

# Malignant peripheral nerve sheath tumor of the breast with epithelial elements: a case report

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## Abstract

MPNST is the preferred term for sarcomas of nerve sheath type because of the heterogeneity of their constituent cells and their inconsistent or poorly reproducible relationship to conventional benign schwannoma or neurofibroma. Common sites include deeper softtissues, usually in the proximity of a nerve trunk. Sporadic MPNST per se arising from the breast is a rare condition in the absence of NF-I. Only a few cases have been documented in the breast. The present of epithelial elements is also a rare finding? We are presenting a case of MPNST of the breast with epithelial elements, which was pan cytokeratin positive, for its rarity.

**Keywords:** MPNST, Breast.

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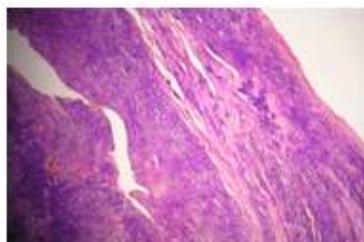
## CASE PRESENTATION

A 40 years old female presented with a painless lump over the left infra axillary region, which was initially thought to be a lipoma. Surgical excision was done and specimen was sent for HPE. Histopathological evaluation revealed a malignant peripheral nerve sheath tumor amidst of breast tissue and also showed islands of epithelial elements. A computed tomography was done

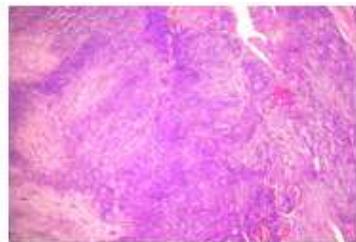
and showed no evidence of metastasis to thorax and abdomen. Immunohistochemistry showed positivity for pan cytokeratin.

## BACKGROUND

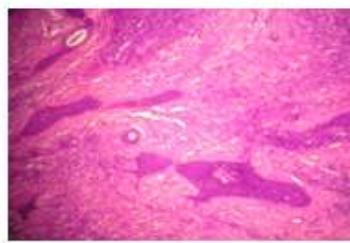
Malignant tumors arising from or displaying differentiation along the lines of the various elements of the nerve sheath (e.g., Schwann cell, perineural cell, fibroblast) are collectively referred to as malignant peripheral nerve sheath tumors (MPNSTs). The diagnosis of MPNST has traditionally been one of the most difficult and elusive among soft tissue tumors in the past because of a lack of standardized diagnostic criteria. Even today, there are no specific biomarkers that can be used to establish the diagnosis with certainty. MPNST of the breast is a rare entity. Only a few cases have been documented so far in the breast. Rarely epithelial squamous islands are encountered.



**Figure 1:** Spindle cell lesion with mammary gland lobular ductules



**Figure 2:** Showing hyper chromatic, pleomorphic spindle cells with increased mitoses



**Figure 3:** Showing epithelial squamous islands (X10) H and E



**Figure 4:** IHC showing Pan cytokeratin positive for epithelial elements (X10)

## DISCUSSION

MPNSTs account for approximately 5% to 10% of all soft tissue sarcomas, and about one-fourth to one-half occur in the setting of neurofibromatosis 1 (NF1)<sup>1</sup>. Primary sarcomas of the breast account for less than 1% of all primary breast neoplasms<sup>2</sup>. They should be distinguished from cystosarcoma phyllodes, sarcomatoid carcinoma and carcinosarcoma of the breast. Malignant nerve sheath tumours (neurofibrosarcoma, malignant schwannoma) have a variable histological pattern, which can include other Mesenchymal elements (Enzinger and Weiss 1983)<sup>3</sup>. Glandular, or epithelial, elements are much rarer (Ducataman *et al.* 1986)<sup>4</sup>. Immunohistochemical studies on these tumours have classed them as predominantly vimentin and S-100 positive (Morgan and Gray 1985)<sup>5</sup> and variably neurone-specific enolase positive (Matsunou *et al.* 1985)<sup>6</sup> in the sarcomatous areas. The epithelial glandular elements have been reported as cytokeratin and carcinoembryonic antigen (CEA) positive (Ducataman *et al.* 1986). The following antigen scan be used to identify nerve sheath differentiation: S-100 protein, Leu-7, myelin basic protein. S-100 immunoreactivity is focal and scattered only in 50–90% of MPNSTs; diffuse reactivity suggests a benign neural tumor. The other two antigens show immunoreactivity in approximately half of the tumors.

## CONCLUSION

Histopathology showed hyper chromatic, pleomorphic spindle shaped cells with increased mitotic figures (Fig 1 and 2). Immunohistochemistry showed S- 100 scattered positive and the epithelial squamous islands are positive

for pan cytokeratin (Fig 3 and 4). As far as our knowledge, this case presentation was a rare in view of MPNST of the breast with squamous islands.

## LIST OF ABBREVIATIONS

**MPNST:** Malignant peripheral nerve sheath tumor

**HPE:** Histo-Pathological evaluation

**NF:** Neurofibromatosis

**HandE:** Hematoxylin and Eosin

## COMPETING INTEREST

The Author(s) declares that they have no competing interest.

## REFERENCES

1. Enzinger FM, Weiss SW. Malignant tumours of peripheral nerves. In Soft Tissue Tumours. St Louis: C.V. Mosby, 2014; 855.
2. Pollard SG, Marks PV, Temple LN, Thompson HH (1990) Breast sarcoma. A clinicopathologic review of 2 cases. Cancer 66: 941–944.
3. Enzinger FM, Weiss SW. Malignant tumours of peripheral nerves. In Soft Tissue Turnours. St Louis: C.V. Mosby, 1983; 625-656.
4. Ducataman BS, Scheithauer BW, Piepgas DG, Reiman HM, IL Strup DM. Malignant peripheral nerve sheath tumours. A clinicopathologic study of 120 cases. Cancer 1956; 57; 2006-2021.
5. Morgan KG, Gray C. Malignant epithelioidschwannoma of superficial soft tissue? A casereport with immunohistology and electron microscopy Histopathology 1985; 9; 765-715.
6. Matsunou H, Shimoda T, Kakimoto S, Yamishitha, I Shikawa, Mukaim. Histopathologic and immunohistochemical study ofmalignant tumours of peripheral nerve sheath (malignant schwannoma). Cancer 1985; 56; 2269-2279.

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