



Case Report:

Malignant peripheral nerve sheath tumour: A rare tumour of the breast

Afeyodion Akhator, Senior lecturer, Dept. of Surgery, Faculty of Clinical Medicine, Delta State University, Abraka,
Osidi CP, Dept. of Surgery, Central Hospital, Warri; Assoc. Lecturer, Dept. of Surgery, Faculty Of Clinical Medicine, Delta State University, Abraka,

Inikori A, Senior Lecturer, Dept. of Radiology, Faculty of Clinical Medicine, Delta State University, Abraka,
Nwachokor FN, Dept. of Pathology, Central Hospital, Warri; Assoc. Lecturer, Dept. of Pathology, Faculty of Clinical Medicine, Delta State University, Abraka

Address For Correspondence:

Dr. Afeyodion Akhator,
Senior lecturer, Dept. of Surgery,
Faculty of Clinical Medicine,
Delta State University, Abraka.
E-mail: doc_akhator@yahoo.com

Citation: Akhator A, Osidi CP, Inikori A, Nwachokor FN. Malignant peripheral nerve sheath tumour: A rare tumour of the breast. *Online J Health Allied Scs.* 2010;9(1):9

URL: <http://www.ojhas.org/issue33/2010-1-9.htm>

Open Access Archives: <http://cogprints.org/view/subjects/OJHAS.html> and <http://openmed.nic.in/view/subjects/ojhas.html>

Submitted: Jan 26, 2010; Accepted: Apr 6, 2010; Published: Jul 30, 2010

Abstract:

Malignant peripheral nerve sheath tumour is a rare tumour of the breast and this report highlights the radiological and histological features and the difficulties in making a diagnosis. A high index of suspicion when dealing with a breast lesion that has both cystic and solid components in a background of von Recklinghausen's neurofibromatosis will make the diagnosis easier.

Key Words: Malignant peripheral nerve sheath tumour, von Recklinghausen's neurofibromatosis, breast

Introduction:

Primary carcinomas of the breast are predominantly of epithelial origin, and non epithelial tumours arising in the breast are very rare.¹ Malignant peripheral nerve sheath tumour (MPNST) accounts for 5-10% of all soft tissue sarcomas commonly in association with von Recklinghausen's neurofibromatosis (VRN), 4% incidence compared with 0.001% in the general population.² MPNST of the breast (with or without VRN) is a rare tumour with only a few cases reported in the literature.³

Case Report:

A 41 years old teacher presented to our surgical outpatient with a 4 year history of recurrent right breast mass. She had 2 excisions of the mass done 5years previously, 3 months apart, only to have it recur. It has increased rapidly in size to now involve the whole right breast. There was no breast pain, nipple discharge or family history of breast disease.

She had developed generalized body nodules 10 years previously during her first pregnancy. She is a Para 4+3, no use of contraceptives; breast fed her children for more than 1 year. There were no associated hearing or balance problems.

On examination, there were generalized multiple nodules on her skin, café au lait spots and some plexiform nodules. The right breast was larger than the left, with an oblique scar; the breast was completely taken up by a large multilobulated mass with hard and cystic components, measuring 15cm x 20cm, no nipple discharge and no palpable axillary lymphnode (Fig 1).

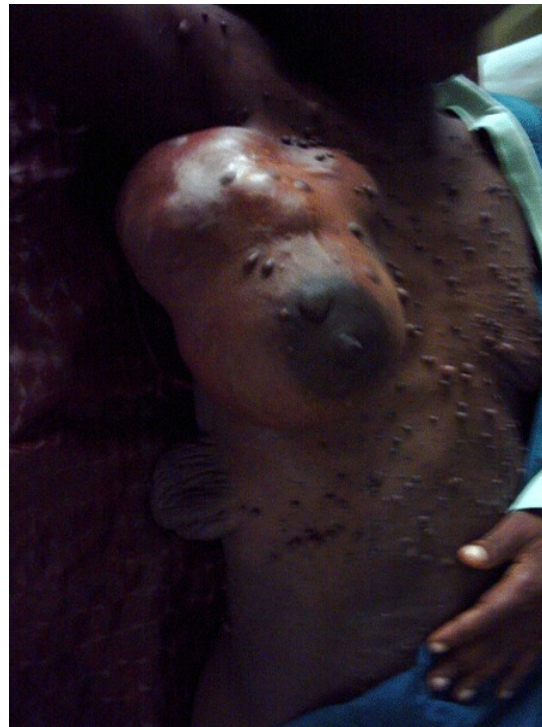


Fig 1: Café au lait spots, plexiform nodules and the multilobulated hard mass affecting the right breast

Assessment of Cystosarcoma phylloides in a patient with VRN was entertained. Patient was requested to do mammography, breast USS and FNAC.

Mammography showed multilobulated masses of varying soft tissue density and of varying sizes seen in the entire right breast, the largest measuring 8 x 10cm. Assessment of Giant fibroadenoma was made.

Breast USS showed multiple, predominantly hypoechoic masses with poorly defined margins seen in the entire right breast, with marked distortion of the parenchyma of the right breast. Some of these lesions show focal anechoic areas consistent with necrosis. Other show associated cystic lesions with posterior acoustic enhancement. The echotexture of the underlying pectoralis muscle is also distorted. Features are highly suggestive of a malignant breast mass.

FNAC – Smears show plump spindle, oval and round cells with little or no cytoplasm. There are no mitotic figures and nuclei are hyperchromatic. The cells form compact isolated sheets. Assessment was of benign spindle cell lesion.

Patient was counseled for mastectomy, at surgery the findings were multilobulated mass completely taken over the right breast involving both the pectoralis major and minor muscles. A modified radical mastectomy with axillary dissection was done.

Histology of the breast show a malignant mesenchymal tumour composed of compact sheets of proliferating spindle cells with moderate eosinophilic cytoplasm. The nuclei are spindle and with blunt ends. Some of the nuclei are round to oval in some areas. Some show attempts at rosette formation. They are arranged mainly in fascicles. Mitotic figures are moderate to frequent in numbers. Epithelioid areas are seen in some sections with rosette-like patterns. Vague pallasading with cellular and acellular areas are seen in some. There is also pleomorphism in some areas. Assessment of Malignant peripheral nerve sheath tumour (MPNST) in a background of Recklinghausens disease was made.

Patient was counseled for radiotherapy and chemotherapy, but she defaulted and returned 7 months later with recurrent tumour of the right breast, which was excised and she was also advised for adjuvant therapy and she has not been able to afford them.

Discussion:

MPNST is a rare sarcomatous tumour, previously known as neurofibrosarcoma, neurogenic sarcoma, malignant neurilemoma and malignant schwannoma. It is believed to arise from a nerve or neurofibroma and usually affects patients who are 20 – 50 years old but develops earlier in patients with VRN.⁵

The clinical diagnosis of MPNST is difficult because of its rarity. Because of the size and consistency of the tumour a diagnosis of cystosarcoma phylloides was entertained initially, but the background setting of VRN should have hinted on the possibility of MPNST.

The radiological features were also unhelpful in making this diagnosis with a discordant between the findings at mammogram and breast ultrasound. Woo et al described the problems of radiological diagnosis of MPNST⁶. The accuracy of cytology in preoperative diagnosis is uncertain and is difficult to distinguish it from benign or malignant spindle tumours⁴ as was in our case. Histological examination therefore remains the most important tool for diagnosis. The presence of spindle shaped cells arranged in dense cellular areas with rosette-like patterns and pallasading is suggestive of MPNST. Electron microscopy and immunohistochemistry is necessary for definite diagnosis of MPNST,⁵ However, these are not yet available in our facility.

Radical or modified radical mastectomy is the treatment for these lesions with post operative radiotherapy helpful in decreasing the local recurrence rate.⁶ Our patient could not afford to go for radiotherapy and had recurrence within 7 months.

References:

1. Majmudar B. Neurilemoma presenting as a lump in the breast. *South Med J.* 1976;69:463-464
2. Ramanathan RC, Thomas JM. Malignant peripheral nerve sheath tumours associated with von Recklinghausen's neurofibromatosis. *Eur J Surg Oncol* 1999;25:190-193
3. Thanapaisal C, Koonmee S, Siritunyaporn S. Malignant Peripheral Nerve Sheath Tumor of Breast in Patient without Von Recklinghausen's Neurofibromatosis: A case report. *J Med Assoc Thai* 2006;89(3):377-379.
4. Dhingra KK, Mandal S, Somak R, Khurana N. Malignant peripheral nerve sheath tumor of the breast: case report. *World Journal of Surgical Oncology* 2007;5:142. doi 10.1186/1477-7819-5-142.
5. Wanebo JE, Malik JM, VandenBerg SR, Wanebo HJ, Driesen N, Persing JA. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 28 cases. *Cancer*1993;71:1247-1253
6. Woo OH, Yong HS, Lee JB, Kim A et al. A giant malignant peripheral nerve sheath tumour of the breast: CT and pathological findings. *The British Journal of Radiology*2007;80:e44-e47. doi 10.1259/bjr/30984808