

Case Report

A case of metaplastic breast carcinoma- an aggressive malignant tumour

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Background

Metaplastic breast carcinoma (MBC) is a rare, aggressive malignancy of the breast with poor prognosis compared to the common types. The presentation, diagnosis, prognosis and treatment strategies are different from other breast cancers. WHO recognized it as a distinct entity only in 2000. Data on presentation, diagnosis, prognosis and treatment strategies are limited. In addition, because of its benign appearance, it may be misdiagnosed at imaging.

Case

A 64 year old woman presented with a painless lump in her right breast of one month's duration. She had no predisposing factors for breast carcinoma. She did not have any symptoms suggestive of metastasis.

Examination revealed a 2x3cm, firm, irregular, ill-defined lump in the upper outer quadrant of the right breast with no skin or nipple changes. It was not attached to deeper structures. There was a 3x4cm mobile right axillary lymph node. Supraclavicular lymph nodes were not palpable. She did not have any signs of distant metastasis.

Ultrasonography of breasts revealed a 2.1x2.4cm irregular, hypoechoic lesion in the upper outer quadrant of the right breast and an axillary lymph node measuring 2.7x4.4cm. The left breast and axilla were normal. Mammography was not done as it was not available in our hospital.

Fine needle aspiration cytology revealed a benign lesion. Trucut biopsy was not feasible due to the cystic nature of the lesion. Excision biopsy of the lesion revealed a grade 2, pure epithelial squamous cell metaplastic breast carcinoma (MBC). Chest X-ray and ultrasonography of abdomen did not reveal any evidence of metastatic disease. She had T2N1Mx disease on clinical TNM staging with Stage IIB disease.

Subsequently she underwent total mastectomy with level II axillary clearance. Histopathology confirmed no residual disease in the breast and 2 out of 17 lymph nodes were positive for metastasis. The tumour stained negative for oestrogen, progesterone and Her-2-neu receptors. A staging CT scan and a bone scan failed to reveal distant metastasis. She was offered 8 cycles of adjuvant chemotherapy followed by radiotherapy to the chest wall and supraclavicular fossa. She is alive and free of disease at last follow up 2 years after diagnosis.

Discussion

Metaplastic breast carcinoma (MBC) is a rare aggressive malignancy of the breast characterized by the co-existence of carcinoma and non-epithelial cellular elements. The prognosis of MBC is less favourable compared to the more common types such as infiltrating ductal or lobular carcinoma. These tumours are usually triple negative.

MBC accounts for 0.25 – 1% of all breast cancers and has a high rate of local and distant metastasis and carries a poor prognosis [1]. MBC presents more commonly as a rapidly growing mass. It has been consistently reported to present with larger lesions (generally greater than 2 cm) than typical breast cancers. Fixation to the underlying deep tissues or to the skin has been reported in one study in over 20% of patients[1].

MBC has been described as having a high density on mammography with either circumscribed, obscured, irregular and/or speculated margins. Both mammography and ultrasonography could be deceptive as the lesions appear well circumscribed or microlobulated without prominent speculations [2]. However, in this case, ultrasonography was suggestive of malignancy. FNAC tends to be an acceptable diagnostic approach and Trucut biopsy is considered if cytology is inconclusive.

The World Health Organization (WHO) classifies MBC into low-grade adeno-squamous carcinoma, fibromatosis-like metaplastic carcinoma, squamous cell carcinoma, spindle cell carcinoma, metaplastic carcinoma with mesenchymal differentiation, mixed metaplastic carcinoma and myoepithelial carcinoma [3].

Metaplastic breast cancer presents with axillary nodal involvement less frequently than standard invasive breast cancer, despite the larger tumour size. Distant metastases can be found in the absence of lymph node metastasis, specifically affecting the brain and lungs. Patients with MBC are more likely to present with stage III and IV disease when compared to patients with other invasive breast cancers [3,8].

The optimal treatment strategies for MBC are still debatable due to low incidence and pathological variability [1]. Surgical treatment modalities have extended from mastectomy to breast conservation therapy. Adjuvant radiotherapy has shown long term survival benefit in these patients [1,4,5,6,7]. Post mastectomy radiotherapy has a limited role in patients with four or more metastatic axillary nodes, large primary tumours and chest wall invasion [4]. This patient was offered adjuvant radiotherapy as she was axillary node positive.

MBC is considered to be chemo resistant. Neoadjuvant chemotherapy is minimally effective at reducing tumour burden and preventing progression of disease. However, patients with MBC are treated with chemotherapy identical to that used in invasive ductal carcinoma (stage II & III) [1,4]. Hence, this patient was offered an anthracycline and taxane based regimen. Hormonal therapy is ineffective as these tumours are triple negative [1]. Although targeted therapy like trastuzumab is ineffective due to Her-2-neu negativity, some metaplastic squamous cell cancers may express HER -1 receptors and may be treated with drugs such as gefitinib (ZD1839) [1].

The five year survival rates ranging from 49% - 68% for patients with MBC is poorer than for those with invasive duct carcinoma (IDC) [1]. The five year disease free rate for MBC is around 40% and overall survival rate is 50%-65% with significantly poorer survival rate in stage III or higher, but with good prognosis for early stage disease[2]. An increased risk of local and distal recurrence has been reported in multiple studies [8]. Patients with triple negative MBC have poor 3-year disease-free survival compared to a similar group of triple-negative IDC patients receiving identical chemotherapy regimens [9]

Summary

Metaplastic Breast Cancer is a rare aggressive breast cancer, which presents in advanced stages associated with poorer prognosis. Both mammography and ultrasonography could be misleading as the lesions may appear benign. Distant metastases can be found in the absence of lymph node metastasis. These tumours are usually triple negative (negative for all 3 receptors) and hormonal therapy is ineffective. MBC is usually chemo resistant. Adjuvant radiotherapy has shown long term survival benefit.

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