Abstract: Medullary carcinoma of breast are rare tumors of breast, they are associated with BRCA1 mutation. They may be high grade histologically but are less aggressive and prognosis is usually good. We present a rare case of medullary carcinoma of right breast.

Keyword: Carcinoma breast, medullary carcinoma breast

Case report
Forty three years old female presented with complaints of lump right breast for two months and pain over lump for one month. On examination mass over right upper quadrant of breast 3x4 cm, firm in consistency, freely mobile not fixed to underlying structure, surface smooth, skin over swelling normal, nipple areola complex normal, no axillary lymphadenopathy. Mammographic study showed a circumscribed mass lesion with speckled calcification BIRADS III and Fine needle aspiration cytology from mass reported as ?fibroadenosis with atypical cells. Patient had no other comorbid conditions, blood investigations, x-ray chest & skeletal survey and ultrasound abdomen were normal. As investigative results were inconclusive, we did excisional biopsy of the mass and sent for histopathological examination which reported as medullary carcinoma of breast which was ER,PR & Her-2 neu negative. As medullary carcinoma is a type of invasive carcinoma of breast after completing the metastatic workup, which was normal, we re-operated and modified radical mastectomy was done. Further, adjuvant chemotherapy with doxorubicin based FAC regimen (5 fluorouracil, Adriamycin, cyclophosphamide) was given.

Discussion
Medullary carcinoma is highly characterized by the presence of lymphocytes, which arise as an immune system response to the cancer. These lymphocytes, which are essentially white blood cells, tend to congregate around the periphery of the tumor. Whether or not lymphocytes are beneficial in combating breast cancer is open to debate, as sometimes lymphocytes can harm normal body tissues. In case of medullary carcinoma, however, the lymphocytes are thought to be helpful. Medullary carcinoma is also found to have certain genetic and hormonal characteristics, including estrogen receptor (ER) negative and progesterone receptor (PR) negative status, and human epidermal growth factor receptor2 (HER-2) negative cells.

Clinical features & investigation
Medullary carcinoma commonly manifests as well-circumscribed mass And cannot be differentiated from fibroadenoma.
by physical examination, on ultrasound larger masses which are round or lobular in shape, with a focally thick wall and anechoic cystic space, and enlarged axillary lymph nodes are more common findings in patients with medullary carcinoma than in patients with fibroadenoma. These sonographic findings help to differentiate medullary carcinoma from fibroadenoma. Mammographically the lesion may be well circumscribed, indistinct or blended, with or without calcification circumscribed pattern was the most frequent type. Breast FNAC samples with syncytial fragments, bizarre nuclei with prominent nucleoli, and a chronic inflammatory infiltrate should raise the possibility of medullary carcinoma. However, the differential diagnosis also includes high-grade ductal carcinoma, lymphoma, or metastasis to breast or intramammary lymph nodes; thus, histopathologic analysis is required for definitive diagnosis.8

**Histology:**
The ‘classic’ histological appearance of medullary carcinoma is a syncytial growth pattern, poorly-differentiated structures, with a high mitotic (cell division) rate1. Medullary carcinoma cells tend to be relatively large, and occur in large ‘sheets’ of cells, making it difficult to distinguish the individual cell membranes separating them. Cell structures tend to be ‘syncytial’ (containing multiple nuclei), poorly-differentiated, and the nuclei tend to bepleomorphic, and with prominent nucleoli. Another distinguishing feature of medullary breast cancer is a clear boundary or margin between the tumor and the surrounding breast tissue. The cancer cells push against the surrounding skin tissues but have less of a tendency to ‘infiltrate’ than other carcinomas. Lymphocytes tend to collect along this margin.9

**Ridolfi criteria**
Ridolfi criteria is useful to diagnose and to grade medullary carcinoma these include: lymphoplasmacytic reaction microscopic circumscription; syncytial growth pattern high nuclear grademitotic rate. A cell that has enough functioning normal DNA to form a specific type of tissue, and behave like that tissue, is “differentiated”. A cell that has so many mutations, that it forms hideously distorted tissues, is poorly-differentiated. The term ‘poorly-differentiated’ from a genetic perspective suggests that a cell started out intending to form a specific type of tissue, and then, according to histological risk grading procedures, it tends to receive a medium to high grade. High grade carcinomas are considered more likely to metastasise and carry a poorer outlook, but medullary carcinoma is actually a fairly mild breast cancer in terms of prognosis, in spite of the histological profile. Since lymphnode metastasis is less likely for medullary cancer when compared with other types of infiltrating ductal carcinoma, it provides more options for treatment. In most cases, chemotherapy is indicated as the most favourable treatment option.5

**Prognosis and treatment**
A curious feature of medullary carcinoma is that, according to histological risk grading procedures, it tends to receive a medium to high grade. High grade carcinomas are considered more likely to metastasise and carry a poorer outlook, but medullary carcinoma is actually a fairly mild breast cancer in terms of prognosis, in spite of the histological profile. Since lymphnode metastasis is less likely for medullary cancer when compared with other types of infiltrating ductal carcinoma, it provides more options for treatment. In most cases, chemotherapy is indicated as the most favourable treatment option.5 Medullary Carcinoma is generally believed to have a slower-growing, less aggressive biological characteristics. Basal-like tumors (which tend to be ‘triple negative’) tend to have a poorer prognosis6. Survival rates for true medullary carcinoma tend to be higher than for ‘atypical’ medullary carcinoma, which are in turn still higher than for non-specified ductal carcinoma. Overall, the prognosis for medullary carcinoma is generally very good, with a 78% five year survival rate. For stage I women the 20 year survival rate is around 95%, and about 62% for stage II cancers.

**Conclusion**
Medullary carcinoma breast usually present as a mass which seems to be benign clinically, high degree of suspicion is needed to diagnose these cancer either by FNAC or by histopathological examination. Though histologically these tumors appear high grade their prognosis is usually good after surgery followed by adjuvant chemotherapy.5 Medullary Carcinoma is generally believed to have a slower-growing, less aggressive biological characteristics. Basal-like tumors (which tend to be ‘triple negative’) tend to have a poorer prognosis. Survival rates for true medullary carcinoma tend to be higher than for ‘atypical’ medullary carcinoma, which are in turn still higher than for non-specified ductal carcinoma. Overall, the prognosis for medullary carcinoma is generally very good, with a 78% five year survival rate. For stage I women the 20 year survival rate is around 95%, and about 62% for stage II cancers.

**References**

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