Multifocal adenoid cystic carcinoma of the breast: not always a quiet course

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Abstract

Adenoid cystic carcinoma (ACC) of breast is a fairly uncommon malignancy, accounting for 0.1% of all cases of breast carcinomas. Though ductal or lobular breast carcinomas can be multifocal/multicentric, examples of the multifocal breast ACC is sparse. Hereby we report a case of retro-areolar multifocal ACC of the breast in a 36 year old female with typical grade 1 histomorphology. Characterization is important as ACC breast has an excellent prognosis with little chance of lymph node and distant metastases. However, our case metastasized after undergoing wide local excision. This example reminds both the physician and pathologist to consider ACC in the differential diagnosis of retro-areolar multicentric/multifocal breast carcinoma and the importance of mastectomy with axillary clearance in all histological grades.

Introduction

Adenoid cystic carcinoma (ACC) of the breast is uncommon, with a frequency of 1 case/ million female years, with a reported less aggressive course [1] than in ACC of other body parts [2]. Hereby, we report a case of multifocal grade 1 ACC of the breast which metastasized.

Case report

A 36 year-old female, with complaints of painless lumps in the right breast, showed two freely mobile ill-defined lumps. One of them was retro-areolar, measuring 3x2 centimetres (cm), while the other was in the upper outer quadrant, measuring 1x1 cm (Figure 1A). The mammogram graded them as BIRADS 5 (Figures 1B & C). PET computed tomogram showed no evidence of metastasis.

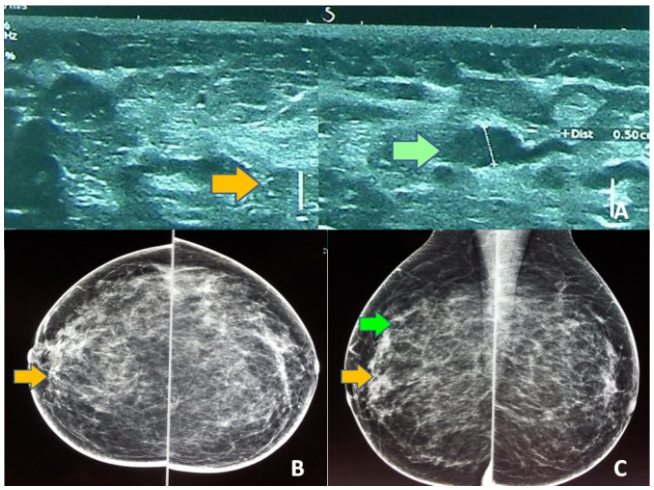

Figure 1. A - Ultrasonography showing right breast retroareolar (yellow arrow) and upper outer quadrant lesions (green arrow); B - Cranio-caudal view of mammography showing BIRADS V lesion in the retroareolar region of the right breast (yellow arrow); C - Medio-lateral oblique view of mammography showing two BIRADS V lesions in the retroareolar (yellow arrow) and upper outer quadrants (green arrow).

Core biopsies showed features of triple negative basaloid invasive carcinomas in both of them. Wide local excision with a sampling of sentinel lymph node was performed as the patient wanted breast conservation. Two circumscribed grey-white tumours, measuring 2.5 cm (retro-areola) and 0.8 cm (upper outer quadrant) were identified, with 1.6 cm width intervening non-tumour stroma. Histological showed solid sheets of basaloid hyperchromatic tumour cells with focal duct centric and cribiform arrangements, and luminal hyalinized cylindrical myxoid material (Figures 2 A-C). CD 117 and smooth muscle actin stains were positive, highlighting epithelial and myoepithelial cells respectively. Ki67 proliferation index was high (Figures 2 D-F). ER, PR and c erbB2 stains were

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negative. The sentinel lymph node biopsy was free of tumour; hence no axillary clearance was done during the second surgery. As the tumours were situated in two different quadrants with 1.6 cm of intervening normal stroma, a diagnosis of multifocal/ centric adenoid cystic carcinoma of the breast was given. Adjuvant first line chemotherapy and radiotherapy were given. However, barely 4 months later, she developed multiple metastatic liver lesions. Hence, second line adjuvant chemotherapy was administered.

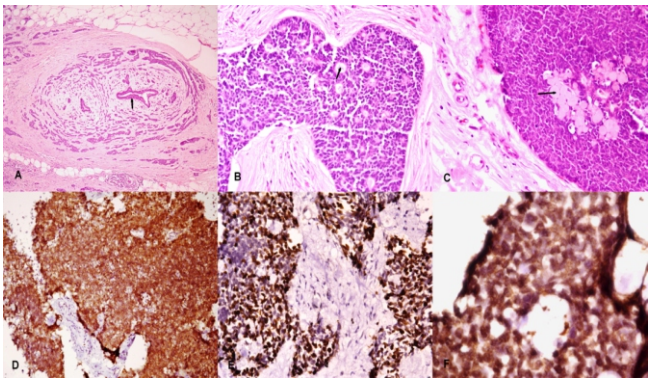

Figure 2. Photomicrographs show infiltrating cords of a basaloid breast tumour around the normal ducts (arrow) (Figure A, H&E x 40), with focal acinar/ rossettoid arrangements (arrow). The angulated tumour cells show hyperchromatic nuclei and scant cytoplasm (Figure B, H&E x 100). With luminal hyaline cylindrical materials (arrow) (Figure C, H&E x 100). CD117 stain shows strong positivity in the tumour cells (Figure D, IHC (CD117) x 100), along with high Ki67 labeling index (Figure E, IHC (Ki67) x 200) and smooth muscle actin positivity (Figure F, IHC (SMA) x 200).

Discussion

The commonest presenting complaint in an ACC is a painful lump of long duration [1]. Examples of multifocal ACC are extremely rare. In a large study by Ghabach et al., out of 338 cases of breast ACCs none of them was multifocal. Though ACC of the breast shows excellent prognosis, lymph node involvement and distant metastasis have been observed, albeit rarely. In the mentioned study only 5 of them showed lymph node metastasis and 1 showed distant metastasis [3]. Qizilbash et al demonstrated amongst 95 cases, one with node metastases and distant metastases in 6 [4]. Positivity for oestrogen and progesterone receptors, is also rare (6.2%) [2].

According to the study by Ro, et al., while local excision sufficed for histological grade 1 lesions; simple mastectomy with or without axillary clearance were required for grade 2 and 3 lesions subsequently [5]. Role of radiotherapy and chemotherapy was not sufficiently evaluated and hormonal therapy is believed not to be of much value [5]. In the index case also being a grade 1 lesion, local excision was performed followed by chemotherapy and radiotherapy. However, development of liver metastasis within a short span of time taught us that even in the multifocal breast ACC, mastectomy might be a better proposition. Robust data in this area is lacking due to the rarity of the disease, therefore a consensus is needed.

References

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Key Points:

- Multifocal breast ACC though extremely rare can be seen in young patients.
- Though they show an overall better prognosis than ACC in other body parts, mastectomy with axillary clearance is preferable irrespective of histological tumour grades.