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Original Research Paper

ADENOID CYSTIC CARCINOMA OF THE BREAST : A CASE REPORT AND LITERATURE REVIEW

Oncology

KEYWORDS: adenoid cystic carcinoma (ACC), triple negative carcinoma, Breast Conservation Surgery (BCS) , adjuvant therapy

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ABSTRACT

Adenoid Cystic Carcinoma of Breast (ACC) is a very rare malignant neoplasm of breast accounting for <0.1 % all patients diagnosed with breast cancer. To date, only 6 cases have been reported in India. It has indolent course with favourable prognosis, despite its triple negative status. Radiologically, its appearance is nonspecific but its diagnosis can be made by FNAC. Here, we report a female with breast lump initially diagnosed as lobular carcinoma on FNAC. She underwent lumpectomy which on histopathology was ACC. Subsequently, she underwent Breast Conservation Surgery for scar and residual lump excision. Histopathology report was the same without neural & nodal involvement. Postoperatively, pt. not advised any adjuvant therapy and currently doing well6 months postoperatively. Although surgery is the main treatment, due to its rarity, there has been no consensus regarding established treatment protocol.

Introduction

ACC of breast was first reported by Geschickter and Copeland in 1945(1). It is previously called as "Cylindroma". It is very rare type of primary breast carcinoma accounting <0.1% of total(2). Although most frequently seen in salivary glands, it is also seen in cervix,skin, lung, kidney, eosophagus & prostate. The affected age group ranges from 19 -97 years, but more common in 50 – 60 year age old(3-5). Typically, patients present as painful breast lump in subareolar region without lymph node involvement(6-7). Histologically, it shows various patterns like cribriform, solid, nodule, tubular, basaloid & nesting. Although the exact site of origin remains unknown, it is estimated to originate from ductal epithelium or myoepithelium. It is triple negative tumours but its prognosis is excellent(3,8). The 10 years survival rate for patients with ACC breast ranges from 85-100% (9). It has low local recurrence and rare distant metastasis (10,11). The aim of this report is to review its clinicopathological features and preferred treatment modalities in the light of existing literature.

CASE REPORT

A 55 years old female came to our hospital with complaints of recurrent lump in right breast since ten days. Patient had history of wide local excision of painful lump since two months at the same site fifteen days back. Patient was having outside FNAC report before excision as suspicious Lobular carcinoma. Histopathology of wide local specimen was Adenoid cystic carcinoma, 2.1cm × 1.6cm × 1.3cm size, moderately differentiated, nuclear grade II, with 25% solid and 10% high grade solid area with necrosis. Minimum negative margin was 2mm.

Patient didnot have family, personal history of any type of cancer,not suffering from any chronic illness and not addicted to any form of tobacco. Patient had history of hysterectomy around 15 years back.

On examination, patient had linear fresh scar 4cm in size over upper outer quadrant of right breast with underlying moderate consistency lump around 3 cmx3cm adhered to scar. Rest of the bilateral breast examinations were normal. There was no palpable lymph node in bilateral axilla & clavicular area. Systemic examinations were unremarkable. All routine haematology were normal.

Slides and blocks sent for review in pathology department. On mammography, ill-defined soft tissue density noted in upper outer quadrant of right breast which on sonography showed 47mm×29mm×43mm sized anechoic lesion below surgical scar site suggestive of postoperative collection. All metastatic work up – chest x-ray, ultrasonography (abdomen & pelvis), CT scan (thorax & abdomen) were normal. The histopathology review report was-specimen CK7 positive, AE1 positive and ER/PR/Her2nu negative. IHC diagnosis was adenoid cystic carcinoma.

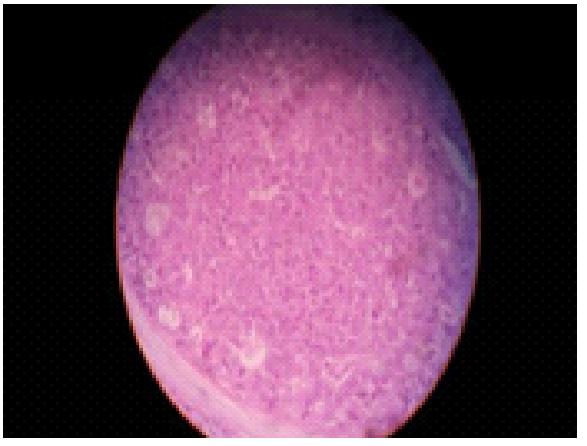
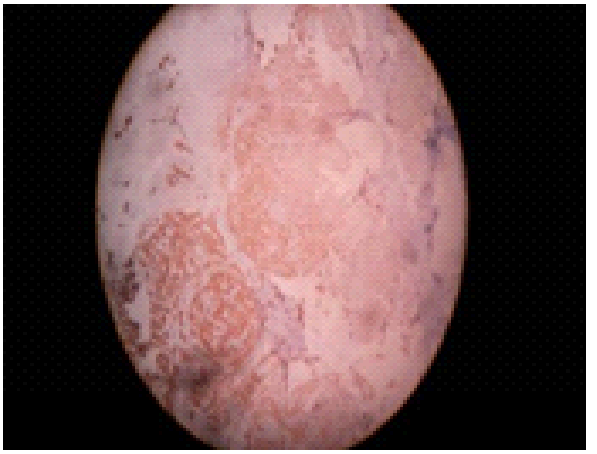


Fig.1 HPE and IHC of lump



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Patient planned and underwent Breast Conservation Surgery for excision of scar and underlying lump. The final histopathology report was – 4.5cm×4cm×2cm hard cystic area with inflammatory changes and no evidence of residual tumour. All soft tissue resected margins and base of resection were free of tumour. Submitted two lymph nodes were free of tumour. Patient not advised any adjuvant therapy. Now patient is 6 months postoperative, on regular follow up and is doing well without any recurrence or metastasis.

Discussion

Adenoidcystic carcinoma was initially described as "Cylindroma " by Billroth in 1856 (3), but ACC of breast was first described by Geschickter in 1945 (1). Its incidence is one per million females per year (1). It is very rare malignancy accounting <0.1% of breast neoplasms (2). Most of the knowledge of breast ACC is derived from individual case reports and few clinical case series. In literature, Only 6 cases of ACC breast have been reported in India (23-28).

Breast ACC are commonly detected in postmenopausal females. However, cases in children, geriatric and male population are also reported in literature (12). The left and right breast are equally affected and there is no tendency for bilaterality. Most common presenting symptoms are palpable breast lump followed by pain and nipple retraction. Qizilbalshet al(13) pointed out a tendency of these tumours to be located in the nipple areola area. Although the tumour size is usually 2-3 cm, the existing literature contains cases with tumour size of 15 cm. our patient profile, presentation and tumour size all were in concordance with the literature but tumour was located in upper outer quadrant not agreeing with the literature.

The diagnosis can be made by FNAC (23,24) but core needle biopsy is confirmatory. It has dual cell composition- luminal epithelium which stains positive for PAS, and basal myoepithelial cells. Morphologically, it has three architectural pattern- tubular, cribriform and solid. The architectural pattern is of prognostic importanceas solid variant has more aggressive course with tendency for lymph node metastasis (7). Immunohistochemistry of breast ACC is generally negative for Estrogen, Progesterone and Her 2 nu receptors, but it doesnot behave like triple negative breast carcinoma (TNBC) (14). It is also CK7, AE1, EGFR, C-Kit positive (15) and androgen receptor (AR) negative (14). Perineural invasion is generally not in seen breast ACC (16). In the present case ER, PR and Her 2 nu receptors were negative and CK7, AE1 were positive as per literature and also having no perineural invasion.On mammography, the tumour is characterised as lobulated lesion with regular or irregular margin with extremely rare microcalcifications. On sonography it appears as hypoechoic solid or heterogeneous mass. The radiological characteristic of this patient exactly matches with the literature.

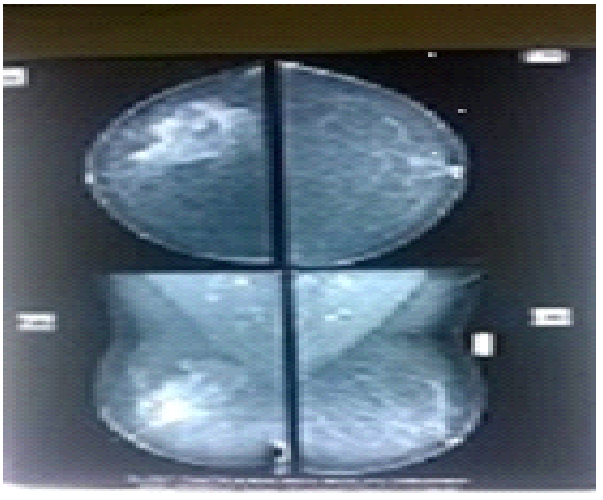

Fig. 2. Non specific mammographic findings of lesion

Treatment of breast ACC is mainly surgical ranging from Breast Conservation Surgery (BCS) to mastectomy(17). There has been no prospective trial to compare their efficacy but in most cohort studies local recurrence after mastectomy and BCS were 0% and 6% respectively (18)though the local recurrence after BCS did not affect overall survival (19). Axillary lymph node involvement is extremely rare occurring in 0-2% of breast cases (3), Sumpio et al reviewed 182 cases who underwent axillary dissection, lymph node metastasis were noted only in 4 patients (17). In Rare Cancer Network (RCN) study of 61 patients, axillary dissection done in 41 patients (67%) and Sentinel lymph node biopsy done in 10 patients (16%), all of them reported negative. An average of 79 months follow up of these patients, there were no axillary, supraclavicular or internal mammary metastasis (18). Therefore, axillary dissection should not be done except in the presence of nodal metastasis. Moreover, sentinel lymph node sampling can be done if tumour is >3cm, high grade or contains other types of breast cancer. Ro et al (19) divided the disease into 03 grades depending on the degree of solid growth pattern into the tumour.

RO et al Histological Gradings of ACC and its Management

GRADES	HISTOLOGICAL CHARACTERISTICS	MANAGEMENT
GRADE I	Cystic without solid componenet	Local Excision
GRADE II	<30% solid components	Mastectomy
GRADE III	>30% solid components	Masectomy & Axillary Dissection

In the present case patient underwent BCS due to lesion being grade II and size <3cm.

A recent large retrospective study by the Surveillance, Epidemiology and End Result (SEER) included 376 patients of breast ACC found that adjuvant radiotherapy after local surgical therapy improved both disease free survival and overall survival (20). Though, the indication for adjuvant radiotherapy in breast ACC patients have not yet been determined. The role of adjuvant systemic chemotherapy and hormonal therapy is controversial. Arpino et al (21) and Mcclenathan and de la Roza (22) reported that adjuvant chemotherapy and hormonal therapy do not increase the survival rate of patients. Here,patient not adviced for adjuvant radiotherapy or chemotherapy.

Prognosis of breast ACC is excellent as there is rare local, regional or distant metastasis. In Surveillance, Epidemiology and End Result (SEER), Ghabach B et al reported 5 year, 10 year and 15 year survival rate to be 98%, 95% and 91% respectively (10). Veeratherapillay R et al reported similar survival rates with 100% 5 year disease free survival (4). The surveillance period of our patient should be long as breast ACC has tendency for late recurrence and she has completed only 6 months postoperatively.

TABLE: Cases Reported in India And Its Clinicopathological Characteristics & Treatment Outcome.

Case NO.	Reference	Age/Sex	Presentation	Surgery	Adjuvant	Recurrence	DFS (months)	Conclusions
1	A. Malik et al	55/F	painless lump lt. breast (cT2N0)	BCS + ALND	CT ->RT	NO	144	FNAC may be diagnostic
2	Amrish N. et al	66/F	Painless lump lt. breast (cT1N0)	WLE	NA	NA	NA	FNAC may be diagnostic
3	Palaniappam M. et al	48/F	B/L painful breast lump (cT3N0)	B/L MRM	CT ->RT	NO	36	ACC may be B/l, painful.
4	Suresh B. et al	30/F	Multiple Painless lump rt. Br. (cT2N1)	Rt. MRM	NA	NA	NA	Axillary LN may be involved
5	Barath R. et al	40/F	Painless lump lt. breast (cT1N0)	BCS + ALND	RT	NO	12	Can be mistaken for DCIS
6	Neha B.et al	59/F	Painful lump lt. breast (cT2N0)	BCS + ALND	NONE	NA	NA	Adjuvants not required
			THIS CASE					
7	Amit k et al	55/F	Painful lump rt. Brest (cT2N0)	BCS	NONE	NO	6	ALND and Adj. not needed

ALND: Axillary Lymph Node Dissection, BCS: Breast Conservation Surgery, B/L: Bilateral, NA: Not Available, Adj. : Adjuvant treatment

Conclusion

Every oncologists must be familier with this carcinoma of breast, that is extremely rare malignancy with extremely good prognosis. Though the additional clinical experiences are needed for established treatment guidelines, Breast conservation surgery without or with axillary lymph node dissection, without or with adjuvant radiotherapy are currently recommended as per literature. Solid variants must be treated aggressively. Also, Overexpression of EGFR and C-Kit in ACC of breast may possibly open doors for targeted therapies. Long term close follow up is required due to rare but possible distant metastasis.

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