

Adenoid Cystic Carcinoma of the Breast, A Case Report and Review of the Literature

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Abstract

Case Report

Adenoid cystic carcinoma (ACC) or mammary cylindroma is a rare type of breast cancer. We report the case of a 52-year-old patient who consulted us for breast swelling. Examination revealed a well-defined nodule, and mammography showed a stellate opacity without microcalcification. The final histological diagnosis was established on the tumorectomy specimen with immunohistochemistry. The patient was treated with conservative therapy associated with adjuvant radiotherapy, achieving complete remission after 19 months.

Keywords: Adenoid cystic carcinoma; Breast; tumorectomy; Radiotherapy.

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INTRODUCTION

Adenoid cystic carcinoma is a rare malignant tumor of the breast, mainly affecting the salivary glands and upper airways. It is a cancer with a good prognosis, and breast localization is less than 1%.

We report a new observation of adenoid cystic carcinoma and study its anatomo-clinical and therapeutic features. No axillary or supraclavicular lymph nodes were found, as was the case for the left breast. Mammography revealed a stellate opacity straddling the right upper quadrants, with no microcalcifications. Complementary ultrasonography revealed a heterogeneous hypoechoic mass with irregular contours and a cauliflower-like appearance measuring 45 mm, classified as ACR 5. Histological examination of the biopsied fragments revealed a high-grade in situ carcinoma.

OBSERVATION

A 52-year-old female patient, nulliparous, with no personal or family history of cancer, consulted for a right breast mass discovered more than a year previously. Clinical examination revealed a patient in good general condition, with a 5 cm mobile mass in the upper quadrants of the right breast, and no signs of skin inflammation or nipple discharge. No axillary or supraclavicular lymph nodes were found, as was the case

for the left breast. Mammography revealed a stellate opacity straddling the right upper quadrants, with no microcalcifications. Complementary ultrasonography showed a heterogeneous hypoechoic mass with irregular contours and a cauliflower-like appearance, measuring 45 mm and classified as ACR 5. Histological examination of the biopsied fragments revealed a high-grade in situ carcinoma.

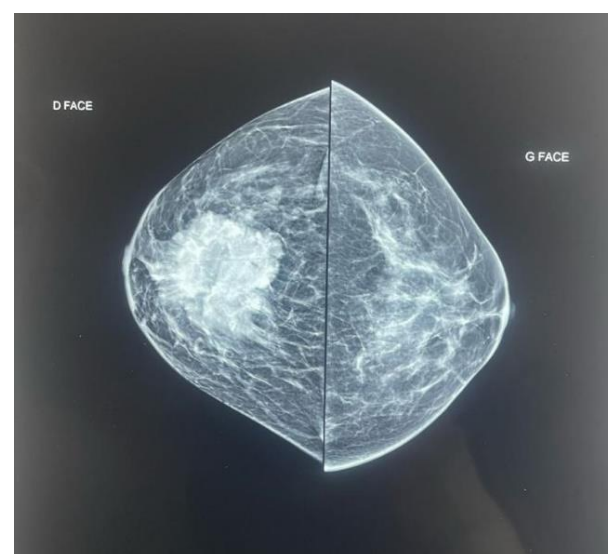


Figure 1: Craniocaudal View of a Mammography Showing the Opacity of the QS Junction

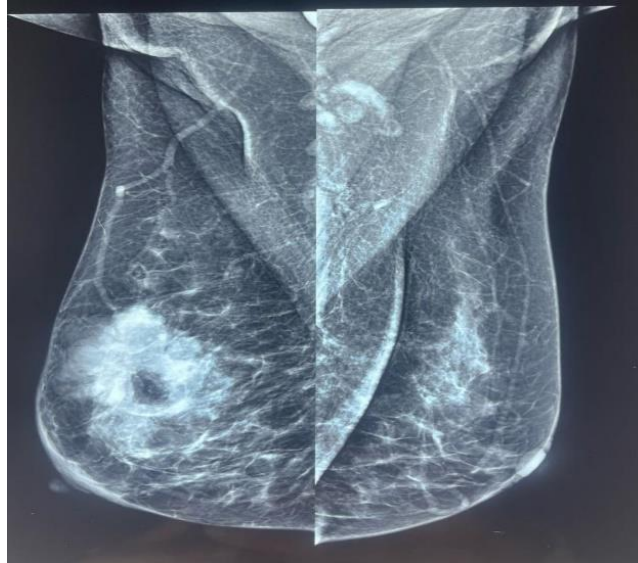


Figure 2: Medio-Lateral Oblique View

A large lumpectomy was performed, and macroscopic examination of the specimen revealed a neoplasm measuring 6 x 6 x 3 cm, with clear resection margins. Microscopically, we found a dual epithelial and myoepithelial proliferation. On immunohistochemistry, there was strong expression of anti-P63, anti-PS100, and anti-CD117 antibodies, with no expression of hormone receptors or HER2.

Thoracic, abdominal, and bone scans were normal. After a multidisciplinary meeting, a

complementary right axillary lymphadenectomy was performed, revealing 8 reactive lymph nodes free of tumor proliferation. The patient received adjuvant radiotherapy to the right breast, totaling 50 Gy in 25 fractions over 5 weeks.

The patient was monitored every 6 months with clinical examinations and annual imaging studies. Nineteen months after the initial treatment, no recurrence was reported.



Figure 3: 1month Postoperative Result

DISCUSSION

Adenoid cystic carcinoma (ACC) of the breast is an extremely rare malignancy, accounting for less than 0.1% of all breast carcinomas [1]. Primarily affecting women between the ages of 60 and 70, it is characterized by low malignant potential and a generally favorable prognosis [2].

The usual clinical appearance of this tumor is that of a firm, mobile, well-defined central mammary mass, which may be confused with an adenofibroma [3]. The mammographic aspects are not specific and are identical to those of other carcinomas; however, a

suggestive element is the absence of microcalcifications, which was the case in our patient's situation [4].

The histological appearance is similar to the cystic adenoid carcinomas seen in other locations (salivary glands, respiratory airways), consisting of two cellular components with a cribriform architecture. The hormonal profile is generally negative [5].

Mastectomy with sentinel node study has long been the treatment of choice for ACC, and the promising 5-year survival results and low local recurrence rate encourage this approach [6]. However, the emergence of

external beam radiotherapy has changed the approach towards a more conservative management.

The KROG study (1992-2022) reported that of 93 patients with ACC, 75 were treated with conservative therapy (CT) followed by breast irradiation, while 18 were treated with radical therapy (RT), including 2 who received axillary radiotherapy for pN+. The CT group (n=75) showed a regional recurrence rate of 1.3%, while the RT group (n=18) showed a rate of 5.6%. These results, together with several observations of isolated cases treated with CT and irradiation, increasingly support a conservative approach [7-8].

Adjuvant radiotherapy in the management of ACC is not clearly defined. Despite these broad indications, it reduces the rate of local recurrence after conservative treatment [9].

Chemotherapy is generally considered ineffective for ACC due to its low proliferation index and distinct histological features, which contribute to its resistance to conventional chemotherapeutic agents. It has also shown limited success in reducing tumor size in neoadjuvant therapy [9], and is therefore not routinely recommended for ACC, except in cases of advanced or metastatic disease [10].

The hormonal status of the tumor is almost always negative; the potential role of hormone therapy in the treatment of these patients is unclear and requires further investigation [9].

CONCLUSION

Adenoid cystic carcinoma of the breast is a rare histological entity. It mainly affects menopausal women. Clinical symptoms and radiological appearance are not specific. The histological appearance is characteristic, but may lead to confusion with other breast cancers, making immunohistochemical techniques particularly useful. Treatment is still poorly established, and codification requires patients to be included in multicenter randomized therapeutic trials. This tumor has a very favorable prognosis; it may recur locally or distantly, justifying regular long-term follow-up.

BIBLIOGRAPHIE

1. Ray JW, Louis M, Grabill N, Ruiz JM, Strom P. A rare case of recurrent adenoid cystic carcinoma of the breast with weak hormone receptor positivity. *Radiol Case Rep.* 2024 Dec 30;20(3):1595-1600. doi: 10.1016/j.radcr.2024.12.011. PMID: 39835301; PMCID: PMC11745780.
2. Muslimani AA, Ahluwalia MS, Clark CT, Daw HA. Primary adenoid cystic carcinoma of the breast: case report and review of the literature. *Int Semin Surg Oncol* 2006;3:17. doi:10.1186/1477-7800-3-17
3. Y. Sbitti a, H. Kadiri a, b, c, d, M. Fetohi a, b, I. Elghissassi a, H. M'rabti a, S. Boutayeb a, M. Ichou b, H. Boussen c, H. Errihani Carcinome adénoïde kystique primitif du sein - 21/01/09 , Doi : 10.1016/j.canrad.2008.10.001
4. De Luis E, Apesteguía L, Noguera JJ, Pina L, Martínez-Regueira F, Miguel C, Sáenz J. Carcinoma adenoide quístico de mama [Adenoid cystic carcinoma of the breast]. *Radiologia.* 2006 Jul-Aug;48(4):235-40. Spanish. doi: 10.1016/s0033-8338(06)73160-5. PMID: 17058651.
5. Okamoto Y, Sumiyama Y, Arima Y, Sakuta M, Okuda T, Noto Y, Naka I, Kiribayashi T, Takahashi K, Kuwajima A. A case of adenoid cystic carcinoma (ACC) of the breast and review of the utility of preoperative imaging diagnose. *Breast Cancer.* 2001;8(1):84-9. doi: 10.1007/BF02967484. PMID: 11180772.
6. Ray JW, Louis M, Grabill N, Ruiz JM, Strom P. A rare case of recurrent adenoid cystic carcinoma of the breast with weak hormone receptor positivity. *Radiol Case Rep.* 2024 Dec 30;20(3):1595-1600. doi: 10.1016/j.radcr.2024.12.011. PMID: 39835301; PMCID: PMC11745780.
7. Lee SM, Jang BS, Park W, Kim YB, Song JH, Kim JH, Kim TH, Kim IA, Lee JH, Ahn SJ, Kim K, Chang AR, Kwon J, Park HJ, Shin KH. Locoregional Recurrence in Adenoid Cystic Carcinoma of the Breast: A Retrospective, Multicenter Study (KROG 22-14). *Cancer Res Treat.* 2025 Jan;57(1):150-158. doi: 10.4143/crt.2024.201. Epub 2024 Jul 12. PMID: 38993091; PMCID: PMC11729329.
8. Romeira D, Cardoso D, Miranda H, Martins A. Adenoid cystic carcinoma: triple negative breast cancer with good prognosis. *BMJ Case Rep* 2016;2016. doi:10.1136/bcr-2015-213704.
9. Arpino, G., Clark, G.M., Mohsin, S., Bardou, V.J. and Elledge, R.M. (2002), Adenoid cystic carcinoma of the breast. *Cancer*, 94: 2119-2127. <https://doi.org/10.1002/cncr.10455>
10. Alshwayyat S, Abu Al Hawa MB, Alshwayyat M, Alshwayyat TA, Sawan S, Heilat G, Hammouri HM, Mheid S, Al Shweiat B, Hanifa H. Personalized treatment strategies for breast adenoid cystic carcinoma: A machine learning approach. *Breast.* 2025 Feb;79:103878. doi: 10.1016/j.breast.2025.103878. Epub 2025 Jan 13. PMID: 39826386; PMCID: PMC11786111.