

# Adenoid cystic carcinoma of the breast: a study of five cases

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## ABSTRACT

Adenoid cystic carcinoma of the breast is a rare type of breast cancer. Most patients present with a bilateral palpable mass. Ultrasound and mammography are non-specific and can sometimes lead to misdiagnosis because of their variable imaging features. Pathological examination is the standard reference. Surgery is the mainstay of treatment for patients. Although adenoid cystic carcinoma has an excellent prognosis, metastatic cases have been reported. This report aims to discuss the clinical and imaging features of one case of adenoid cystic carcinoma with a poor prognosis and four cases with a good prognosis at our center.

## CASE SERIES

### CASE SERIES

Five women with a mean age of 55 years (range, 49-61 years) were referred to our center with palpable masses from 2014 to 2018 (Table 1). Patient 4 and 5 were in their menopausal period, while patients 1, 2, and 3 were not. All patients had no relevant family history. Four patients were located in the outer quadrants while patient 3 was in the inner quadrant.

All patients underwent breast ultrasound and mammography examinations (Figures 1-5). Each tumor presented as a hypoechoic solid or heterogeneous mass with minimal vascularity on ultrasound. Ultrasound did not detect suspicious axillary lymph nodes in any of the patients. Each tumor presented as a dense node without calcification on a

mammogram. Patient 1 presented with a large regularly shaped tumor with a circumscribed margin, while the other four patients had relatively small tumors, which were irregularly shaped with uncircumscribed margins. All patients were category 4 or 5 according to the Breast Imaging-Reporting and Data System (BI-RADS) assessment, and core biopsy was recommended. More details are shown in Figures 1-5.

Four patients underwent mastectomy, while patient 3 underwent breast-conserving surgery (BCS). The tumor diameter ranged from 20 mm to 65 mm. After surgery, patients 2 and 3 underwent extra adjuvant chemotherapy with fluorouracil, epirubicin, cyclophosphamide, and taxinol for four cycles, and doxorubicin and cyclophosphamide for one cycle. Patient 3 underwent adjuvant radiotherapy. Bone and

lung metastases were detected in patient 1 in the fourth year after surgery, while all other patients remained recurrence-free during the follow-up period. The deadline for follow up was June 2020. More details are shown in Table 1.

## DISCUSSION

### Etiology & Demographics:

Adenoid cystic carcinoma (ACC) is among the most common salivary gland malignancies. It also affects other exocrine tubulo-acinar glands, such as those found in the breast. ACC of the breast is a rare type of cancer that accounts for less than 1% of all breast cancer [1,2].

A large number of affected patients are female; however, males are also affected [3,4,5]. Although ACC can occur at any age, most cases are diagnosed in the fifth or sixth decades of life [6,7]. Consistent with current studies, the mean and median ages of the five patients in this study were 55 years and 54 years, respectively (range, 49-61 years), which is the age at which breast cancer is most likely to occur in Chinese women [8]. In Ghabach's research, 338 cases of ACC were analyzed in the United States from 1977 to 2006 with mean and median ages of 63 years and 62 years, respectively (range, 33-97 years). Women aged 50 years or older had an 11-fold higher incidence of ACC compared with younger women [2].

### Clinical & Imaging findings:

A palpable mass was the chief complaint in most patients. Imaging features were variable. The imaging characteristics of breast ACC have not been well described in previous studies. Previous studies with a relatively large number of patients have mainly focused on the pathology, immunohistochemical characteristics, treatment, and prognosis. For example, 933 cases of ACC from the National Cancer Data base were analyzed in Kulkarni's article [7], while 338 cases of ACC from the Surveillance, Epidemiology, and End Results Program were analyzed in Bassam Ghabach's research [2]. However, these two studies, which had large sample sizes, did not examine the imaging features of ACC. According to Wei Tang's research into the use of multiple imaging techniques in 11 patients, ACC presents as an irregular or lobulated mass with indistinct or spiculated margins on mammography. Calcification are not detected. Sonographically, most ACCs appear as hypoechoic solid or heterogeneous masses with minimal vascularity on color Doppler examinations [9]. These features are consistent with our findings. The margins are often not well circumscribed; rather, they are angular, indistinct, or microlobulated [9], which makes them easy to categorize as BI-RADS category 4 for further biopsy. However, the tumor margin can also be circumscribed, as it was in patient 1 in the present study. In Huang's case report of ACC, sonography revealed a homogeneous hypoechoic mass with a circumscribed margin and posterior enhancement [10], which mimicked a benign tumor. This observation can challenge radiologists to make a correct diagnosis. Further multi-center analyses are needed to systematically analyze and compare the imaging features of other pathological types of breast cancer.

Histopathologically, ACC is characterized by a biphasic pattern, consisting of a proliferating glandular component and a stromal or basement membrane component [11]. Different architectural growth patterns, such as cribriform, tubular, trabecular, and solid, or a mixture of the above, can be observed in ACCs [1]. ACCs can be divided into three categories: grade 1 tumor without a solid component, grade 2 tumor with a solid component of less than 30%, and grade 3 tumor with a solid component of more than 30% [12]. ACC is distinguished from other subtypes of breast cancer by immunohistochemistry. It is often estrogen receptor (ER)-negative and progesterone receptor (PR)-negative, and it does not express the human epidermal growth factor receptor 2 [13], which is consistent with our study. However, the National Cancer Data base review of ACC found that 15.4% of cases were ER-positive and 13.3% were PR-positive [7]. The two distinct ACC cell types, basaloid and luminal, are best appreciated by immunohistochemical staining. Basaloid cells express basal cytokeratins (Cks), such as Ck14 and Ck17, vimentin, S-100, actin, calponin, and p63, while epithelial cells stain positive for luminal Cks, such as Ck7, carcinoembryonic, epithelial membrane antigen, and CD117 (c-KIT) [14].

### Treatment & Prognosis:

There is no consensus regarding the optimal treatment strategy for ACC. BCS is performed more commonly in patients with ACC compared with patients with invasive ductal carcinoma (IDC) (69.8% vs. 59.8%, respectively;  $p < 0.0001$ ) in the United States [7]. However, in our study, only one of five patients underwent BCS. Patients' needs for BCS, medical costs and disease severity should be considered when deciding the surgical plan. According to Fan Lei's review of breast cancer in China, even in developed urban areas, such as Beijing and Shanghai, the incidence of BCS was only 12.1% in 2005, climbing to 24.3% by 2008 [8]. Some older patients with breast cancer prefer to undergo mastectomy. To add, radiation therapy needs to be administered as part of BCS. The cost of radiotherapy will increase the financial burden of some patients, which impacts the confidence of patients. Romeira recommended local excision for grade 1 ACC, simple mastectomy for grade 2 ACC, and modified mastectomy for grade 3 ACC based on a pathology classification system [15]. Welsh suggested that surgical axillary staging may not be necessary for patients with pure ACC subtypes. Preoperative axillary ultrasound with fine needle aspiration of suspicious nodes accurately predicts pathological nodal stage; therefore, axillary surgery might be omitted safely in patients with pure ACCs and a clinically negative axilla [16]. Thompson suggested that unless there are palpable lymph nodes or other reasons to suspect nodal involvement, routine axillary lymph node biopsy or excision may not be warranted, especially with T1 tumors [6].

ACC is often a triple-negative breast cancer [13], but it has an excellent prognosis with an overall 5-year relative survival rate of 98.1% due to its indolent growth [2]. Local recurrence rates are low (3%-18%), and if growth dose occurs, more than half of these patients can be cured with further surgery [17]. Axillary lymph node involvement is observed in <2% of patients. ACC Metastasis is uncommon, but when it does occur, the lungs are most commonly affected, followed

by the liver, bones, and kidneys. Rare cases of brain and scalp metastases have also been reported [18,19]. So far, the reason for the poor prognosis in patients with ACC is not clear. As the size of the tumor increases, the incidence of positive lymph nodes also increases [6]. Breast cancer prognosis is strongly associated with tumor size at diagnosis [20]. In our study, one of the apparent differences between patient 1 with a metastatic prognosis and other patients with a recurrence-free prognosis was tumor size, which was associated with tumor staging. Patient 1 was pathologically staged as T3 after surgery, while the other four patients were staged as T1 or T2. This indicates that the risk of ACC metastasis may be proportional to tumor size. In Ghabach's research, the mean and median tumor sizes among 229 women diagnosed in 1988 or later were 2.1cm and 1.8cm, respectively [2]. Mhamdi reported a case of ACC measuring 8 cm (T3) with pulmonary, kidney, and brain metastases [18]. Spiliopoulos reported a case of ACC measuring 6 cm (T3) with axillary lymph node metastasis [21]. These reports suggest that a small tumor size might positively impact the prognosis of patients with ACC, while a large tumor size might be a risk factor for poor prognosis. Detection, diagnosis, and treatment in the early stages of disease are important for a good prognosis. Regular follow up is also important after surgery. The prominent difference on ultrasound between patient 1 and the other four patients in this study was the tumor margin. Patient 1 had a circumscribed tumor margin and the other patients had uncircumscribed tumor margins, such as indistinct, angular, or microlobulated margins. A large number of patients with ACC are needed to study the difference between patients with a good prognosis and those with a poor prognosis.

#### *Differential Diagnoses:*

A painless solitary mass with a heterogeneous texture, a circumscribed or microlobulated margin, posterior enhancement, minimal vascularity and without calcification and suspicious lymph nodes can be a differential diagnosis for ACC.

ACC image features are variable. When ACC presents as an irregular mass with uncircumscribed margin, it should be differentiated from other malignancies. However, it is not difficult for a radiologist to classify it as BI-RADS category 4 for further biopsy to avoid a missed diagnosis. However, when ACC presents as a regular mass with a circumscribed margin, it is hard to distinguish it from benign tumors, such as fibroadenoma and phyllodes tumor. Histopathology and additional immunohistochemistry are helpful for differential diagnoses [11].

#### **Invasive ductal carcinoma:**

IDC accounts for approximately 50%-80% of breast cancers [22]. IDC may have a variable posterior acoustic appearance that ranges from shadowing to unchanged to enhanced. An unchanged posterior signal is more common with IDC than either shadowing or enhancement when images using ultrasound, while posterior enhancement is more common with ACC. Microcalcification can be observed by mammography in patients with IDC, while ACC usually has no calcification. On T2-weighted magnetic resonance imaging (T2W MRI), larger adenoid cystic lesions demonstrate very

strong signals, whereas IDC appears as hypointense or isointense in most cases [10,23].

#### **Fibroadenoma:**

Fibroadenoma is a common benign neoplasm of the breast. Fibroadenoma usually presents as a painless, firm, mobile breast lump, often in younger women aged 20-35 years. Fibroadenomas often appear as well-circumscribed isohypoechoic homogeneous masses on ultrasound. Mammograms sometimes show an oval or lobulated circumscribed mass. Dense coarse calcification may be seen in some cases. On T2W MRI, fibroadenoma appears as homogeneous and hypointense or isointense in most cases. In addition, some fibroadenomas have non-enhancing internal septations, while the internal septations of ACC lesions have been described as delay-enhanced, especially with larger lesions [10,24].

#### **Phyllodes tumor:**

Phyllodes tumors are rare fibroepithelial neoplasms of the breast that accounts for 0.3%-1.0% of all breast tumors. Phyllodes tumors can occur at any age with a peak incidence between 30 and 40 years, while ACCs usually occur in the fifth decade of life. Most phyllodes tumors present with large, well-defined, rounded or lobulated masses on medical imaging. Calcification is rarely detected with mammography. Phyllodes tumors usually appear as palpable, enlarged, painless masses with a diameter over 3 cm, while ACCs usually present as a small mass with a median tumor size of 1.8 cm [2,25,26].

#### TEACHING POINT

Adenoid cystic carcinoma (ACC) is a rare type of breast cancer that often presents as a painless solitary mass with a heterogeneous texture, a circumscribed or microlobulated margin, posterior enhancement, and minimal vascularity without calcification or suspicious lymph nodes on clinical and imaging examinations. Sometimes ACC can mimic a benign tumor, such as a fibroadenoma or a phyllodes tumor. Even if ACC resembles a benign tumor, prognosis can be poor.

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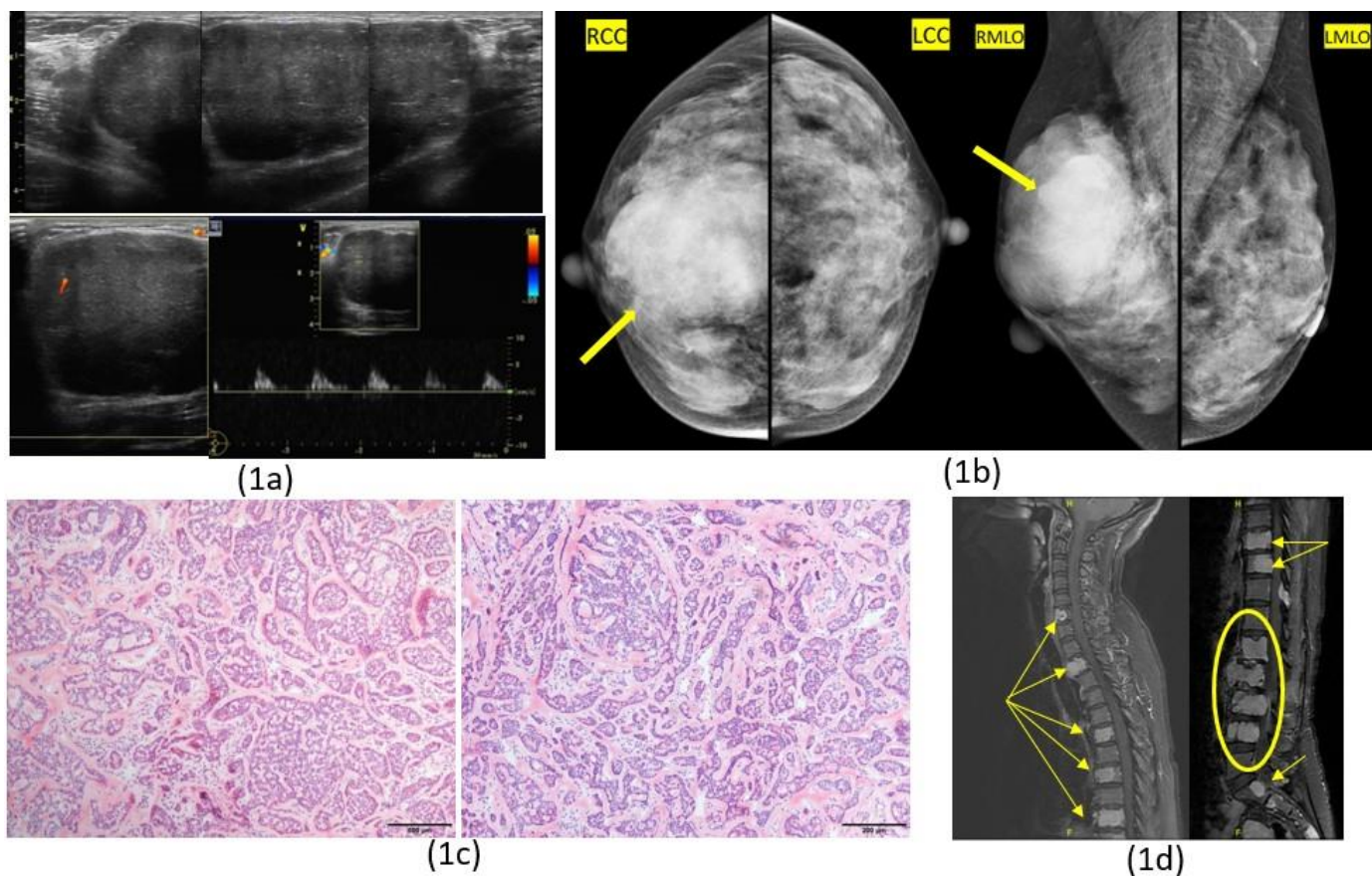
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## FIGURES



**Figure 1:** 49-year-old woman with adenoid cystic carcinoma of the right breast.

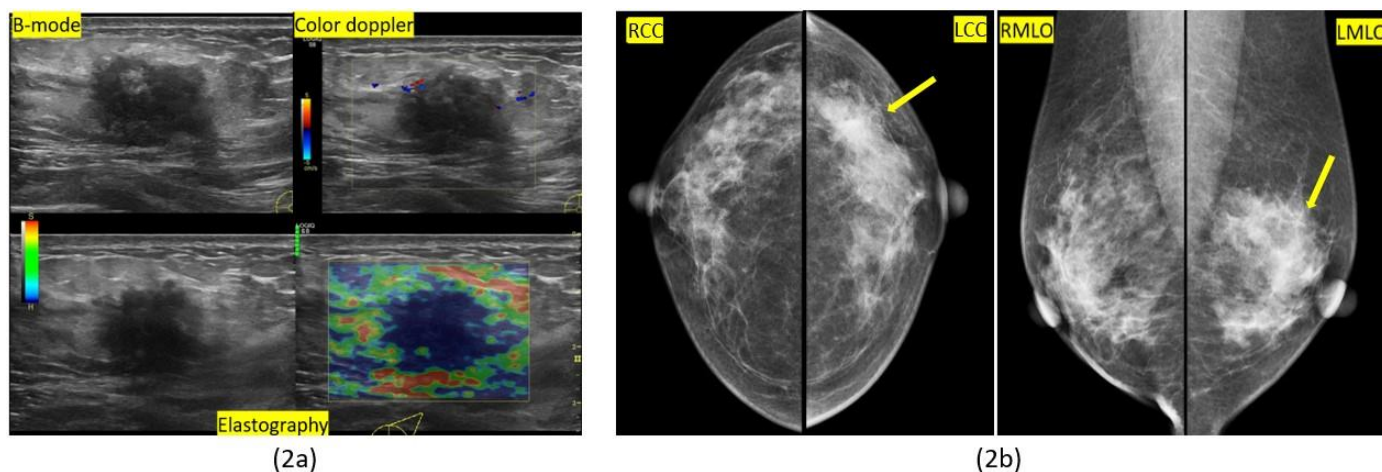
## FINDINGS:

Figure 1a An oval hypoechoic heterogeneous solid mass ( $41 \times 31$  mm) with a circumscribed margin, a parallel orientation, and posterior enhancement is shown on B-mode ultrasound. Color Doppler shows minimal vascularity and an arterial spectrum is detected by pulse wave Doppler. Figure 1b Mammography shows bilateral diffuse heterogeneous dense breasts. A hyperintense irregular large mass ( $56 \times 52$  mm, yellow arrow) with a sharp margin but without calcification is seen in the upper outer quadrant of the enlarged right breast. Figure 1c Hematoxylin and eosin stained sections show tumor cells arranged in small dispersed nests with cribriform growth and moderate cytoplasmic staining. Figure 1d After 53 months, sagittal contrast-enhanced T1-weighted magnetic resonance imaging demonstrates multiple lesions with strong signals (yellow arrow and circle) which reflect vertebrae metastases.

## TECHNIQUE:

Figure 1a Ultrasound using the GE Vivid 7 (GE Healthcare, USA) ultrasound machine with a 12L linear transducer. Figure 1b Mammogram using the Senographe DS (GE Healthcare, USA) (28 kVp and 48mAs). Figure 1c Hematoxylin and eosin stained sections:  $\times 500$  and  $\times 200$ . Figure 1d magnetic resonance imaging using the SIGNA Pioneer (GE Healthcare, USA); sagittal SE water T1-weighted IDEAL +C, 3.0 T; slice thickness, 3 mm; repetition time, 444 ms, echo time, 10.144 ms, contrast material, 10ml of Multihance (Bracco, Italy)





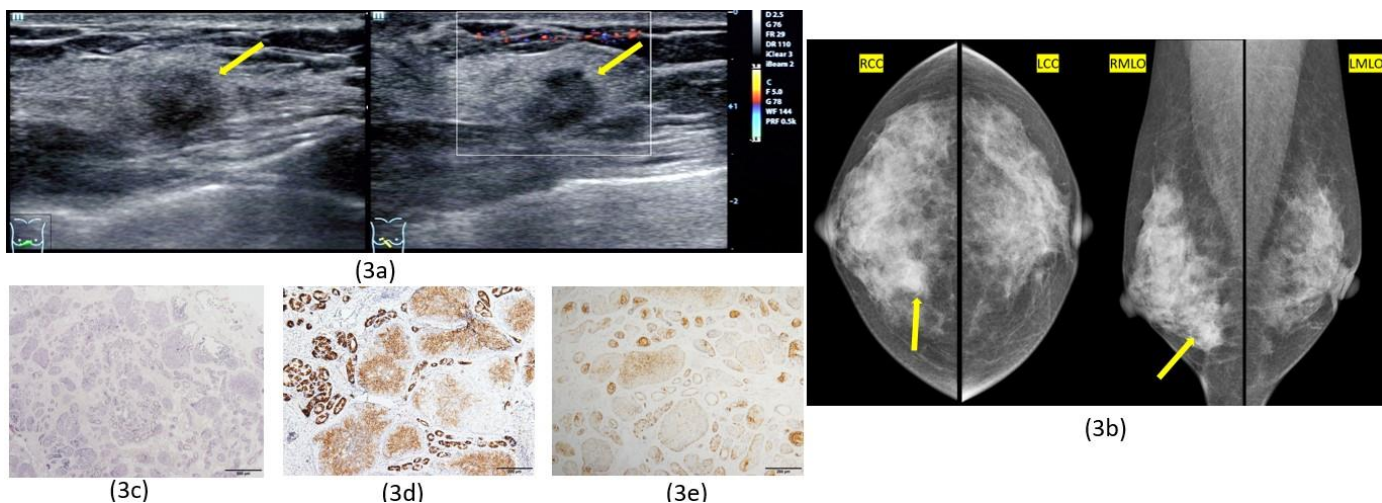
**Figure 2:** 50-year-old woman with adenoid cystic carcinoma in the left breast.

#### FINDINGS:

Figure 2a An irregular heterogeneous mass (20 × 17 mm) with an indistinct, angular, microlobulated margin, a parallel orientation, and posterior shadowing is shown using B-mode ultrasound. Color Doppler shows minimal vascularity. The entire area of the tumor and its surrounding area are blue on elastography. The elasticity imaging score of the lesion was 5. Figure 2b A mammogram demonstrates bilateral scattered fibroglandular tissue. An irregular hyperintense mass (14 × 11 mm) with an indistinct margin is seen in the outer quadrants of the left breast (yellow arrow). No suspicious microcalcification is noted.

#### TECHNIQUE:

Figure 2a Ultrasound using the LOGIQ S8 (GE Healthcare, USA) ultrasound machine with a 6-15L linear transducer. Figure 2b Mammography using the Senographe DS (GE Healthcare, USA) (31 kV and 32 mAs).



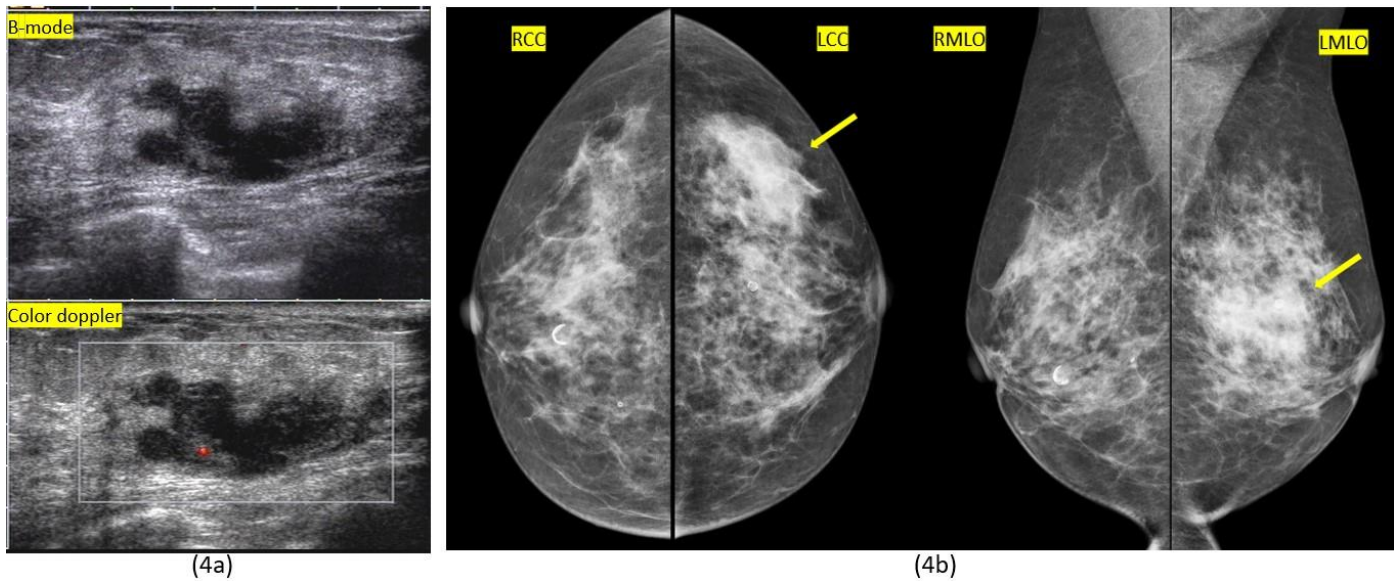
**Figure 3:** 54-year-old woman with adenoid cystic carcinoma of the right breast.

#### FINDINGS:

Figure 3a A heterogeneous hypoechoic solid mass (11 × 8 mm, yellow arrow) with a microlobulated margin, an unparallel orientation, and posterior enhancement is shown on ultrasound. No vascular signal is detected inside the tumor on color Doppler. Figure 3b Mammography demonstrates bilateral heterogeneous dense breasts. A hyperintense irregular mass (14 × 11 mm, yellow arrow) is shown within the lower inner quadrant of the right breast. No suspicious microcalcification is observed. Hematoxylin and eosin staining shows cribriform and tubular growth (Figure 3c, ×500). Immunohistochemical staining shows CD117 (Figure 3d, ×200) and Ck5/6 (Figure 3e, ×200) positivity.

#### TECHNIQUE:

Figure 3a Ultrasound using the DC-8 (Mindray, China) with a 12L-3 linear transducer. Figure 3b Mammography using the Senographe DS (GE Healthcare, USA) (34 kV and 22 mAs). Figure 3c Hematoxylin and eosin stained sections, (×500). Figure 3d & 3e Immunohistochemical staining, (×200).



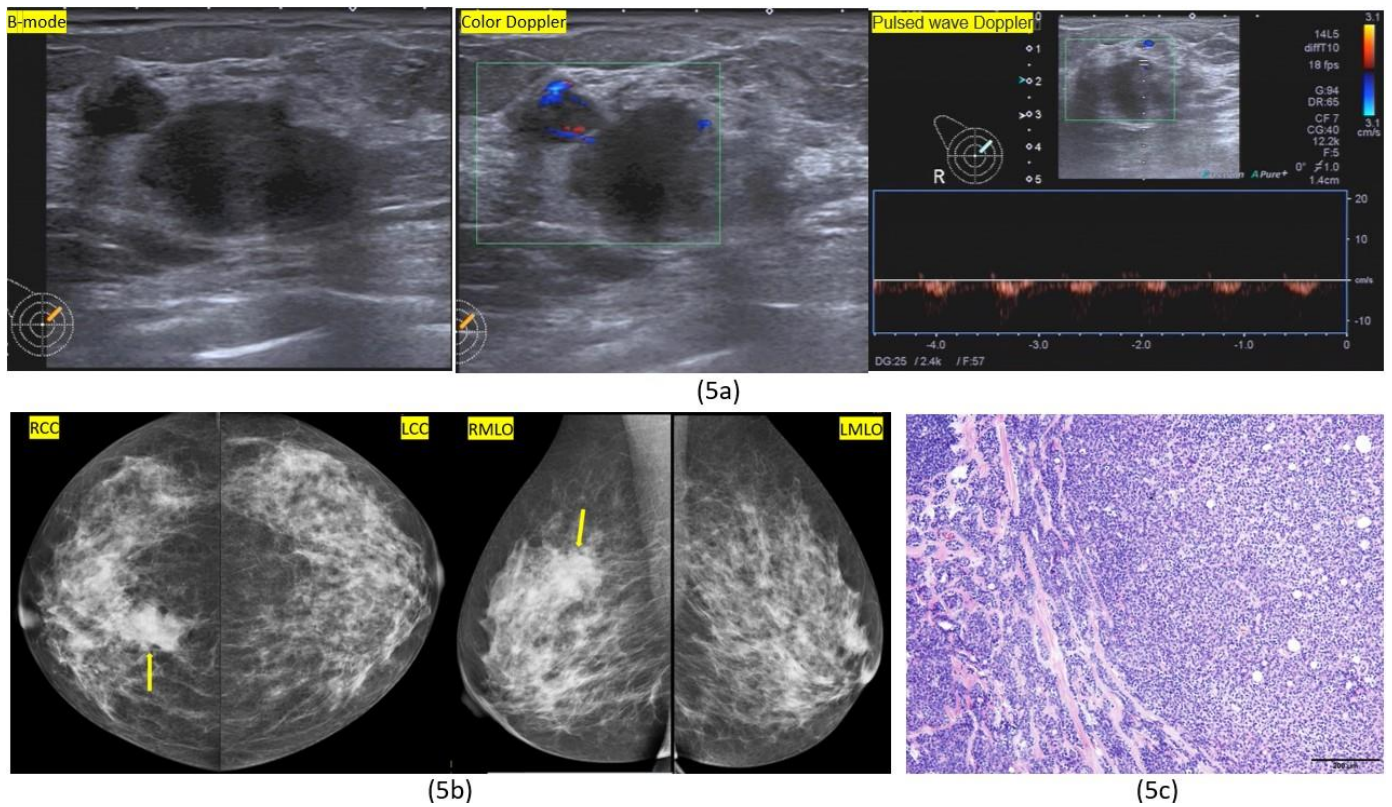
**Figure 4:** 60-year-old woman with adenoid cystic carcinoma in the left breast.

#### FINDINGS:

Figure 4a An irregular heterogeneous hypoechoic mass ( $11 \times 8$  mm) with a parallel orientation, a microlobulated margin, and posterior enhancement is shown on B-mode ultrasound. Color Doppler shows minimal vascularity in the mass. Figure 4b Mammography demonstrates bilateral scattered fibroglandular tissue. An irregular hyperintense mass ( $14 \times 11$  mm) with an indistinct margin is shown in the outer quadrant of the left breast. No suspicious microcalcification is observed.

#### TECHNIQUE:

Figure 4a Ultrasound using the ACUSON Sequoia 512 (Siemens, USA) with a 15L-8 linear transducer. Figure 4b Mammography using the Senographe DS (GE Healthcare, USA) (31 kV and 20 mAs).



**Figure 5:** 61-year-old woman with adenoid cystic carcinoma in the right breast.

#### FINDINGS:

Figure 5a Two adjacent irregular tumors with a parallel orientation and microlobulated and angular margins are shown on B-mode ultrasound. Color Doppler shows minimal vascularity and an arterial spectrum is detected on pulse wave Doppler. Figure 5b Mammography demonstrates bilateral heterogeneous dense breasts with an irregular hyperintense mass in the right breast (yellow arrow). Figure 5c Hematoxylin and eosin staining ( $\times 200$ ) shows cribriform and solid growth.

#### TECHNIQUE:

Figure 5a Ultrasound using the AplioTM 400 (Toshiba, Japan) with a 14L-5 linear transducer. Figure 5b Mammography using the Senographe DS (GE Healthcare, USA) (32 kV and 25 mAs). Figure 5c Hematoxylin and eosin stained sections ( $\times 200$ ).



Cases		Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age(year)		49	50	54	60	61
Menopausal status		Premenopausal	Premenopausal	Premenopausal	Postmenopausal	Postmenopausal
Family history		No	No	No	No	No
Symptoms		Palpable mass	Palpable mass	Palpable mass	Palpable mass	Palpable mass and breast pain
Location		Upper outer quadrant in the right breast	3 o'clock in the left breast	5 o'clock in the right breast	3-4 o'clock in the left breast	1-2 o'clock in the right breast
Ultrasound	Shape	Oval	Irregular	Irregular	Irregular	Irregular
	Orientation	Parallel	Parallel	Not parallel	Parallel	Parallel
	Margin	Circumscribed	Not circumscribed, indistinct, angular, microlobulated	Not circumscribed, angular, microlobulated	Not circumscribed, microlobulated	Not circumscribed, angular, microlobulated
	Echotexture	Heterogeneous	Heterogeneous	Hypoechoic	Heterogeneous	Heterogeneous
	Posterior features	Enhanced	Shadowing	Enhanced	Enhanced	Enhanced
Mammogram	Mammographic density	Diffuse heterogeneous dense	Scattered fibroglandular	Heterogenous dense	Scattered fibroglandular	Heterogenous dense
	Mass	Irregular dense mass	Irregular dense mass	Irregular dense mass	Irregular dense mass	Irregular dense mass
	Calcification	None	None	None	None	None
Size at gross specimen(mm)		65 × 50 × 30	33 × 25 × 23	40 × 25 × 10	20 × 10 × 10	30 × 20 × 10
TNM staging		T3N0M0	T2N0M0	T2N0M0	T1N0M0	T2N0M0
Immunohistochemistry	Subtype	Triple negative cancer	Triple negative cancer	Triple negative cancer	Triple negative cancer	Triple negative cancer
	Ki67	1%	10%	30%	15%	55%
	CD117	Negative	Positive	Positive	Not determined	Positive
	CK7	Positive	Positive	Not determined	Positive	Positive
	CK5/6	Positive	Positive	Positive	Positive	Positive
P63	Positive	Positive	Positive	Positive	Positive	
Surgery		Modified radical mastectomy and axillary lymph node dissection	Modified radical mastectomy and axillary lymph node dissection	Breast-conserving surgery and sentinel lymph node biopsy	Simple mastectomy and sentinel lymph node biopsy	Modified radical mastectomy and axillary lymph node dissection
Adjuvant treatment		No	Chemotherapy	Chemotherapy and radiotherapy	No	No
Follow-up time(month)		74	47	41	42	27
Recurrence free survival (month)		53	47	41	42	27
Prognosis		Metastasis to multiple bones and left lung.	No recurrence or metastasis	No recurrence or metastasis	No recurrence or metastasis	No recurrence or metastasis

**Table 1:** Characteristics of five patients with breast adenoid cystic carcinoma and their respective tumors.

<b>Etiology</b>	Neoplastic
<b>Incidence</b>	ACC is a rare type of tumor that which accounts for less than 1% of breast cancers.
<b>Gender ratio</b>	Predominantly female disease. Males are mentioned in a few reported cases.
<b>Age predilection</b>	Mostly in the fifth or sixth decades of life
<b>Risk factors</b>	Unclear
<b>Treatment</b>	There is no consensus regarding optimal treatment. Mastectomy has been widely accepted.
<b>Prognosis</b>	Excellent prognosis with an overall 5-year relative survival rate of 98.1%.
<b>Imaging findings</b>	Non-specific. Usually heterogeneous masses with unsharp margins and minimal vascularity. No microcalcification.

**Table 2:** Summary table for breast adenoid cystic carcinoma.

	<b>Ultrasound</b>	<b>Mammogram</b>	<b>MR</b>
<b>Adenoid cystic carcinoma</b>	A hypoechoic solid or heterogeneous mass with posterior enhancement, and minimal vascularity on color Doppler examination. Suspicious axillary lymph nodes are rarely detected.	An irregular or lobulated mass with indistinct or spiculated margins but without calcifications.	Larger lesions demonstrate an extensive heterogeneous high signal on T2WI. Delay-enhanced internal septations.
<b>Infiltrating ductal carcinoma</b>	An irregular solid mass with an uncircumscribed margin. Unchanged posterior signal is more common. Microcalcification can be seen. Abundant blood signals are shown on color Doppler. Suspicious axillary lymph nodes are commonly detected.	An irregular dense mass with spiculated margins. Microcalcification can be seen in some cases.	Variable signal intensity. Hypointense or isointense in most lesions on T2WI.
<b>Fibroadenoma</b>	A hypoechoic circumscribed oval mass with homogeneous internal echoes or coarse calcifications.	An oval or lobulated circumscribed mass sometimes with dense coarse calcifications.	Homogeneous high signal on T2WI. Non-enhancing internal septations.
<b>Phyllodes tumor</b>	An oval shaped circumscribed homogeneous hypoechoic large solid mass with a parallel orientation and fluid-filled spaces.	A large, well-defined rounded or lobulated high-density mass. Calcifications are rare.	Isointense on T1WI and heterogeneously hyperintense on T2WI with heterogeneous internal enhancement on contrast-enhanced T1WI.

**Table 3:** Differential diagnosis table for breast adenoid cystic carcinoma.

#### ABBREVIATIONS

ACC = Adenoid cystic carcinoma  
 BCS = Breast-conserving surgery  
 IDC = Invasive ductal carcinoma  
 MR = Magnetic resonance  
 SE = Spin echo  
 T1WI = T1-weighted imaging  
 T2WI = T2-weighted imaging

#### KEYWORDS

Breast Adenoid cystic carcinoma; Ultrasound; Mammography; Clinical features; Prognosis

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