Case Report

Adenoid Cystic Carcinoma of Breast: A Common Tumour at An Uncommon Site - A Case Report and Literature Review

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DOI: 10.21276/APALM.3277

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Submitted: 28-Aug-2023
Final Revision: 12-Sep-2023
Acceptance: 18-Sep-2023
Publication: 08-Dec-2023

Abstract

Adenoid Cystic Carcinoma is an uncommon malignancy of breast with favorable prognosis and distinct histomorphology. We present a case of 47-year-old female with palpable lump in right breast. Mammogram revealed an irregular hypoechoic solid mass at 10 'O clock position with indistinct margins. Trucut biopsy sections revealed a tumour showing pseudo glandular and true gland-like spaces lined by epithelial and myoepithelial cells and filled with abundant homogenous eosinophilic material. Immunohistochemistry revealed triple negative tumour cells, low Ki 67 (1-2 %) with p63 positive in myoepithelial cells and CD117 positive in epithelial cells. With these findings, it was opined as Adenoid cystic carcinoma right breast and the patient underwent wide local excision which we received for histopathological examination. On serial slicing, a solid circumscribed tumour measuring 1x1x2 cm was seen. Sections revealed concurrent morphological and IHC findings as seen on trucut biopsy. Patient responded well to surgery followed by radiotherapy. There has been no loco-regional recurrence or distant metastasis after a follow-up of 18 months.

Keywords:

Adenoid cystic carcinoma, breast, triple negative, malignant tumour

Introduction

Adenoid Cystic Carcinoma (AdCC) is primary salivary gland tumour which is also seen in sites like endocervix, skin, esophagus, uterine cervix, lungs, prostate and kidneys.[1,2] Being rare, it accounts for less than 0.1% of all invasive breast carcinomas and has a favourable prognosis.[3,4] Histomorphology shows proliferating epithelial, basal and myoepithelial cells with solid, tubular and cribriform patterns along with abundant basement-membrane like material.[1,5] Lumpectomy is considered gold standard as this tumour has very rare occurrence of local and distant metastases. We present a case diagnosed as classic AdCC on histopathological examination (HPE) along with literature review on subtypes and treatment.

Case Report

A 47 years old female presented with a history of palpable lump in the right breast of two weeks duration. On palpation, there was a small tender lump in upper-outer quadrant of right breast. Mammography of right breast revealed an irregular hypoechoic solid mass measuring 1.1 X 1.0 X 1.3cm , 6cm from the nipple at 10 ‘o clock position with indistinct margins (Fig.1a-b). There was no calcification in the mass and no enlarged axillary lymph nodes were noted. Contra-lateral breast appeared normal. Based on these findings, it was graded as BIRADS 4c - highly suspicious for malignancy and further confirmation by HPE was suggested.

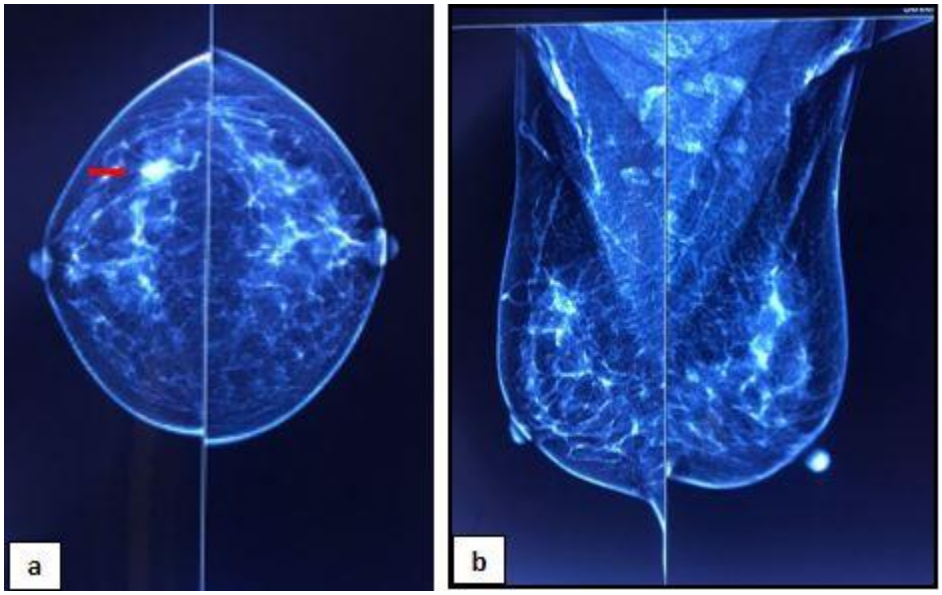

Figure 1: (a, b)Mammograph bilateral breasts: Right breast with an irregular hypoechoic solid mass measuring 1.1 x 1.0 x 1.3 cm at 10 ‘o clock position with inditinct margins (red arrow).

We received six linear cores following trucut biopsy from right breast, largest measuring 1.9x0.2x0.2cm. Hematoxylin and eosin (H&E) stained sections from biopsy revealed a biphasic tumour composed of inner ductal and outer myoepithelial cells arranged in tubular and cribriform pattern forming pseudoglandular and true glandular spaces. Myoepithelial cells had scant cytoplasm with hyperchromatic angulated nuclei. Gland like spaces were filled with abundant homogeneous eosinophilic membrane-like material (Fig.2a-b). Keeping in view the characteristic histomorphological findings, the diagnosis of AdCC- right breast was considered which was further confirmed by immunohistochemistry (IHC).

On IHC, tumour cells were triple negative (ER, PR and Her2 neu negative) with a low Ki 67 expression (1-2 %). Myoepithelial cells were highlighted by p63 and ductal epithelial cells showed positivity with CD 117 (Fig. 3a-b). Hence, final opinion of AdCC right breast was given in this case. Following the histopathological opinion on trucut biopsy, patient underwent wide local excision (WLE) along with right sentinel lymph node excision biopsy. WLE specimen measured 5x5x3cm and on serial slicing, a solid circumscribed whitish tumour measuring 1x1x2 cm was seen at the centre of the specimen. No areas of haemorrhage or necrosis were noted. Sentinel lymph node measured 3x2cm and appeared uninvolved on gross examination. H&E sections showed concurrent characteristic morphological and IHC findings as seen on trucut biopsy. Section from sentinel lymph node showed normal histology with no metastatic deposits.

Patient responded very well to WLE surgery followed by adjuvant radiotherapy and there has been no loco-regional recurrence or

distant metastasis after a follow-up of 18 months.

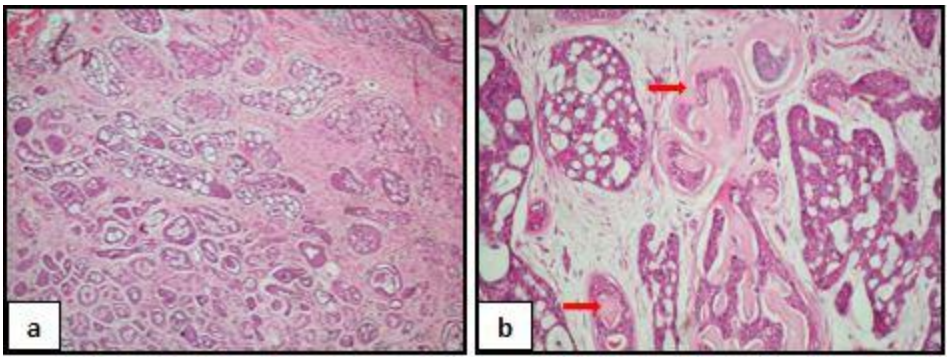

Figure 2: (a) H&E, 100x: Tumour arranged in tubular and cribriform pattern; (b) H&E, 400x: glandular and pseudo glandular spaces filled with and surrounded by eosinophilic basement-membrane like material (red arrow).

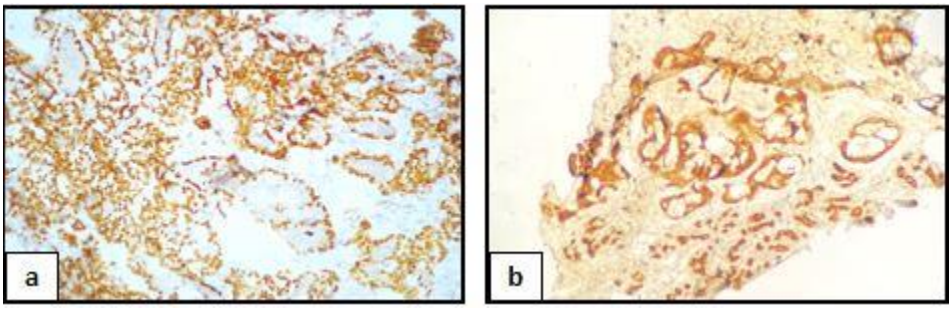

Figure 3: Immunohistochemistry, 200x: (a) p63 highlighting the myoepithelial cells; (b) CD117 highlighting the epithelial cells.

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Discussion

AdCC was earlier termed as “cylindroma” by Billroth in 1856. Later in 1945, Geschickter first described AdCC of breast.[4] Being most common in salivary glands and bronchi, this tumour is also seen at sites like endocervix, skin, esophagus, uterine cervix, lungs, prostate and kidneys.[1,2] Being rare, AdCC accounts for less than 0.1% of all primary breast invasive carcinomas and as opposed to the aggressive course with poor prognosis in salivary glands, it has a favourable prognosis as primary breast tumour.[3,4]

Being predominantly seen in fifth and sixth decade, AdCC of breast can occur over a wide age range of 28 to 90 years.[3] It usually presents as a well circumscribed tender mass in subarealor region which can range from 1 to 12cm in size (mean 3cm). Some may present with nipple retraction. There is no side predilection and is rarely bilateral.[2,4,6] Our patient also presented with tender breast lump which was within the mean size range as mentioned in literature. Nodal and distant metastasis is very rare though have been reported in lung, bone, kidney and liver.[6] Mammography is of limited use in differentiating AdCC from other invasive carcinomas as there are no characteristic findings attributing to diagnosis of AdCC. It usually appears as a sharply marginated and circumscribed lesion with extremely rare microcalcifications, which sometimes may be missed on imaging. On ultrasonography, the lesion appears hypoechoic[1,2,7] In our case, lesion was well picked up by mammography as a malignant tumour. Fine needle aspiration cytology and core needle/ trucut biopsy are commonly performed methods to reach at a conclusive diagnosis of breast lump.[3]

Histopathologically, AdCC is a mixed tumour where tumour cells differentiate towards epithelial, basal and myoepithelial cells which may show three types of patterns: solid, tubular and cribriform. There is abundant laying of hyaline or myxoid basement-membrane like material. There are two subtypes of salivary gland AdCC in WHO classification: (a) tubular/cribriform subtype and (b) solid subtype.[1,5] As commonly seen in salivary gland AdCC, perineural invasion is rare in AdCC of breast. Grading of AdCC is based on percentage of solid component in whole tumour and higher grade implies an aggressive behaviour with poorer prognosis. Grade I tumours have only tubular/cribriform pattern with no solid component; grade II tumours have <30% of solid component; and grade III tumours have ≥ 30% of solid component.[1,2] Our case had <30% solid areas, and so it was graded as grade II AdCC of breast. Although AdCC breast is a triple negative tumour (ER, PR, Her2 neu negative) on IHC, it still behaves as low-grade malignancy.[1,2] Other important IHC markers are p63 and CK5/6 highlighting the basal cells whereas ductal epithelial cells are positive for CD117 and CEA. Some of the differential diagnoses which need to be considered include: collagenous spherulosis and cribriform pattern of ductal carcinoma in situ (DCIS) for the tubular/cribriform subtype and solid papillary carcinoma, neuroendocrine carcinoma and metaplastic carcinoma of solid subtype.[7] Though myoepithelial cells in both collagenous spherulosis and AdCC express nuclear positivity for p63, ductal epithelial cells in AdCC show cytoplasmic and membranous positivity with CD117.[8] Cribriform DCIS is ER, PR positive and lacks myoepithelial layer, hence it does not express p63. Only a very small proportion of cases of cribriform DCIS are positive for CD117. Invasive cribriform carcinoma is negative for both p63, CD117 and Her2 neu while it expresses ER/PR.[8,9] Our case expressed p63 in myoepithelial cells and CD117 in the ductal epithelial cells, along with being triple negative for hormone receptors, thus, favoring the diagnosis of AdCC of breast.

Simple resection with lumpectomy without lymph node dissection is the treatment of choice in these tumours. Sentinel lymph node biopsy is usually considered enough as nodal and distant metastases are extremely rare. There is some evidence of improvement in loco-regional disease control by giving post-operative adjuvant radiotherapy.[2] In our case also, patient underwent WLE with sentinel lymph node biopsy followed by adjuvant radiotherapy and response to surgery has been very well as after a follow-up of 18 months, patient is disease free with no evidence of local/ distant metastases.

Conclusion

AdCC of breast is a rare malignant tumour with a favourable prognosis. Prompt diagnosis can save the patient from undergoing mastectomy for invasive carcinoma as lumpectomy with adjuvant radiotherapy is the most widely accepted treatment protocol for this tumour. Follow-up post surgery is required to keep a watch on rare but likely occurrences of local and distant metastases.

Abbreviations

AdCC: Adenoid Cystic Carcinoma
HPE: Histopathological examination
H & E: Hematoxylin and eosin
IHC: Immunohistochemistry
WLE: Wide local excision
DCIS: Ductal carcinoma in situ

Acknowledgements : *None*

Declarations: Nil

Statement of informed consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patient(s) understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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