

Multicentric Medullary Carcinoma Breast - Uncommon Histologic Subtype

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INTRODUCTION

Ridolfi et al. in 1977 defined medullary breast carcinoma (MBC) as one of the invasive and malignant subtypes of breast cancer.¹ It is a rare breast malignancy accounting for 5 % of breast carcinomas.² Although MBC is considered as having good prognosis, patients reported as triple-negative breast cancers are associated with aggressive clinical behaviour and poor prognosis.³ Also factors like local invasion, evidence of regional metastasis, etc. will determine the progress of this type of cancer.

PRESENTATION OF CASE

A female patient of 85 years came to Department of Oncosurgery with a complaint of painful lump in the right breast of 8-months duration, which was rapidly increasing in the last 3 months. The right breast lump was 9 x 8 cms., firm, mobile, tender and located in upper outer quadrant. There was retraction of nipple. The skin over mass showed ulcerations. Right axillary lymph nodes were palpable. There was no family history of breast cancer. She was not on any medication or hormonal therapy. On general physical and systemic examination no other significant findings were noted. Patient had obstetric history of G3P3. Routine and haematological investigation were normal. HER2, CA - 125 was 15.9 μ / mL, serum CEA was 5.9 ng / mL. On mammography, showed a mass in right breast, having BIRADS V, with perimass inflammation and oedema. It was reported as suggestive of malignancy. Mass was adherent to the pectoral muscle. Right axillary lymph nodes were enlarged suggestive of metastasis. On fine needle aspiration cytology reported as suggestive of breast carcinoma. She underwent modified radical mastectomy with right axillary clearance.

Medullary carcinoma of the breast presenting as multicentric tumour is rare. The medullary carcinoma has high-grade cytological features. However, it has prognostically having relatively good outcome. An 85-year-old female presented with lump in right breast of 8 month which rapidly increased in last 3 months. The lump was 9 x 8 X 6 cm firm, mobile, tender and located in upper outer quadrant. Right axillary lymph nodes were palpable. Her CA - 15.3 was 15.9 μ / mL, serum CEA was 5.9 ng / mL. On mammography showed a mass in right breast having BIRADS-V, with perimass inflammation and oedema. It was suggestive of malignancy. Right axillary lymph nodes were enlarged suggestive of metastasis. On fine needle aspiration cytology reported as suggestive of breast carcinoma. She underwent modified radical mastectomy with right axillary clearance. On histopathological findings reported as multicentric medullary carcinoma breast-invasive breast carcinoma specific type of right breast. Conclusion - We are reporting uncommon case of multicentric medullary carcinoma breast for its clinical, radiological, histopathological and immunohistochemical findings.

CLINICAL DIAGNOSIS

Breast Cancer with Axillary Lymphadenopathy - Right Breast

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DIFFERENTIAL DIAGNOSIS

- Inflammatory carcinoma.
- Lymphoma
- EBV (Epstein-Barr Virus) Associated Carcinoma
- Undifferentiated Ductal Carcinoma

PATHOLOGICAL DISCUSSION

Gross

We received the specimen of modified radical mastectomy. On gross examination, right breast specimen was measuring 20.5 x 12.5 x 6.8 cm and weighing 558 gms. The covering skin flap measured 15.2 x 12.3 cm and showed irregular ulcer (Figure 1) with large slough and oedema measuring 8.5 cm X 6.2 cm. Nipple was retracted. Serial cut sectioning revealed a tumour measuring 8.5 x 8 x 6.5 cms, located in the upper, inner and upper outer quadrant of right breast. Tumour is soft to firm in consistency. Tumour had pushing borders and cut section was grey white to grey-yellow. Deep surgical margin was 1 mm away from tumour. Overlying skin was 1 mm away from tumour. Nearest peripheral surgical margin was 0.5 cm away from tumour. Adjacent breast tissue was unremarkable. Also noted two tumour nodules located in the upper outer quadrant at a distance of 1.6 cm and 2.3 cm from the main tumour respectively. One tumour nodule measured 4.4 x 3.6 x 2.5 cm and cut section of which was soft to firm, grey white to grey yellow and shows focal areas of haemorrhage. Other tumour nodule measured 2.2 x 1.5 x 1.3 cm and cut section of which was soft to firm, grey white to grey yellow and showed focal area of haemorrhage (Figure 2). The right axillary clearance showed fibro-fatty grey yellow piece of tissue measuring 7 x 5.5 x 1.8 cm. Serial cut sectioning revealed 9 lymph nodes. Largest lymph node measured 1.2 x 1 x 0.8 cm. Cut section of which was grey white and firm.



Figure 1.
Gross Specimen of Breast Showing Tumour with Irregular Skin Ulcer



Figure 2.
Breast Specimen - Cut Section Showing Multiple Tumours

Microscopic Examination

Histopathological findings reported as multicentric medullary carcinoma breast of right breast. RB (Bloom Richardson) score was $3 + 3 + 2 = 8$, Grade III. Moderate to marked lymphoplasmacytic infiltrate within and at periphery of tumour was noted. (Figure 3, 4). Tumour showed extensive necrosis and scattered bizarre tumour giant cells. Right axillary clearance revealed 9 lymph nodes in which 3 were involved by tumour.

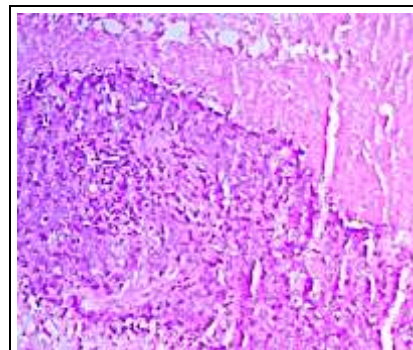


Figure 3.
Tumour Syncytial Growth Pattern, Marked Diffuse Lymphoplasmacytic Infiltration (H & E Stain, 100X)

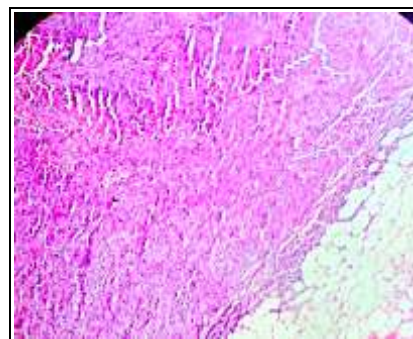


Figure 4.
Tumour Syncytial Growth Pattern, Necrosis and Marked Diffuse Lymphoplasmacytic Infiltration (H & E Stain, 40X)

DISCUSSION AND MANAGEMENT

Modified radical mastectomy is the surgical treatment. Adjuvant chemotherapy and radiation are recommended similar to other triple negative tumours and invasive carcinoma respectively. Pure medullary carcinoma has a better prognosis; therefore, strict specific diagnostic criteria are critical to provide better healthcare.

MBC is uncommon and represents about 5 % of all breast cancers. MCB is of two types that is typical medullary breast carcinoma (TMBC), and atypical medullary breast carcinoma (AMBC). The criteria used to diagnose TMBC are as 1) syncytial growth pattern- > 75 % of tumour, 2) microscopically circumscribed tumour, 3) moderate to marked diffuse mononuclear stromal infiltrate, 4) micro glandular and intraductal components are not seen, and 5) moderate or marked nuclear pleomorphism is noted.¹

Medullary carcinoma breast can develop at any age, but more commonly affects in young women in their early 50s.⁴ It is more common in Japan than in North America. Patients with mutations of the suppressor gene BRCA - 1 are having more risk to develop TMBC.⁵ Medullary breast cancer tends to occur ipsilateral breast, but approximately 3 - 18 % of patients may develop multicentric tumours. In our case it was in ipsilateral breast with three lesions.

A study by Chu Z et al found in total of 117 diagnosed with TMBC, one patient presented with 2 lesions in different quadrants of ipsilateral breast.⁶ MBC, on gross examination are usually single, large distinct mass with a circumscribed margin and soft to firm consistency. There may be haemorrhages and cystic changes. In our case it showed multicentric lesion with three lesions. Most of the tumours are of 2 - 3 cm. The tumours are usually soft to firm and discrete. Tumour shows lobulated or nodular appearance. Also, areas of necrosis and haemorrhages are noted. Young *et al.* have reported cases of bilateral medullary carcinomas in about 3 - 18 % of patients. He observed multicentric medullary carcinoma in one breast.⁷ In our case patient presented in late age. Rarely in 1.5 % of cases are found in the 75 – 79 years age group.⁸ Along with typical five criteria of MBC other common findings are spindle cell metaplasia, tumour giant cells, cystic degeneration, and absence of calcification. Many cases show extensive tumour necrosis. In our case showed extensive necrosis and scattered bizarre tumour giant cells. Immunohistochemical profile of typical MBC is usually characterized by features of basal like carcinomas. These tumours triple negative i.e. for ER, PR and HER 2 / neu receptors. The triple negative for medullary carcinoma were reported 70 %.⁹ The lymph node metastasis in medullary carcinoma to axillary node is common but is limited to few nodes only. The TMBC have an axillary lymph node metastasis in about 26.5 % cases. Most published studies have reported incidence of axillary lymph node involvement in patients with medullary carcinoma (19 % - 46 %).¹⁰

FINAL DIAGNOSIS

Multicentric medullary carcinoma breast of right breast. RB score was 3 + 3 + 2 = 8, Grade III.

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Disclosure forms provided by the authors are available with the full text of this article at jebmh.com.

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