can establish the diagnosis of MPNST as shown by the positivity of vimentin, S-100, and KI-67 in our case. Immunohistochemical staining being negative for epithelial membrane antigen, desmin, calponin, and 88 smooth muscle actin rules out rhabhdomyosarcoma, synovial sarcoma, and leiomyosarcoma. Treatment is complete surgical excision of the tumor with negative margins along with radiotherapy [6]. Dissection of the axillary tail is not the protocol as the mode of dissemination is primarily hematogenous [7].

Conclusion

MPNST arising in the breast is uncommon. MPNST should be considered in the differential diagnosis when a sarcomatous lesion is found in the breast due to its high risk of recurrence and dismal prognosis [8,9]. The unusual location and size of the tumor make our study noteworthy.

Owing to the infrequent occurrence of the tumor, there are no definite guidelines for its management. We recommend wide local excision with postop radiotherapy as the optimum treatment for this condition.

What's new?

Primary MPNST of the breast is an extremely rare variety of soft tissue tumors of ectomesenchymal origin. The unusual location and size of the tumor make our study noteworthy. Owing to the infrequent occurrence of the tumor, there are no definite guidelines for its management. We recommend wide local excision with postop radiotherapy as the optimum treatment for this condition.

List of Abbreviations

FNAC	Fine needle aspiration biopsy
IHC	Immunohistochemistry
MPNST	Malignant peripheral nerve sheath tumor
MRI	Magnetic resonance imaging

Conflict of interest

The authors declare that they have no conflict of interest regarding the publication of this report.

Funding

None.

Consent for publication

Informed consent was obtained from the patient.

Ethical approval

Ethical clearance was taken from the Institutional committee.

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Summary of the case

1	**Patient (gender, age)**	Female/22 years
2	**Final diagnosis**	MPNST left breast
3	**Symptoms**	Painless progressive mass left breast
4	**Medications**	None
5	**Clinical procedure**	Wide local excision
6	**Specialty**	Breast oncology