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Case Report

Primary mucinous cystadenocarcinoma of breast with sternal metastasis: A diagnostic challange

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ABSTRACT

The WHO categorizes the breast tumors in various distinct histological types based on cell morphology and architecture. A widely accepted fact that the aggressive nature of the breast cancer can be determined by its histological type, grade, nodal status, and metastasis holds the ground even in the era of immunohistochemistry (IHC) and molecular. An extremely rare variant of primary breast carcinoma belongs to the family of mucin producing carcinoma sharing the same histology as the mucinous cystadenocarcinoma of the ovary and pancreas. The tumour typically occurs in postmenopausal women with a median patient age of 61 years. Due to the rarity of these tumors, there is an extreme paucity of literature on this topic. We report a case of 35 year old female with left breast lump and presternal swelling. Breast lumpectomy and total sternectomy was done and final diagnosis was given as Primary mucinous cystadenocarcinoma with sternal metastasis and Estrogen receptor (ER) and Progesterone receptors (PR) positivity.

1. Introduction

Mucinous cystadenocarcinoma is an uncommon histological subtype of breast cancer composed of cystic structures with tall columnar cells with abundant intracytoplasmic mucin, resembling pancreaticobilliary, ovarian and appendix tumours.[1] The tumor typically occurs in postmenopausal women with a median patient age of 61 years. The incidence of breast mucinous cystadenocarcinoma is about 1-6% of primary breast cancers.[2,3] On immunohistochemistry, ER and PR receptors and HER2 neu are usually negative.[4,5] Due to the rarity of these tumors, there is an extreme paucity of literature on this topic. We report a case of 35 year old female with left breast lump and parasternal swelling. Breast lumpectomy and total sternectomy was done and final diagnosis was given as Primary mucinous cystadenocarcinoma with sternal metastasis and ER & PR positivity.

2. Case History

A 35-year-old female referred to the Zydus Cancer Centre for lump in the breast and sternal lesion. Peviously operated for left breast lump with axillary dissection at outside Centre. At that time the patient was diagnosed as Papillary DCIS with abundant mucin lakes in adjacent area. After that presternal swelling small biopsy was done. The diagnosis given as Adenocarcinoma with glandular and papillary architecture and low to intermediate nuclear grade. At that time IHC showed ER and 34BetaE12 positivity and suggested probably breast origin. The patient was then finally referred to Zydus Cancer Centre.

On sonography, There is heterogenous and well circumscribed mass in left breast at 3O'clock position, mass

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measuring 21x11 mm. Two rounded to oval circumscribed solid cystic masses without significant vascularity seen in 4o'clock position, measuring 15x9 mm and at 9o'clock position measuring 16x13 mm.

On MRI, Large confluent expansile lytic lesion involving almost entire sternum from the level of manubrium upto lower end of body. Two closely placed soft tissue nodular lesions were seen in central -outer quadrant of left breast.

2.1. Histopathological examination

The patient was admitted and underwent lumpectomy surgery with total sternectomy. Intraoperative frozen section of breast lump received (Figure 1, gross image). The lump showed multiple cystic lesions at 4 to 5 o' clock and 3o'clock position. The surgical margins were free from the lesions. The frozen section diagnosis was given as-Neoplastic lesion favours mucinous carcinoma. Paraffin sections were awaited for final diagnosis.

Fig. 1: The lump received in frozen section. Cut surface of sternum shows tan brown solid and cystic areas

On gross examination, multiple sections were taken and stained by hematoxylin and eosin. Microscopically, neoplastic tissue was composed of dilated ducts, filled with papillary projections, lined by mild to moderate atypical tall columnar cells with scattered mitoses and without distinct, myoepithelial layer mostly floating in mucinous lakes with invasion in surrounding stroma (Figure 2, microscopy). Focus of ductal carcinoma in situ including papillary pattern was seen. All surgical margins were free of tumor. There was no perineural or lymphovascular invasion.

On gross examination of total sternectomy specimen, solid and cystic mass was identified, measuring 9.5x3.5x2 cm, involving sternum.(Figure 1). The attached 1^{st} to 7^{th} rib margins are away from mass. The microscopic examination is same as the breast lesions with sternal bone involvement. (Figure 3)

Immunohistochemistry performed on both the sites sternum and breast. IHC profile of breast tumour revealed CK7, ER, PR positivity and HER2 NEU, CK20 and CDX2 negativity (Figure 4 -IHC). In sternum lesion CK7, GATA 3, ER, PR were positive and CK20 and CDX2 were negative.

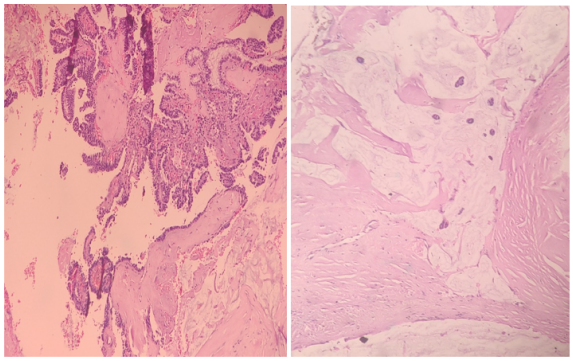

Fig. 2: Left breast lump

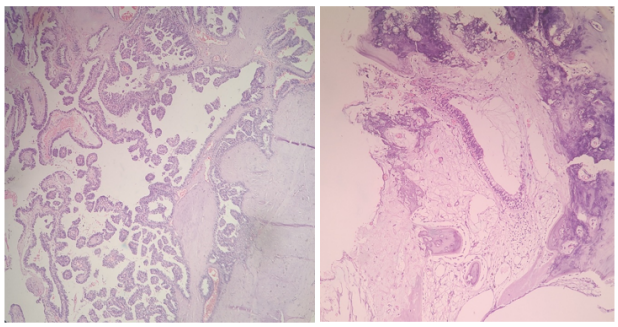

Fig. 3: Sternal metastasis

Based on this morphological and IHC findings the diagnosis was given as Primary mucinous cystadenocarcinoma of breast metastasizing to sternum. The patient was then finally discharged with regular follow-up.

3. Discussion

Mucinous cystadenocarcinoma of the breast is a rare variant of breast carcinoma which has a higher incidence in peri and post-menopausal women.[6] Mucinous carcinoma typically has two subtypes, the cystic (mucinous cystadenocarcinoma) and solid (columnar cell mucinous carcinoma).[7] Microscopically, there are cystic spaces which are lined by mostly bland-looking columnar mucinous cells with some papillary formations. Nuclear atypia is evident in some foci.[8] On immunohistochemical stain, estrogen receptor (ER), progesterone receptor (PR), Her2neu are negative.[4,9] We report a first case of a 35-year-old female with a primary mucinous cystadenocarcinoma of the breast metastasizing to sternum in addition to the immunophenotype ER (+), PR (+) and Her2 (-).

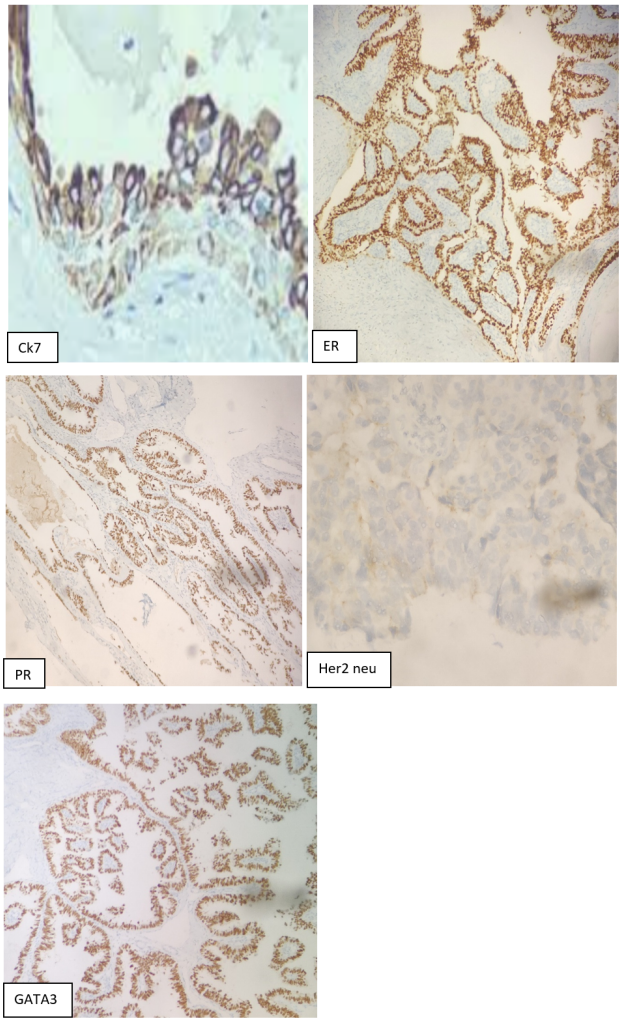

Fig. 4: IHC markers

4. Conclusion

A proper histological diagnosis and a special mention of the rare histologic subtypes are required to formulate clear recommendations of their treatment protocols. In the next decade with a routinary molecular evaluation could be possible to propose tailor treatments not only with drugs but surgical treatments as well.

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