Scholars Journal of Applied Medical Sciences (SJAMS) **ISSN 2320-6691 (Online)**
Sch. J. App. Med. Sci., 2013; 1(6):681-683 **ISSN 2347-954X (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjams.2013.v01i06.006

Case Report

Medullary Carcinoma Breast: A Case Report

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Abstract: Invasive ductal carcinoma is the most common form of breast cancer, attributing to about 70 to 80 percent of all breast cancers; the medullary variety constitutes 1 to 10 percent of this carcinoma. A case review of the medullary carcinoma is presented with regards to clinical and pathological characteristics.
Keywords: medullary carcinoma, incidence, modified radical mastectomy, prognosis

INTRODUCTION

In 1949, Moore and Foote described their experience with a well-circumscribed form of breast carcinoma with lymphoid stroma [1]. It was called medullary carcinoma and was recognised to have a favourable prognosis compared with other histological subtypes. Medullary breast carcinoma accounts for less than 5% of all invasive breast cancers and is a frequent phenotype of *BRCA1* hereditary breast cancer [2]. Women with this cancer have a better 5-year survival rate than those with NST or invasive lobular carcinoma.

CASE REPORT

A 48 year old female presented with complaints of right breast lump and fungating growth over the right breast since 6 months. Growth was associated with foul smelling bloody discharge. She gave history of similar lump in the right breast 2 years back for which she underwent lumpectomy, but did not receive any chemo-radiation. On examination an ulcero-proliferative mass was present in the upper and outer quadrant of the right breast measuring 7x7cms (Fig 1, 2). Mass was not fixed to the chest wall. An anterior axillary lymph node was palpable measuring 2x2cms in size. Patient was staged IIIa [$T_3N_1M_0$]. All routine investigations were within normal limits. Mammogram of the opposite breast was normal. Patient underwent modified radical mastectomy with axillary clearance. Specimens were sent for histopathological examination and hormonal status. Histopathological analysis of the mass was suggestive of medullary carcinoma and showed well circumscribed and capsulated lesion showing pleomorphic cells arranged in sheets and cords (Fig 3). Cells were round – oval with hyperchromasia and O-1 nucleoli and scant cytoplasm. Sheets of cells exhibited pushing borders without evidence of any infiltration. There was no evidence of vascular or capsular invasion. All the margins and base of the specimen were free of dysplastic or malignant cell infiltration. Lymph node sent was not involved. On immuno-histochemistry tumor was triple negative. Post-operative period was uneventful and patient was referred to higher centre for chemo-radiation.

Fig. 1: Gross specimen showing the growth

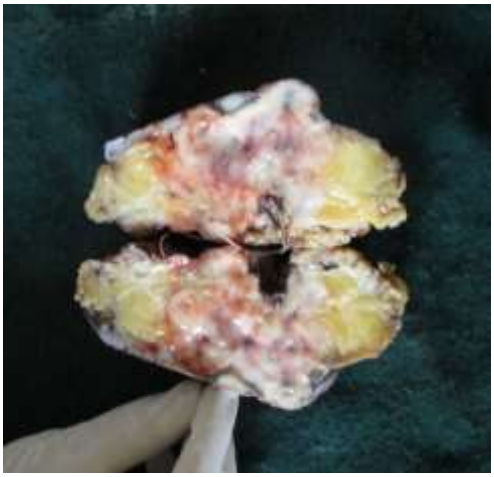

Fig 2: Gross specimen (cut-section) showing the tumor

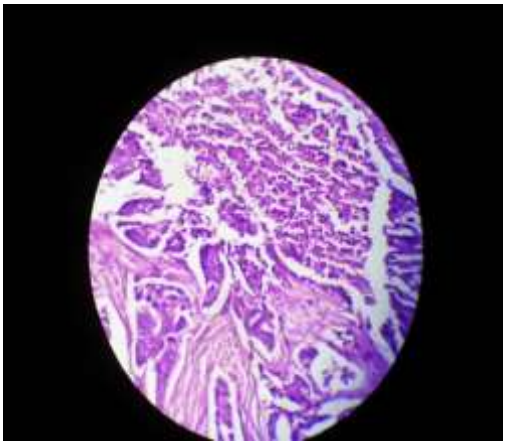

Fig 3: Histopathology slide showing pleomorhic cells with lymphocytic infiltrate

DISCUSSION

According to WHO over 1.2 million women are diagnosed with breast cancer annually worldwide. Infiltrating ductal carcinoma is a broad entity which comprises of tumors that exhibit one or more characteristics of specific types of breast cancers. There is evidence of tubular, papillary, medullary or mucinous differentiation microscopically [3]. Medullary carcinomas account for fewer than 5% of all invasive breast cancers [4].

Patients with medullary carcinoma usually present at a relatively younger age than patients with other breast cancers; the mean age at presentation is in the late fifth and early sixth decades, with a wide age range reported.The majority of patients with medullary carcinoma present with a palpable mass, usually in the upper outer quadrant. Of interest, some patients with this tumor type exhibit axillary lymphadenopathy at the time of presentation, suggesting the presence of metastatic disease. Histologic examination of the lymph nodes in such cases, however, typically reveals benign reactive changes. Rare examples of medullary carcinoma have been reported in men [15].

Though this is essentially a histopathologic diagnosis, certain features on the imaging modalities are described which underline the malignant nature of the lesion. On mammogram, these features include irregular shape, spiculated or irregular margins, or high density of the lesion. The density of the lesion can be described as low, intermediate or high by comparing it with an area of normal breast tissue on mammogram. Generally, benign masses tend to be of lower density than carcinomas; however, it is not reliable as a distinguishing mammographic sign. Sonographic findings suspicious for cancer include spiculation or thick echogenic halo, microlobulations, hypoechogenecity, calcification, acoustic shadowing, duct extension [16]. In gross appearance, these lesions are well-circumscribed, soft, tan-brown to grey tumors that bulge above the cut surface of the specimen. A multinodular appearance may be appreciated in some cases. Areas of hemorrhage, necrosis, or cystic degeneration may be present in tumors of any size, but prominent necrosis is usually seen in larger tumors [15].

Three similar but distinct classification systems for the histologic diagnosis of medullary carcinomas have been proposed by Ridolfi *et al.* [3], Wargotz and Silverberg [5] and Pedersen *et al.* [14]. All three classification schemes recognize the following attributes of medullary carcinomas, but the relative importance and the mandatory nature of each are stressed to different degrees: (a) syncytial growth pattern of the tumor cells in more than 75% of the tumor, (b) admixed lymphoplasmacytic infiltrate, (c) microscopic circumscription, (d) grade 2 or grade 3 nuclei, and (e) absence of glandular differentiation Tumors that lack a variable number of these characteristics (depending on the system used) are classified either as atypical medullary carcinoma or invasive ductal carcinoma. Structural variations that characterize atypical medullary carcinoma include invasive growth at periphery of tumor, well-differentiated nuclear cytology, diminished lymphoplasmacytic reaction, low frequency of mitoses and glandular, trabecular or papillary growth with fibrosis[6].

There are recent publications of classification of breast cancer on the basis of gene expression profile analysis which have proposed that medullary carcinoma can be part of basal-like carcinoma spectrum made up of ER-negative, PR-negative and HER2-negative phenotype [7]. However, there are number of reports in literature that it can be ER, PR and/or HER2 positive, indicating the heterogeneity of this type of breast carcinoma [7-10].

Most studies indicate that the incidence of axillary lymph node metastases is lower in patients with medullary carcinomas (19% to 46%) than in those with atypical medullary carcinomas (30% to 52%) or invasive ductal carcinomas (29% to 65%).

The prognosis of medullary carcinoma, whether typical or atypical, is better than that of high grade infiltrating ductal carcinoma [11]. It has been proposed that extensive presence of plasma cells and lymphocytes helps to keep the medullary carcinoma in check, preventing it from growing and spreading quickly [12]. The overall 10-year survival rate is 74% and more than 90% in patients with negative lymph nodes [13]. It is interesting to note that pure medullary carcinoma has better prognosis as compared to the one with atypical features. Hence, strict adherence to diagnostic criteria is critical to provide a more accurate prognosis.

The treatment for medullary carcinoma, whether typical or atypical, is similar to invasive ductal carcinoma [6]. The division into typical and atypical

subtypes has prognostic significance and does not modify treatment options. The treatment includes modified or radical mastectomy along with radiation or chemotherapy depending on the stage or histologic grade.

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