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Malignant Peripheral Nerve Sheath Tumor of the Anterior Chest Wall Presenting As a Breast Mass- A Case Report

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Abstract: Malignant peripheral nerve sheath tumor is a rare, aggressive tumor accounting for 5-10% of all soft tissue sarcoma. In the thorax they are usually located in the posterior mediastinum. We report a case of MPNST of the anterior chest wall presenting as a breast mass in a 22 year lactating woman. Diagnostic work-up included sonomammography, MR mammography, and core biopsy and was treated primarily by surgical resection.
Keywords: Malignant Peripheral Nerve Sheath Tumour, anterior chest wall, Sonomammography, MR Mammography, Immunohistochemistry.

INTRODUCTION:

Malignant peripheral nerve sheath tumour (MPNST) also known as neurogenic sarcoma, neuro fibrosarcoma and malignant schwannoma are the sarcoma arising from a peripheral nerve [1]. MPNST is a rare neoplasm that accounts for 5-10% of all soft tissue sarcomas and is often associated with Neurofibromatosis type 1 and previous radiation exposure [1, 2]. MPNST are highly aggressive affecting the head, trunk, and extremities with poor survival rate[3]. Neurogenic tumours in the thorax usually occur in the posterior mediastinum [4]. We report a case of MPNST of the anterior chest wall presenting as a breast mass in a 22 year lactating woman treated primarily by surgical resection

CASE REPORT:

A 22 year old lactating female patient presented in surgery outpatient department with a painful lump in the left breast for last 2 years. The lump was initially small, showing rapid progression for last 6 months. On clinical examination a large soft cystic mass was noted in the breast predominantly involving the upper outer quadrant and adjoining upper chest wall. The overlying skin was stretched up by the mass without any fixity. The mass was fixed to the underlying chest wall. No clinically palpable axillary or supraclavicular lymph nodes noted. She had no signs and symptoms or family history of NF-1.There was no past history of radiation exposure.

The blind core biopsy done outside 2 months back revealed a lactating adenoma. Sonomammography revealed a large (more than 5 centimetres), well defined heterogenous predominantly hypoechoic mass with cystic spaces and few septations in the upper outer quadrant of the left breast extending to the upper outer chest wall and adjoining axilla.

MR Mammography revealed a T2 hyper intense, well circumscribed mass with lobulated margin located in the retro mammary space extending to the upper outer chest wall and axilla. The mass is seen displacing the breast parenchyma anteriorly. It measures approximately 16.5 cm x 10.2cm x 11.69cm in size. The mass showed true peripheral diffusion restriction .No diffusion restriction was noted in the central part. Heterogenous peripheral rim enhancement with central area of low signal intensity was noted after intravenous administration of contrast. Pectoralis major muscle was thick, stretched and oedematous.MR Spectroscopy revealed a choline peak at 3.2 ppm.

Wide local surgical excision of the mass was done and the specimen was sent for histopathological examination and immunohistochemistry. Gross examination of the resected mass showed greyish white to greyish brown encapsulated soft tissue mass measuring 18cm x13cm x12 cm. On cut section, variegated appearance with large areas of necrosis was seen. One cystic space was noted measuring 6cm with an area of growth within it. Microscopy revealed histological features of a highly cellular spindle cell tumor with alternate hyper-cellular and hypo-cellular (myxoid) areas. Cells were arranged in sweeping fascicles with frequent nerve like whorl formations.

Individual cells showed wavy nucleus at places. A fair number of thick walled vessels, perivascular concentration of cells and hemangio pericytomatous pattern were seen. Brisk mitotic activity was noted. Areas of geographic necrosis and small areas of metaplastic cartilage were present.

Immunohistochemistry examination revealed that the tumor cells stained positive for S100 and Nest in consistent with the diagnosis of MPNST.

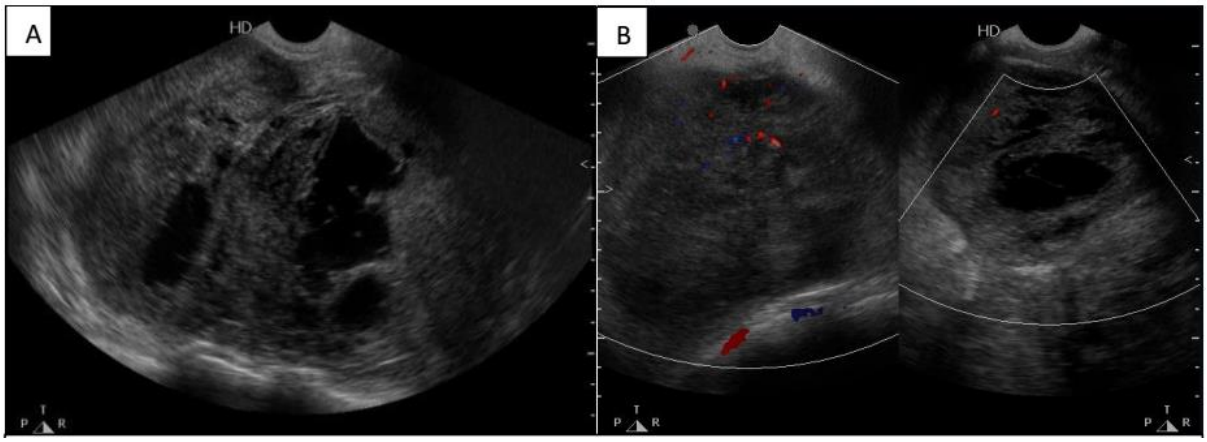

Fig 1: Malignant peripheral nerve sheath tumor in a 22 year old lactating woman with a palpable mass in the left breast. Sonomammography of the left breast shows: A.a large, well defined heterogenous predominantly hypoechoic mass with internal cystic spaces in the upper outer quadrant extending to the axilla; B. increased vascularity on color Doppler imaging.

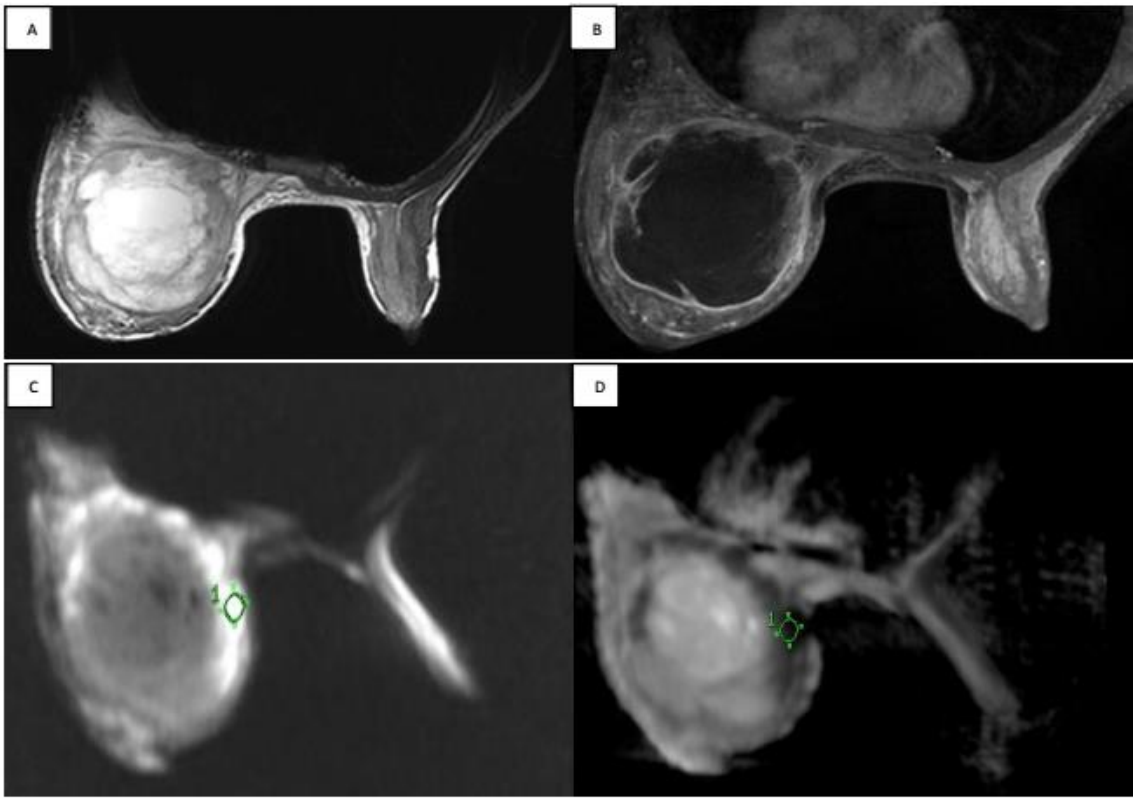

Fig 2: MR Mammography of both breast in a 22 year old lactating women with MPNST of the left breast.2A: Axial T2 weighted image reveals a well-defined high signal intensity mass with peripheral hypo intensity in the left retromammary space displacing the breast parenchyma anteriorly.2B: In axial post contrast MRI mass shows enhanced rim on the periphery and a central area of low signal intensity. Few enhancing septations were also noted.2C & D: Non-contrast DWI image obtained with a b-value of 800 shows peripheral high signal with corresponding low signal in ADC mapping.

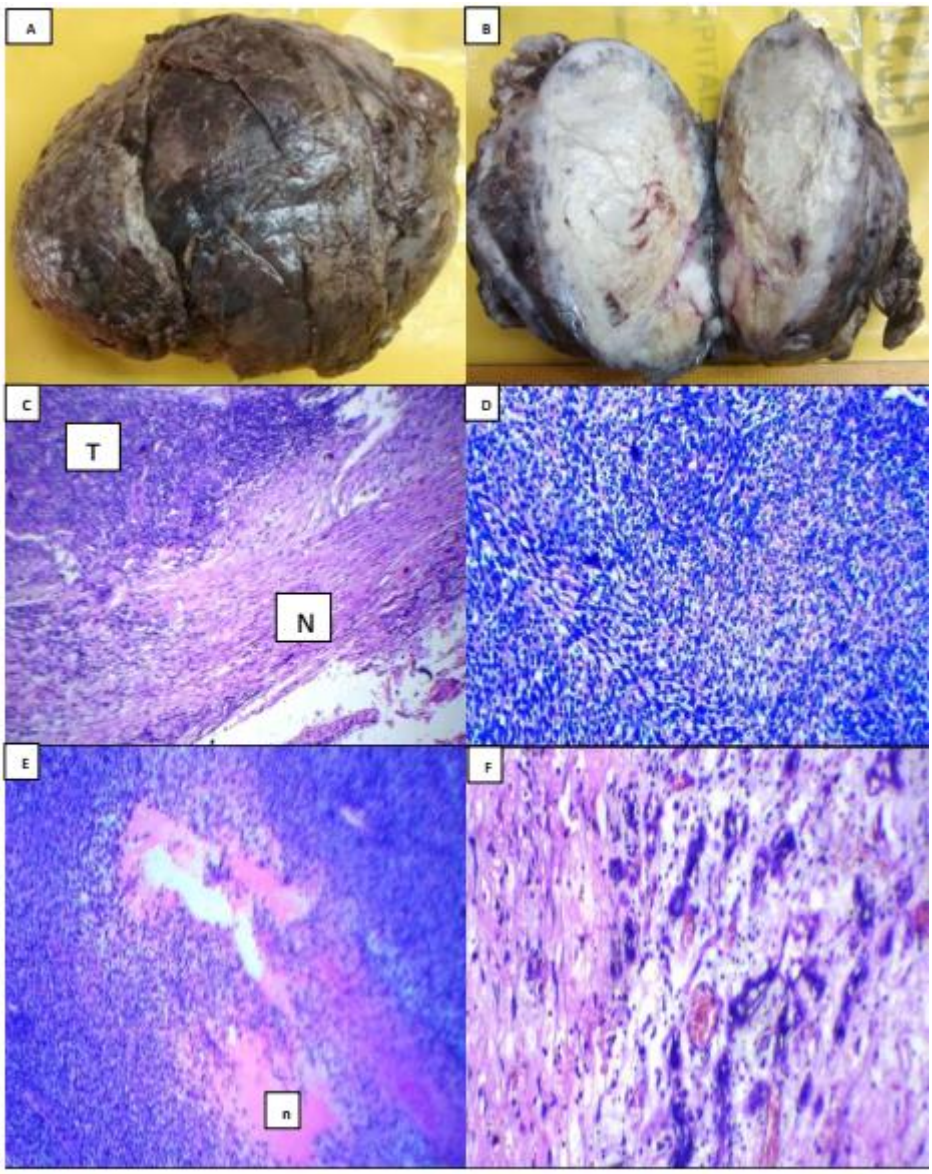

Fig 3: Photograph and microscopic appearance of tumor. A. Specimen of the excised tumor measuring 18cm x 13cm x 12 cm; B. Cut section showing whitish tumor mass with rim of breast tissue; C. Photograph showing areas of tumour (T) along with adjacent nerve bundle (N) [H & E, 100x]; D. High power view [H & E, 400x] showing whirling arrangement of cells; E. Figure showing areas of necrosis(n); F. Tumor with the normal breast tissue.

DISCUSSION:

Malignant peripheral nerve sheath tumor are aggressive, rare neoplasm arising from the peripheral nerve cells[5]. Variously known as malignant schwannoma, neurogenic sarcoma, neuro fibrosarcoma, MPNST is the current term used by the World Health Organization for a spindle cell sarcoma arising from nerve or neurofibroma or demonstrating a nerve tissue differentiation[6, 7]. It comprises 4-10% of all soft tissue sarcomas[1]. MPNST usually affects the head ,trunk, and extremities and have poor survival rate[3]. The risk of development of MPNST is higher in patients with von Recklinghausen's disease(NF-1) and with a history of previous irradiation[1]. The incidence of MPNST is 0.001% in the general population and 4.6% in patients with NF-1[1]. Usually patients are in age group of 20-50 years with equal sex predilection and presents with an enlarging mass, pain, symptoms of nerve deficit or a combination of these[1, 8, 9].

Primary soft tissue tumors of the chest wall are comparatively rare and imaging features particularly those which are locally aggressive are non-specific in most cases [10]. The role of imaging mainly lies in lesion localization and commenting on its morphology and tumor extent. In breast ultrasonography the lesions originating from the chest wall will displace the breast tissue anteriorly, may make an obtuse angle to the chest wall and a retro mammary fat layer is usually seen anterior to the lesion[11].

Neurogenic tumors of the chest wall which include schwannoma, neurofibroma, malignant peripheral nerve sheath tumor arises from the intercostal nerves, spinal nerve roots or sometimes distal branches of brachial plexus and show considerable overlapping imaging features[10]. In ultrasound schwannomas usually appear as homogenous eccentric mass with posterior acoustic enhancement, intratumoral cystic changes and may show increased vascularity on color doppler imaging while the neurofibromas appear as coarse echogenic mass with hypovascular pattern on color doppler imaging[11]. Most MPNST are homogenous and hypoechoic in echotexture and may show peripheral nerve congruity on ultrasonography. A sudden increase in size of the lesion with indistinct margins may suggest the malignant nature of the lesion[12]. Although irregular infiltrative margins along with intratumoral heterogeneity is more common in malignant nerve sheath tumor , it can also be present in benign neurofibromas[13]. Neurofibromas commonly contain multiple cystic spaces of varying sizes due to myxoid degeneration [14]. On MR Imaging neurogenic tumors show T2 hyperintensities with target like enhancement pattern. Peripheral enhancement is commonly observed in MPNST in comparison to focal central enhancement seen in benign neurogenic tumors [15].

In our patient the unusual location of the tumor lead to diagnostic dilemmas. Since the patient presented with a breast mass Sonomammography and MR mammography were performed to know the nature, size, location and extent of the mass. Sonomammography revealed a large heterogeneous mass with cystic spaces occupying upper quadrants of the left breast and extending to the left upper outer chest.MR mammogram allowed differentiation of the mass from benign entities like breast abscess .Although the exact origin of the tumor could not be well delineated MR mammogram showed that the tumor is not arising from the breast tissue rather the epicenter of the mass is lying in the retromammary fat plane in close proximity to pectoralis major muscle.

Histopathological examination and Immunohistochemistry is the mainstay for the diagnosis of MPNST. Histopathologically MPNST are characterized by spindle cells arranged in sweeping fascicles with frequent nerve like whorl formation associated with collagenous or myxoid matrix, hyper cellularity, nuclear polymorphism , brisk mitotic activity, perivascular concentration of cells and areas of geographic necrosis[1, 2, 16]. These tumors show focal S-100 positivity and diffuse positivity for vimentin[16]. Nest in, an intermediate filament protein is expressed in neuroectodermal stem cells is a sensitive marker of MPNST[17]. These features are consistent with our case.

The multimodality treatment of MPNST includes wide local excision with minimum 4cm safety margins, adjuvant chemotherapy and radiotherapy[18]. First line of treatment is surgical resection with complete removal of the tumor. Postoperative irradiation and chemotherapy are necessary in case of incomplete resection due to high risk of recurrence although they are often used as adjuvant therapy even if complete surgical excision with wide resection margins have been done[6]. Despite aggressive therapy, local recurrence (seen in 40% of patients) and distant metastasis (seen in 60% of patients) are common[7]. MPNST commonly metastasizes to the lungs, bone, pleura and retroperitoneum[7]. The overall 5-year survival rate for patients with MPNST without NF-1 is 50% but it drops to 10% for MPNST patients with NF-1[19].

In our patient a complete wide excision of the anterior chest wall mass was done .Histopathology showed negative surgical margins. Patient is asymptomatic with no evidence of recurrence till date.

CONCLUSION:

In conclusion the authors would like to highlight the diagnostic dilemma due to location of MPNST in the anterior chest wall presenting as a breast mass. Although neurogenic tumors are more common in the posterior mediastinum it should be included in the differential diagnosis of anterior chest wall tumors. The role of imaging is unreliable in differentiating benign from malignant nerve sheath tumor; presence of distant metastatic foci is a helpful sign along with sudden rapid growth and presence of pain on clinical examination. Cross-sectional imaging by MRI with multiplanar reformation is an important tool in localizing the lesion, commenting on its morphology (shape, margin and composition) and describing the extent. Histopathology and immunohistochemistry are the gold standard for diagnosis.

TEACHING POINT:

Anterior chest wall lesion may have clinical features suggestive of a breast lump because of the anatomic relationship between the chest wall and the breast. While evaluating a breast lump by sonomammography or MR mammography it is important to localize and characterize the lesion and describe its extent.

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