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Anterior Abdominal Wall Leiomyoma Arising De Novo in a Perimenopausal Woman – Diagnostic Enigma

Metaplastic Carcinoma of Breast – A Rare Tumour with Uncommon Presentation

Anticonvulsant and Anxiolytic Properties of the Leaves Extracts of *Cymbopogon Proximus*

Comparing Clinical Learning Effectiveness Among Lecture-Based Training, Simulation-Based Training and Training Using Animal Tissue Models

Generation Y (Gen Y) Issues in Medical Education at Private University in Shah Alam, Malaysia: Bridging the Gap

Anterior Cruciate Ligament: Single Vs Double Bundle

A Case Report of Kienbock's Disease in A Thirteen Year Old Girl

Can Witty Introductory Quotes Help Rivet Attention in Chemical Pathology?

Does Linking a Medical Learning Point to a Relevant Fictional Character Enhance Knowledge Acquisition?

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Metaplastic Carcinoma of Breast – A Rare Tumour with Uncommon Presentation

Krishna S; Naveen L; Shenoy R; Kadage V

Institution

Kasturba Medical College,
Manipal, Karnataka, India

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Abstract

Introduction: Metaplastic breast carcinoma (MBC) is a rare primary breast carcinoma, which is characterised by presence of carcinoma with co-existence of non-epithelial cellular elements. It comprises of 0.05 – 0.1 % of all breast carcinomas. Because of the rarity of the disease, there is paucity of information regarding the outcome of this variant of breast carcinoma.

Case: In our case, 45-year-old female presented with a large ulcerated lump in the left breast with foul smelling discharge and maggots. Patient presented in sepsis and hence she was taken for emergency surgery after initial resuscitation. Histopathology report was given as high grade MBC – squamous variant with triple negative receptor status.

Discussion: MBC is a rare variety of breast carcinoma which forms only 0.1% of all breast carcinomas. MBC usually has different characteristics compared to IDC with large tumour size, high-grade tumour, less nodal involvement, hormone receptor negativity and poor outcomes. Patients with MBC are to be treated more aggressively than IDC, but specific protocols for management of this type of carcinoma are not framed due to rarity of cases and presently are treated on the guidelines of IDC.

Conclusion: Although a rare variety of carcinoma, MBC is of considerable interest because of its pathological heterogeneity and differences in clinical behaviour compared to typical breast carcinomas. The poor disease free survival associated with MBC suggests that further research exploring mechanisms of carcinogenesis and identifying clinically relevant prognostic factors is needed to direct optimum clinical care.

Key Words

Metaplastic; Non-Epithelial; Squamous Cell; Sepsis; Mastectomy

Corresponding Author:

Dr Lella Naveen; E-mail: naveen_lella@yahoo.co.in

Introduction

Metaplastic Breast Carcinoma (MBC) is a rare heterogeneous group of malignant lesions comprising 0.05 – 0.1 % of breast carcinomas¹. The unique nature of MBC is that it has an epithelial or mesenchymal cell population mixed with adenocarcinoma. Pathologic classification of MBC is generally divided into two main groups:¹ homogeneous spindle cell/sarcomatoid carcinoma, and² heterogeneous carcinosarcoma/ carcinoma with sarcomatous differentiation (osseous, chondroid and rhabdoid). Pure epithelial malignant tumours with metaplasia, such as adenosquamous and pure squamous cell carcinoma, are also considered as MBC subtypes.^{1,2}

Case Presentation

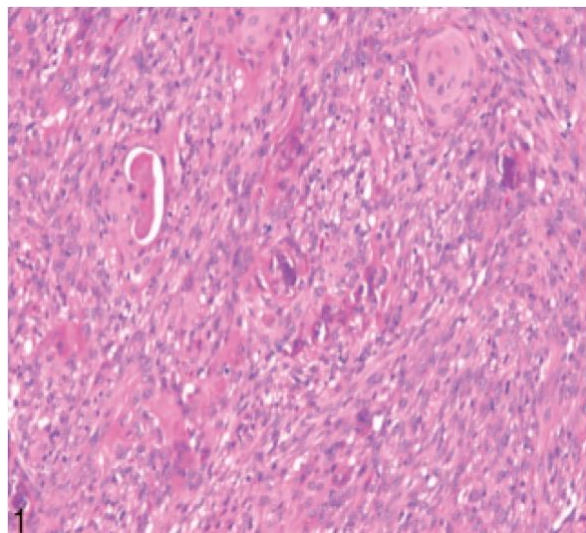
A 45-year-old female with no known comorbidities, presented to the casualty with a large lump in the left breast with ulceration and foul smelling discharge with maggots. Lump was there since 10 years, which was initially small, non-progressive in

size with no other complaints. The swelling increased in size over the past 1 – 2 months along with ulceration over the skin and discharge from the ulcer for past 10 days. She noticed maggots since two days and had low grade fever for one day.

On examination patient had fever (99.40F), tachycardia (pulse rate – 120/min) and blood pressure was 90/60 mm of Hg.

A large mass of size 15 x 10 cm involving almost all quadrants was present in the left breast. There is ulceration of skin over the lump of around 10 x 7 cm with necrotic tissue and foul smelling discharge. Hundreds of maggots were present in the ulcer. A lymph node of size 2 x 1 cm is present in the left axilla, which was firm, non-tender and freely mobile.

In view of the large size of the mass, our clinical diagnosis was malignant phylloids tumour with skin ulceration and sepsis.



Laboratory data showed leukocytosis with neutropilia.

As patient was in sepsis at presentation, she was taken up for emergency simple mastectomy after initial resuscitation and specimen was sent for histopathological examination.

Post-operative recovery was uneventful. Histopathological report showed high grade metaplastic carcinoma of breast, subtype – squamous cell carcinoma. Tumour cells are ER/PR and HER 2 neu negative.

After recovery from the acute situation, patient underwent metastatic workup with contrast enhanced CT of chest and abdomen and bone scan

which showed metastasis to L2 vertebra. Patient was started on palliative chemotherapy in view of stage IV disease.

Discussion

Metaplastic breast carcinoma is a rare variety of breast carcinoma, which contains admixture of epithelial and non-epithelial components. Non-epithelial elements consist of spindle –shaped cells, bizarre giant cells, bone, cartilage or rarely skeletal muscle cells. Some classification systems also consider adenosquamous type and pure squamous cell variants under MBC based on the metaplastic cell component^{1,2}.

Pathological classification is challenging due to its histologic diversity, rarity of the disease and lack of significant consensus regarding appropriate system

of classification of the disease. MBC with prominent spindle cell component consists atypical spindle cells with fascicular arrangements. The amount of atypia varies from pleomorphic tumours resembling high-grade sarcomas to mild cellular atypia resembling benign fibrous lesion^{1,2}.

MBCs are usually high grade but low grade MBCs with fibromatosis-like phenotype have been recently described and must be considered in the differential diagnosis of low grade or benign looking fibrous lesions of breast. However, as in case of high-grade tumours, demonstration of epithelial component is necessary to label it as MBC³.

Cells of origin for MBC are thought to be epithelial in nature with sarcomatous components representing areas of metaplasia or de-differentiation. Tumours with predominant squamous cell differentiation represent metaplasia of malignant ductal epithelial cells⁴.

Most of the studies show that MBC is typically large at presentation with median size being 3.7 cm. In our case, size at presentation was 15cm, which is far larger than the average. MBC is usually associated with lower incidence of axillary lymph nodal involvement than would be expected from similar sized typical breast carcinoma, which is around 6-26%^{2,3}. In our case, palpable lymphnode was present in the axilla, though no tissue diagnosis is available to confirm the metastatic nature of the node.

MBC very less frequently expresses oestrogen and progesterone receptors (0-14%). The ER/PR receptor status in our case was negative. There is only limited data regarding HER 2/neu overexpression. A study showed HER2/neu over expression in 72% of cases with spindle cell and squamous cell variant where as 33% of cases with chondroid variant^{3,4}.

Patients with MBC have poorer outcomes compared to classic IDC of breast with high risk of recurrence following primary surgery. In various studies, disease free survival (DFS) after primary surgery after a follow-up of 20-25 months was 50-60%. Adenosquamous cell variant has little better prognosis compared to other variants^{1,5}.

Systemic therapy is rarely used for MBC and data regarding the same is limited because of the rarity of the disease. Different studies, where systemic therapy has been used for MBC, show that there is no significant response or increased survival benefit

with use of systemic chemotherapy or hormonal therapy in MBC^{1, 2, 5, 6}.

Conclusion

MBC is a recently classified rare subtype of breast carcinoma, which is aggressive compared to IDC.

It is characterised by advanced stage at presentation, relatively less involvement of axillary nodes, hormonal receptors negative tumour with increased recurrence rates and less median DFS.

Role of chemo-radiotherapy in MBC is limited since there is no significant survival benefit.

More studies including large number of patients with MBC are to be called for to study the disease entity in more detail and to explore novel prognostic and therapeutic targets for better survival in this disease.

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