



PRIMARY SMALL CELL CARCINOMA OF BREAST, RARE BREAST CANCER: A CASE REPORT.

Oncology

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ABSTRACT

Primary small cell carcinoma of breast is a very rare variety of breast cancer. There is no defined standard treatment of primary small cell carcinoma of breast in the literature due to rarity of the disease. It constitutes <1% of all breast cancers. Optimal management strategy may include surgery (non- metastatic cases) followed by systemic chemotherapy commonly cisplatin and etoposide based and chest wall irradiation with 50 Gy in 25 fractions, there is no clear guidelines regarding the dose of radiation, therefore it is justifiable to use 50 Gy in 25 fractions. We are reporting a case of right sided primary breast small cell carcinoma in a 34 years old premenopausal female, treated with right modified radical mastectomy followed by etoposide and cisplatin based chemotherapy 4 cycles and chest wall irradiation 50 Gy in 25 fractions. She completed her treatment and regularly on follow up.

KEYWORDS

rare breast cancer, small cell breast cancer, radiation of small cell breast cancer.

INTRODUCTION

Primary small cell carcinoma of breast is a very rare variety of breast cancer. WHO in 2003 defined small cell cancer of breast as a separate subtypes. The morphological & immunohistochemical (IHC) features of these tumors are similar to that of small cell neuroendocrine cancer of the lung, so a thorough search must be performed to rule out any metastatic disease from another primary site. Extrapulmonary small cell cancer comprises of only 5% of all small cell carcinoma [1]. Small cell carcinoma of the breast constitutes <1% of all cases of breast cancers [2]. There is no defined standard treatment of primary small cell carcinoma of breast in the literature due to rarity of the disease. The population based study by Hare et al., included 199 patients with small cell carcinoma of the breast, who were compared with patients with small cell lung carcinoma [3]. Treatment strategies have been described those were based on treatment of breast cancer & small cell cancer of lung [4]. We are going to describe a case of 34 yrs old female presented to General surgery OPD at AIIMS, Patna with right sided breast lump for last 8 months, she underwent right sided MRM followed by systemic adjuvant chemotherapy and planned for adjuvant radiotherapy.

CASE PRESENTATION

34 yrs old female presented to general surgery department of AIIMS, Patna in august 2018 for evaluation of right sided breast lump for last 8 months which was progressive in nature. She was premenstrual and had two living issues, both of the children whom she had breastfed for more than 6 months. She had no medical comorbidities and also there is no history of major medical or surgical ailments in the recent past. There is no family history of malignancy. Clinical examination revealed that there was a mobile lump in the right breast involving upper outer quadrant of about 12 x 8 cm not fixed to chest wall & skin was also not involved. There was a single, mobile, non tender palpable lymph node in medial wall of axilla of about 2 x 1 cm. Left breast, axilla & supraclavicular fossa was within normal limit.

USG breast confirms right sided BIRADS 4a & left sided BIRADS 1. USG guided FNAC from right breast lump revealed possibility of neuroendocrine tumor. Trucut biopsy from the right breast lump showed lobular carcinoma with neuroendocrine differentiation (fig: 1). IHC from trucut biopsy specimen revealed that tumors cells are CK (fig: 2), Chromogranin A (fig: 3), EMA (fig: 4), E-cadherin positive (fig: 5) & negative for Synaptophysin, CD56, CD45, ER, PR, GATA-3, TTF-1. Ki-67 was 80%. IHC impression was consistent with small cell neuroendocrine carcinoma.

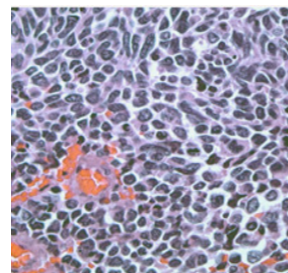


Fig: 1 H&E staining showing neoplastic cells with nuclear pleomorphism, mitotic features, hyperchromatic nuclei & scanty of cytoplasm

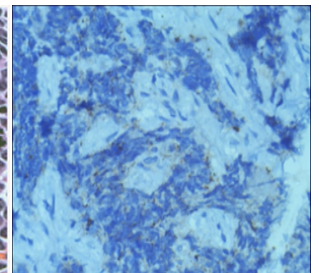


Fig: 2. IHC 400X showing positive staining for CK consistent with small cell carcinoma

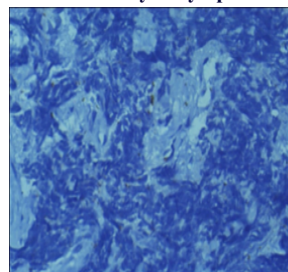


Fig: 3. IHC at 400X showing positive staining for chromogranin

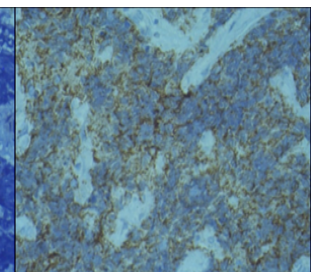


Fig: 4. IHC at 400X showing positive staining for E cadherin

Metastatic workup did not revealed any systemic metastatic disease with normal CT scan thorax, normal CT scan whole abdomen & normal TC^{99m} bone scan study. Patient underwent right sided MRM with axillary lymph node dissection on 27/9/18. Post operative recovery was uneventful. Post operative histopathological examination report revealed neuroendocrine carcinoma breast right side, grade 2, all margins negative, lymphovascular invasion and perineural invasion was not seen, all lymph nodes are reactive out of 19 dissected nodes. Serum chromogranin A was 149ng/ml (post operative). Patient was started on systemic chemotherapy cisplatin & etoposide based regimen. She received 4 cycles of systemic chemotherapy completed in March 2019 with manageable systemic chemotherapy related toxicity. She also received right sided chest wall, right axillary and right supraclavicular irradiation of 50 Gy in 25 fractions, 2 Gy per fractions over 5 weeks. Her treatment was completed in May 2019. She is on regular follow up with no evidence of local or systemic recurrence till December 2019.

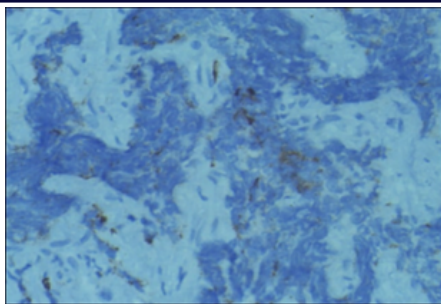


Fig: 5. IHC at 400X showing positive staining for EMA

DISCUSSION

In current scenario breast cases are diagnosed early due to increased awareness among common people. Small cell cancer of breast is one of the most aggressive subtypes of cancer and commonly found in lungs, but may arise at any other sites with high potential of metastases at diagnosis. It is very important to rule out any other primary site that may have lead to the metastases to the breast. Primary small cell carcinoma of breast generally presents as palpable breast lump mainly in females with median age of 60 yrs, at diagnosis 50-60% of patients may have involved lymph nodes [5]. As due to rarity of the disease and limited availability of data there is no defined guideline for the management approach. The treatment of our patient consisted of combination of approach to breast cancer management and management principles of small cell carcinoma of lung due to similarity in histological and clinical behaviour. The standard management of small cell carcinoma of lung consists of 4 cycles of etoposide and cisplatin based chemotherapy followed by thoracic irradiation and prophylactic cranial irradiation to patients who responds to chemotherapy and radiotherapy [6]. The question remains unanswered that whether the same principle for management of small cell carcinoma of lung can be applied in the management of extra pulmonary small cell carcinoma. The answer may be drawn from the study of Peter MacCallum Cancer Centre in East Melbourne, Australia 2011. This study concluded that the overall 5-year survival rate was 25.4% for patients with limited disease and 0% for patients with extensive disease. Improved overall survival was seen in patient use of definitive radiation therapy along with sequential chemotherapy [7]. There was low risk of brain metastasis in cases of extra pulmonary small cell carcinoma, therefore prophylactic cranial irradiation is not recommended [8]. Our patient was managed with right sided modified radical mastectomy followed by adjuvant chemotherapy with etoposide and cisplatin based regimen and she was planned for 4 cycles of chemotherapy followed by radiotherapy, due to negative ER/PR receptor status hormonal therapy was not considered. During the preparation of this case report she has completed her treatment and she is on regular follow up.

CONCLUSION

Primary small cell carcinoma of breast is very rare and aggressive subtype of breast cancer. It shows similar hisopathological and IHC features to that of small cell lung cancer, therefore diagnostic workup must include the investigations to rule out other primary sites. As the small cell cancer of breast is rare entity there is no standard guideline for its management. Management strategy may include treatment of breast cancer and treatment of small cell cancer of lung. The role of prophylactic cranial irradiation in small cell breast cancer is not defined due to lack of evidence as compared to small cell lung cancer. Further studies are required to formulate the standard treatment for this rare subtype of breast cancer.

Conflict of interest

The authors have no conflict of interest to declare.

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