Malignant Peripheral Nerve Sheath Tumour of the Male Breast with Nodal Metastasis

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Abstract Malignant peripheral nerve sheath tumour (MPNST) is a rare soft tissue sarcoma with high rate of local recurrences and metastases. Common sites include deeper soft tissues, usually in the proximity of a nerve trunk. MPNST of breast is a rare occurrence. To our knowledge there are only less than 10 case reports of MPNST of breast. We present a young male patient with MPNST and lymph node metastases and also discuss diagnostic modes and management challenges in such rare difficult scenarios.

Keywords Malignant peripheral nerve sheath tumor of breast · Sarcoma · Male breast cancer

Introduction

Malignant peripheral nerve sheath tumour (MPNST) is a rare soft tissue sarcoma with high rate of local recurrences and metastases. Common sites include deeper soft tissues, usually in the proximity of a nerve trunk. MPNST of breast is a rare occurrence. To our knowledge there are only less than 10 case reports of MPNST of breast.

Case Report

A 17 year old male presented with a recurrent right breast lump post excision biopsy 2 months earlier which was report-

ed elsewhere as adenocarcinoma of the breast. He had no family history of breast cancer. On examination, a 6×6cm, well defined, lobulated, non tender mass with a scar overlying it on the lateral aspect. There was no skin fixity or ulceration (Fig. 1). Multiple mobile 1×1cm firm lymph nodes were palpable in the right axilla. There were no other subcutaneous swellings, café au lait spots, or Lisch nodules to suggest underlying neurofibromatosis.

Ultrasonography of the breast demonstrated a large, lobulated, markedly hypoechoic lesion with few necrotic areas and increased vascularity with multiple axillary nodes (Fig. 1). Chest radiograph and ultrasonography of the abdomen did not reveal metastasis. The core tissue biopsy was reported as poorly differentiated malignant neoplasm.

The patient underwent a wide excision of the mass including the nipple areolar complex and breast disc; the chest wall defect was reconstructed with a latissimus dorsi flap. Serial sectioning of the breast tissue showed large nodules, with a firm to hard, grey white cut surface. The axillary tail had 16 lymph nodes, with grey white to congested cut surface. On microscopic examination, breast parenchyma and lymph nodes were infiltrated by a tumour composed of sheets and nests of polygonal cells with moderate amounts of eosinophilic cytoplasm and moderately pleomorphic nuclei, fine chromatin and prominent nucleolus, surrounded by hyalinised septae. At some places, the tumour showed a peritheliomatous growth and are arranged in palisading pattern (Fig. 1). Mitotic activity was 4–5/10 hpf. Areas of stromal sclerosis and necrosis were present. Tumour cells were positive for S-100, CD56, CD99 and negative for pancytokeratin in immunohistochemistry and it was reported as MPNST (Fig. 1).

The patient received 50Gy adjuvant radiotherapy to the chest wall. One month later he developed a small 1×1 cm in the right gluteal swelling. Fine needle aspiration cytology was reported as MPNST confirming metastasis. Wide local excision was performed for palliation.

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Fig. 1 Image of the patient with the tumor. Ultrasound image of the tumor. Histopathology of the tumor. Immunohistochemistry image

Discussion

MPNST is the term coined by World Health Organisation to represent all tumours of peripheral nerve or nerve sheath origin. MPNST has replaced all other terms previously used such as malignant schwannoma, malignant neurilemmoma, neurofibrosarcoma. They constitute 10 % of all soft tissue sarcomas [1]. MPNST arise de novo in 50 % and in association with Neurofibromatosis 1 (NF1) in other 50 % cases, rarely also occur in sites of previous irradiation [2]. MPNST presenting as a breast lump is rare and its presentation in a male is even rarer [3]. Most studies have reported a mean age of presentation of 37 years (ranged from 9 to 84 years) with slight female preponderance [4].

Because of its rarity, it is difficult to diagnose MPNST of breast clinically and commonly it is misdiagnosed as phyllodes tumour because of its size and its cystic nature. Histopathological examination has a definitive role in the diagnosis rather than its radiological appearance or cytology [5].

Most of the tumours are greater than 5 cm in size as in the present case. Histopathology is characterized by spindle cells arranged either in palisading or spiral pattern. Heterologous and epithelioid differentiation is seen in 15 % of MPNST. Heterologous components includes rhabdomyoblasts, neuroendocrine, smooth muscle, cartilage and bone. The most

common heterologous component in MPNST is rhabdomyoblast differentiation but our patient had an epithelioid variant.

Immunohistochemical (IHC) expression helps to exclude other diagnoses. Commonly used neural markers includes S-100, CD56, PGP9.5 (protein gene product). Marker S-100 is not 100 % sensitive (50–90 % sensitivity) and CD 56, PGP 9.5 are sensitive but not specific. In high grade tumour cells, S-100 is sparsely positive. Nestin, an intermediate filament protein is proven to be more sensitive than other neural markers in recent studies [6].

Optimal treatment for MPNST is radical excision with a good margin of normal tissue which would involve mastectomy as in our case of recurrent MPNST. In critical areas where resection with wide margin of normal tissue is not feasible, excision should be combined with radiotherapy [7]. Chemotherapy has shown to have significant benefit in disseminated disease rather than localized disease. Regimes like doxorubicin – ifosfomide have been reported [8].

In reported cases with NF1, metastasis has been seen in 16 % patients and recurrences in 38 % patients. 5 year survival rate is 16 % in patients with neurofibromatosis and 53 % for patients without it. Radical resection improves survival than subtotal resection. Large tumours with size >5 cm and the presence of neurofibromatosis have unfavorable prognosis [7].

Conclusion

MPNST of breast is difficult to diagnose clinically but it requires high index of pathological suspicion to diagnose and confirm it by IHC. It needs early optimal intervention as it tends to recur locally and have poor prognosis.

Ethical statement There is no conflict of interest among any of the authors and no financial disclosures.

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