Metastatic Metaplastic Breast Carcinoma

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A 33 years old Saudi single female from a suburban village presented with a rapid progressive increase of the left breast over a period of seven months. She was alarmed one month prior to presentation with copious spontaneous bloody discharge associated with progressive heaviness of the left coupled with intractable neck and shoulder pain. The patient claimed that she had regular menstrual cycles with no prior hormonal, radiation or surgical treatment. Past history and family history were negative.

Clinical Examination

Revealed a young frail female with normal Both vital signs and systemic examination. Local examination revealed asymmetrical breasts with normal examination on the right breast. There was massive left breast enlargement, red stretched skin, nipple retraction and copious serosanguious nipple discharge. The surface of the breast contained painless multiple masses with variable consistencies. There were no palpable axillary lymph nodes. (Fig. I a,b)

Investigations

Mammogram of the right breast was within normal limits, however, mammogram of the affected side could not be performed due to hard consistency of the enlarged breast. Ultrasound has shown the breast in involved with a large mass out of range for measurement with both solid and cystic components. Vascularity of the lesion is limited. (Fig. II). Chest x-ray revealed scattered multiple bilateral lung fields rounded lesions suggesting disseminated metastasis. (Fig. II) CT. Scan of the chest confirmed the presence of the enlarged breast lesion measuring 17.2x12.6cm with heterogeneous density and variable areas of necrosis and confirmed the chest x-ray findings of multiple hypo-attenuating pulmonary nodules seen scattered throughout both lung fields. (Fig. IV) Ultrasound guided biopsy reported revealed atypical cells. The remainder of the metastatic workup was negative.

Figure I (a,b): massive enlargement of the left breast with multiple irregular masses

Figure II: Ultrasound shows an irregular large mass of both cystic and solid components with minimal internal vascularity

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Management

After detailed discussion with the multidisciplinary team palliative mastectomy was the treatment of choice for symptom relief. The mastectomy specimen weight was 2.7 kilogram with multiple cystic spaces, hemorrhage and extensive necrosis. (Fig. IV) Histo-pathological evaluation of the specimen reported exclusive spindle cell differentiation without epithelial differentiation. The neoplastic cells were pleomorphic, with indistinct cell borders, inconspicuous nucleoli, accompanied with scattered osteoclast-like giant cells, frequent mitotic figures and extensive necrosis. (Fig. VI a&c The rim of residual breast parenchyma was unremarkable. Immunohistochemical studies showed focal positive reactivity for cytokeratin AE1/AE3 (Fig. VI c)), CK 5/6 and CK8/18. However, they were negative for epithelial membrane antigen, CK7, BCL-2, Beta-catenin, estrogen receptor, progesterone receptor and HER2.

Based on these findings the patient was diagnosed with Metaplastic Breast carcinoma.

Figure III: Chest x-ray revealed multiple bilateral lung lesions suggestive of disseminated lung metastasis

Figure IV: CT-Scan of the chest demonstrating enlarged left breast mass with multiple lung metastases

Figure V: The cut surface showing area hemorrhage and necrosis

Figure VI (a,b): Low and high power microscopic examination shows the exclusive spindle cells
Discussion

Despite the emerging sporadic case reports over the last decades, Metaplastic breast carcinomas (MBC) remain rare primary breast malignancies. The co-existence of carcinoma with non-epithelial cellular elements certainly characterizes this entity. They can be classified as monophasic spindle cell carcinoma, biphasic carcino-sarcoma, adenocarcinoma and squamous cell carcinomas [1].

It affects women in their forties yet younger women with more aggressive disease have been reported. The active young breast is inviting for the more aggressive sub-types of breast cancer with dismal prognosis [2].

Metaplastic carcinoma of the breast is a rare accounting for only 0.25% of all breast cancers [3]. They are characterized by its aggressive nature and progresses rapidly into enormous size with less likely nodal involvement but inclination to distal metastases. [4, 5] Reported variation is only in age range yet the overall tumor characteristics are similar [6]. Reported sizes range between 3.4–>13 cm with the histological features dictating the diagnosis. [7-9]

Despite the active nature of MBC its poor prognosis is mainly determined by the extent type, and the presence extensive sarcomatous component [10].

The best treatment option of MBC to avoid local recurrence is wide and adequate surgical excision sparing the axillae. Adjuvant chemotherapy may cause a paradoxical increase in size and role of radiotherapy results is not promising. [1, 11]

We are skeptical regarding reports encouraging the adoption of breast conserving surgery in MBC since the role of adjuvant postoperative external beam is radiotherapy is limited [12, 13].

Metastatic disease expected to develop at a mean of 10.5 and 14.5 months from the initial diagnosis and demise may follow within 20 months of the onset of metastasis. [1] Like invasive carcinoma common sites for metastasis include lung, bone, and liver, with the lung metastasis in 31-70% of patients. [8] It is the agreed upon that the treatment options are limited. It currently remains that the only successful treatment is simple Mastectomy to achieve negative margins sparing axillary node dissection [14-16].

We conclude that MBC remains a rare entity with poor response to both chemo-radiotherapy, histological diagnosis is challenging and surgery is the main option currently available.

References


