

Case Report

## Adenoid Carcinoma of Breast: A Case Report

Dr Surita Kantharia<sup>1</sup>, Dr Susan Cherian<sup>2</sup>, Dr Anita Gadgil<sup>3</sup>

<sup>1</sup>Consultant, Radiology Unit - BARC Hospital, Mumbai.

<sup>2</sup>Head of Department, Pathology Unit-BARC Hospital, Mumbai

<sup>3</sup>Head of Department, Surgery Unit -BARC Hospital, Mumbai

Corresponding Author: Dr Surita Kantharia

### ABSTRACT

Adenoid cystic carcinoma (ACC) of the breast is a subtype of breast cancer characterized by the presence of a dual cell population of luminal and basaloid cells arranged in specific growth patterns. Besides, it does not express estrogen receptor  $\alpha$ , progesterone receptor, or human epidermal growth factor receptor 2 and hence also termed as a “triple negative” breast carcinoma. Though uncommon, there are various case series published, but none from India. The present case report describes a case of ACC of breast which we came across with an overview of clinical, histopathological, and molecular genetic features

**Key words:** Adenoid Cystic Carcinoma of breast, Triple negative breast carcinoma, Uncommon Breast Carcinoma

### CASE REPORT

A 52 year old postmenopausal woman presented with heaviness in the right breast and mastalgia. She was a known case of multiple simple bilateral breasts cysts and on follow-up since 3 years. On examination, a hard lump was palpable in upper and outer quadrant of right breast. She was subjected to Mammography and breast Ultrasound. Mammography (figure 1&2) revealed an irregular high density mass, approximately 6x 3.5 cm in size, with indistinct margins in upper and outer quadrant of right breast, at 10-11 o clock position, 2.5 cm away from the nipple with a skin depth of 1.3 cm. On Ultrasound (figure 3 &4), the mass was seen as heteroechoic irregular lesion, 5.7x3.5 cm in size with angular margins in outer quadrant at 10 o clock position. A core

biopsy performed, was reported as Adenoid cystic carcinoma of an intermediate grade. Immunohistochemistry revealed ER /PR and Her2neu-negative (score 0). She was subjected to Modified Radical Mastectomy. The histopathology revealed adenoid cystic carcinoma with skin, nipple and surgical margins free of malignancy. It had a single focus of invasive carcinoma with largest invasive focus of > 1mm. All the 14 nodes dissected were free of tumor. The Pathological stage (pTNM) as per, AJCC 8th edition was pT2N0. Immunohistochemistry done showed ER /PR, Her2neu, Chromogranin and Synaptophysin negative in tumor cells. CK5/6, EMA were Focal positive and CK7 diffuse positive in tumor cells and C-kit showed diffuse membranous positivity

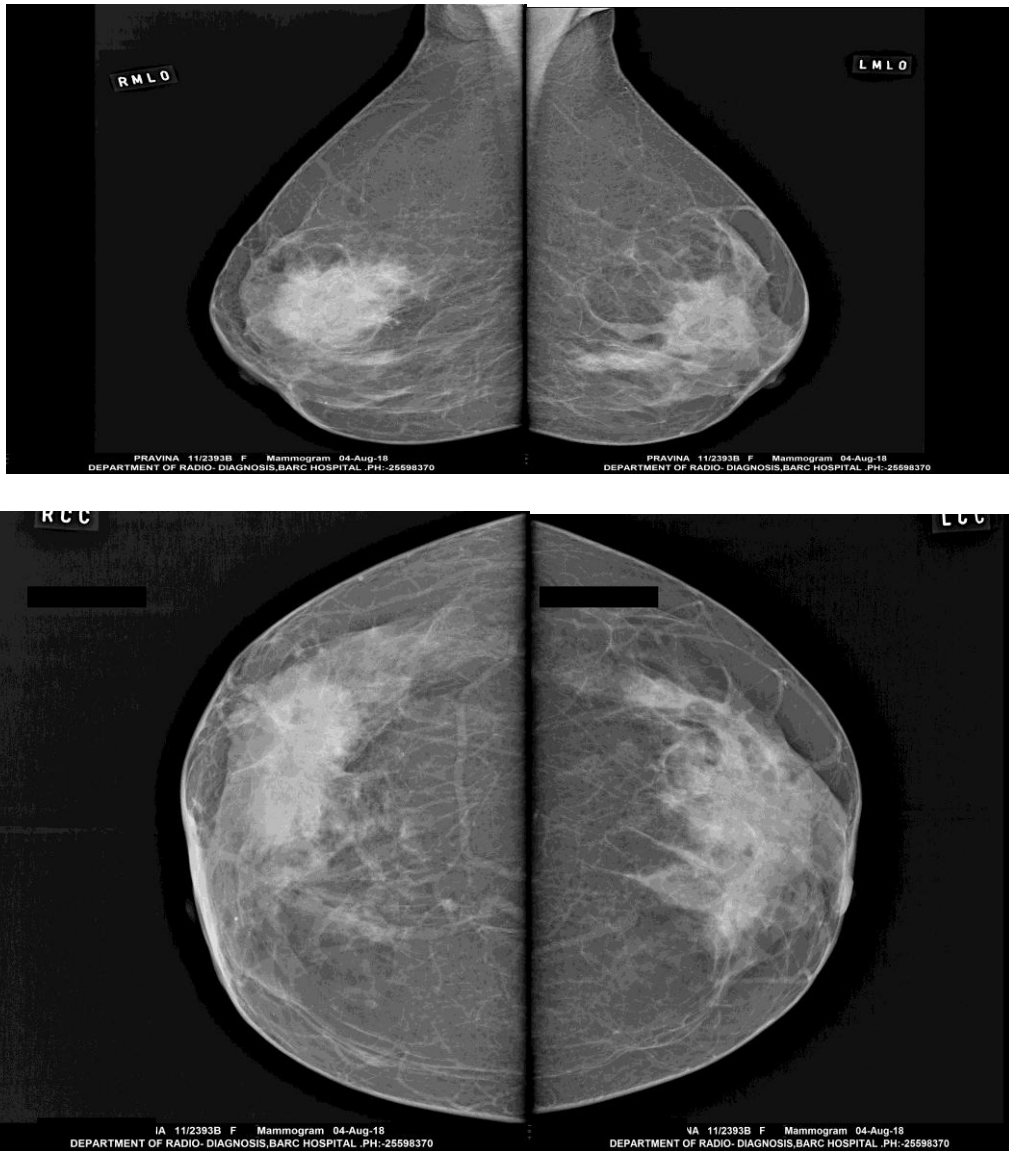


FIGURE 1&2: Mammography showing irregular high density mass, approximately 6x 3.5 cm in size, with indistinct margins in upper and outer quadrant of right breast

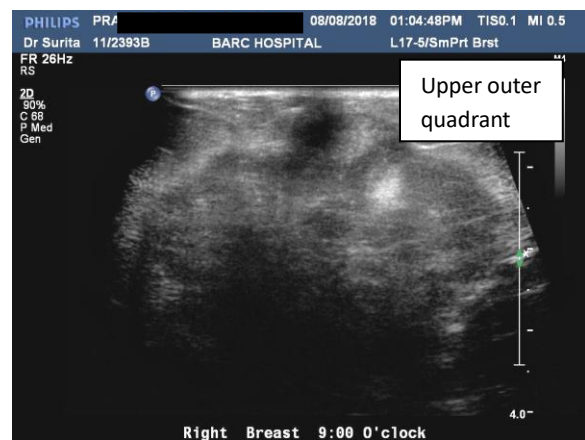


FIGURE 3&4: Ultrasonography showing heteroechoic irregular lesion, 5.7x3.5 cm in size with angular margins in upper and outer quadrant at 10 o'clock position

## DISCUSSION

Invasive breast carcinoma comprises of a heterogeneous group, with varied clinical, morphologic and molecular features. [1,2] Adenoid Carcinoma (ACC) of the breast is a rare basal-like breast cancer, [3,4] accounting for 0.1% of all breast carcinoma. [5,6] It was first described in the year 1856 by Billroth, [7] and was termed as Cylindroma, because of its morphological appearance mimicking that of a carcinoma occurring at salivary gland, lung and skin. [3,8] Geschickter was the first to describe the breast Adenoid Cystic Carcinoma in 1945. [9]

The age-adjusted incidence ratio (IR) for ACC is 0.92 per 1 million person-years as reported by Ghabach et al. [10] It predominantly affects postmenopausal women. [11] As per the Connecticut Tumor Registry the mean age of occurrence is 64 years. [12] A palpable tender mass is a common presentation. [13,14] It affects both the left and right breasts equally. Though the occurrence of tumor is not specific in any particular quadrant of breast, about 50 percent of tumors are found in a subareolar region. [15] A wide variation in tumor size has been described with the mean of tumor size being 1.8 to 3.7 cm. [11,16,17] It has been noted that the tumor may exist many years prior to its diagnosis. [18,19] The incidence of axillary LN involvement is reported to be as low as 8% [11], though the solid variant of ACC with basaloid features is associated with a higher incidence of axillary lymph node metastases. [20,21] Very rarely it may have a multifocal presentation. [22,23] It very rarely metastasizes in less than 20% of cases, the common site of metastasis being to lung and bone. [11]

The radiological findings are not very specific, and is very often diagnosed as a benign lesion. [6] On ultrasonography, the lesion is usually seen as an as well-defined, irregular, heterogeneous, or hypoechoic mass. On Mammography the lesion is seen as an asymmetric density or an irregular mass. It exhibits minimal vascularity on color Doppler imaging and on positron emission tomography scan. [24]

ACC of the breast needs to be differentiated from invasive carcinoma of the breast and, collagenous spherulosis a rare, benign condition. Besides at times, the morphologic features of the solid (basaloid) variant of ACC also overlap with that of small cell carcinoma (neuroendocrine carcinoma), solid papillary carcinoma, and metaplastic carcinoma. In such cases Immunohistochemistry is of help to distinguish among the histopathologic subtypes. [17] The characteristic histopathological features of ACC of breast is dual cell population of luminal and basaloid cells arranged in specific growth patterns and absence of estrogen receptor, progesterone receptor, and human epidermal growth factor receptor 2 expression. ACC differs from basal-like breast carcinoma at the genomic level as it less frequently harbors gains of 1q, 6p, 8q, and 10p and losses of 4p, 5q, 12q, and 15q. [25] MYB-*NFIB* gene fusion too, is a consistent genomic alteration seen in ACC and is believed to be key oncogenic mechanism in the pathogenesis of ACC. [25,26,27]

Breast -conserving surgery, such as wide excision or quadrantectomy with or without radiotherapy is the surgery of choice in ACC of breast due to its indolent clinical course and favorable outcome. [10,28,29] Mastectomy is considered in patients with invasive lesions, when the tumor has a high-grade pattern and when a cosmetically satisfactory excision is not possible. [30,31]

Study from California Registry has shown that adjuvant radiotherapy has a positive impact both on the overall survival and disease free survival. [32] However the role of adjuvant chemotherapy remains controversial and is usually reserved for high-risk early-stage patients and those with metastatic disease. [30]

The overall outcome following treatment is reported to be good with 10 year survival being more than 90 % . [10,28,33] However, patients of ACC require a long term follow up, as they have a risk of developing secondary malignancies [14,33]

and also the risk of distant metastasis increases with time.

## CONCLUSION

ACC is a subgroup of low-grade tumor with an indolent clinical behavior displaying a triple-negative, basal-like phenotype. Immunohistochemistry and genomic molecular studies are of help in distinguishing ACC of breast from the Invasive carcinoma of breast. Breast Conservative surgery with adjuvant radiotherapy is the treatment of choice, with a good overall outcome. It warrants long term follow-up, to detect secondary malignancy and secondary metastasis.

## REFERENCES

1. Schnitt S.J. and Collins L.C.: Biopsy interpretation of the breast. Philadelphia: Lippincott Williams & Wilkins, 2009.
2. Badve S., Dabbs D.J., Schnitt S.J., et al: Basal-like and triple-negative breast cancers: a critical review with an emphasis on the implications for pathologists and oncologists. *Mod Pathol* 2011; 24: pp. 157-167.
3. Marchio C, Weigelt B, Reis-Filho JS: Adenoid cystic carcinomas of the breast and salivary glands (or 'The strange case of Dr Jekyll and Mr Hyde' of exocrine gland carcinomas). *J Clin Pathol*. 2010, 63: 220-228.
4. Weigelt B, Horlings HM, Kreike B, Hayes MM, Hauptmann M, Wessels LF, de Jong D, Van de Vijver MJ, Van't Veer LJ, Peterse JL: Refinement of breast cancer classification by molecular characterization of histological special types. *J Pathol*. 2008, 216: 141-150.
5. Rosen P.P.: Adenoid cystic carcinoma of the breast. A morphologically heterogeneous neoplasm. *Pathol Annu* 1989; 24: pp. 237-254
6. Glazebrook K.N., Reynolds C., Smith R.L., Gimenez E.I., and Boughey J.C.: Adenoid cystic carcinoma of the breast. *AJR Am J Roentgenol* 2010; 194: pp. 1391-1396.
7. Billroth T: Die cylindergeschwalst. Untersuchungen ueber die entwicklung der blutgefasse. 1856, Berlin: G. Reimer.
8. Bennett AK, Mills SE, Wick MR: Salivary-type neoplasms of the breast and lung. *Semin Diagn Pathol*. 2003, 20: 279-304.
9. Geschickter CF: Diseases of the Breast: Diagnosis, Pathology, and Treatment. 1945, Philadelphia, PA: J.B. Lippincott.
10. Ghabach B., Anderson W.F., Curtis R.E., Huycke M.M., Lavigne J.A., and Dore G.M.: Adenoid cystic carcinoma of the breast in the United States (1977 to 2006): a population-based cohort study. *Breast Cancer Res* 2010; 12: pp. R54
11. Semir Vranic MD, Richard Bender MD, Juan Palazzo MD and Zoran Gatalica. A review of adenoid cystic carcinoma of the breast with emphasis on its molecular and genetic characteristics. *Human Pathology*, 2013-03-01, Volume 44, Issue 3, Pages 301-309.
12. Sumpio BE, Jennings TA, Merino MJ, Sullivan PD: Adenoid cystic carcinoma of the breast. Data from the Connecticut Tumor Registry and a review of the literature. *Ann Surg*. 1987, 205: 295-301.
13. McClenathan JH, de la Roza G: Adenoid cystic breast cancer. *Am J Surg*. 2002, 183: 646-649.
14. Millar BA, Kerba M, Youngson B, Lockwood GA, Liu FF: The potential role of breast conservation surgery and adjuvant breast radiation for adenoid cystic carcinoma of the breast. *Breast Cancer Res Treat*. 2004, 87: 225-232.
15. Azzopardi JG. Problems in breast pathology. London: Saunders Company; 1979.
16. Azoulay S, Lae M, Freneaux P, Merle S, Al Ghuzlan A, Chnecker C, Rosty C, Klijanienko J, Sigal-Zafrani B, Salmon R, Fourquet A, Sastre-Garau X, Vincent-Salomon A: KIT is highly expressed in adenoid cystic carcinoma of the breast, a basal-like carcinoma associated with a favorable outcome. *Mod Pathol*. 2005, 18: 1623-1631
17. Shin SJ, Rosen PP: Solid variant of mammary adenoid cystic carcinoma with basaloid features: a study of nine cases. *Am J Surg Pathol*. 2002, 26: 413-420.
18. Horner MJ, Ries LAG, Krapcho M, Neyman N, Aminou R, Howlader N, Altekruse SF, Feuer EJ, Huang L, Mariotto A, Miller BA, Lewis DR, Eisner MP, Stinchcomb DG, Edwards BK, (Eds): SEER

- Cancer Statistics Review, 1975-2006. 2009, Bethesda, MD: National Cancer Institute
19. Alis H, Yigitbas H, Kapan S, Kalayci M, Kilic G, Aygun E: Multifocal adenoid cystic carcinoma of the breast: an unusual presentation. *Can J Surg.* 2008, 51: E36-E37.
  20. Edge S.B., and Byrd D.R.: *AJCC cancer staging manual.* Berlin: Springer, 2010.
  21. Fukuoka K., Hirokawa M., Shimizu M., et al: Basaloid type adenoid cystic carcinoma of the breast. *APMIS* 1999; 107: pp. 762-766.
  22. Alis H., Yigitbas H., Kapan S., Kalayci M., Kilic G., and Aygun E.: Multifocal adenoid cystic carcinoma of the breast: an unusual presentation. *Can J Surg* 2008; 51: pp. E36-37
  23. Franceschini G., Terribile D., Scafetta I., et al: Conservative treatment of a rare case of multifocal adenoid cystic carcinoma of the breast: case report and literature review. *Med Sci Monit* 2010; 16: pp. CS33-9
  24. Sheen-Chen S.M., Eng H.L., Chen W.J., Cheng Y.F., and Ko S.F.: Adenoid cystic carcinoma of the breast: truly uncommon or easily overlooked? *Anticancer Res* 2005; 25: pp. 455-458
  25. Wetterskog D., Lopez-Garcia M.A., Lambros M.B., et al: Breast adenoid cystic carcinomas constitute a genomically distinct subgroup of triple-negative and basal-like breast cancers. *J Pathol* 2012; 226: pp. 84-96.
  26. Brill L.B., Kanner W.A., Fehr A., et al: Analysis of MYB expression and MYB-NFIB gene fusions in adenoid cystic carcinoma and other salivary neoplasms. *Mod Pathol* 2011; 24: pp. 1169-1176
  27. Persson M., Andrén Y., Mark J., Horlings H.M., Persson F., and Stenman G.: Recurrent fusion of MYB and NFIB transcription factor genes in carcinomas of the breast and head and neck. *Proc Natl Acad Sci USA* 2009; 106: pp. 18740-18744
  28. Khanfir K, Kallel A, Villette S, Belkacémi Y, Vautravers C, Nguyen T, Miller R, Li YX, Taghian AG, Boersma L, et al. Management of adenoid cystic carcinoma of the breast: a Rare Cancer Network study. *Int J Radiat Oncol Biol Phys.* 2012;82:2118–2124.
  29. Xue Y, Liu X, Poplack S, Memoli VA. Adenoid cystic carcinoma of the breast in reduction mammoplasty. *Breast J.* 2012;18:611–613.
  30. Rosen PP. Adenoid cystic carcinoma. In: Rosen PP, editor. *Rosen's breast pathology*, third edition. Philadelphia: Lippincott Williams & Wilkins; 2009. pp. 590–604.
  31. Youk JH, Kim MJ, Kim EK, Lee JY, Oh KK, Park BW. Recurrence of adenoid cystic carcinoma in the breast after lumpectomy and adjuvant therapy. *J Ultrasound Med.* 2006;25:921–924.
  32. Coates J.M., Martinez S.R., Bold R.J., and Chen S.L.: Adjuvant radiation therapy is associated with improved survival for adenoid cystic carcinoma of the breast. *J Surg Oncol* 2010; 102: pp. 342-347.
  33. Page D.L. Adenoid cystic carcinoma of breast, a special histopathologic type with excellent prognosis. *Breast Cancer Res Treat* 2005; 93: pp. 189-190.
  34. Sinn H.P., Lehnert T., and Otto H.F.: Adenoid cystic cancer of the breast. Case report and meta-analysis of the literature. *Chirurg* 1993; 64: pp. 198-202.
  35. Vranic S., Frkovic-Grazio S., Lamovec J., et al: Adenoid cystic carcinomas of the breast have low Topo II $\alpha$  expression but frequently overexpress EGFR protein without EGFR gene amplification. *Hum Pathol* 2010; 41: pp. 1617-16

How to cite this article: Kantharia S, Cherian S, Gadgil A. Adenoid carcinoma of breast: a case report. *Int J Health Sci Res.* 2019; 9(6):388-392.

\*\*\*\*\*