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Breast adenomyoepithelioma, a case report

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ABSTRACT

BACKGROUND: Adenomyoepithelioma is a rare tumor of the breast characterized by a dual/biphasic proliferation of two cell populations: the epithelial cells and the myoepithelial cells. The first case was reported in 1970. The majority of the cases are benign, but few malignant cases were reported in literature.

CASE PRESENTATION: A case of a 66-year-old lady presenting with an asymptomatic breast mass, of 18 × 17 × 15 mm size with irregular borders and negative metastatic workup. The patient was operated for wide local excision of the tumor, with a confirmed negative margins intraoperatively. The final pathology was Adenomyoepithelioma.

CONCLUSION: We report this rare case to encourage physicians to keep this etiology in mind as part of the differential diagnosis of breast mass.

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1. Introduction

Adenomyoepithelioma of the breast is a rare tumor that was first described by Hamperl in 1970 [1]. The second case was reported several years later, in 1983 [2]. These tumors have a variable biological behavior. The majority of the cases reported were benign, but the tumor tends to recur locally. Malignancy was reported in up to 40 cases [3]. Imaging studies are not conclusive and cannot build definite diagnosis. Local excision is the best treatment and histopathology is the gold standard for diagnosis [3].

Here, we report a case of breast adenomyoepithelioma, in an elderly female patient, then we proceed to review of literature concerning the presentation, pathology, and optimal treatment.

This case was reported in accordance with the SCARE criteria [4].

Abbreviations: AME, Adenomyoepithelioma; CT, Computed Tomography; ER, Estrogen Receptor; FNA, Fine Needle Aspiration; MRI, Magnetic Resonance Imaging; PR, Progesteron Receptor; SLND, sentinel lymph node dissection.

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2. Case history

We report a case of a 66-year-old female patient, with an unremarkable medical or family history, operated with cholecystectomy and hysterectomy. The patient presented with a right breast mass, that was discovered by self-examination. She denied any nipple retraction or discharge, breast pain, redness or swelling.

The patient had a good general condition. She denies any systemic complaint. The physical examination was unremarkable except for a palpable right breast mass in the lower inner quadrant. There is no palpable axillary masses.

Mammography showed an oval opacity in the lower inner quadrant, with hazy margins, containing scattered microcalcifications. Ultrasound described the mammography findings as a heterogeneous soft tissue mass, measuring 18 × 17 × 15 mm, with irregular borders, with no obvious vascularization. no axillary lymph nodes visible. Imaging of the left breast was unremarkable (Fig. 1). A breast core biopsy was done and showed a fibroadenoma changes.

In the metastatic workup, a CT scan of the chest, abdomen and pelvis was done, with injection of intravenous contrast showing no significant changes (Figs. 2 and 3).

She was planned for nipple sparing right partial mastectomy, and the mass was sent for frozen section, and was reported as low-grade carcinoma, reaching the surgical margins. Wide local excision of additional tissue was done and negative surgical margins were achieved. The procedure was completed with a right axillary lymph

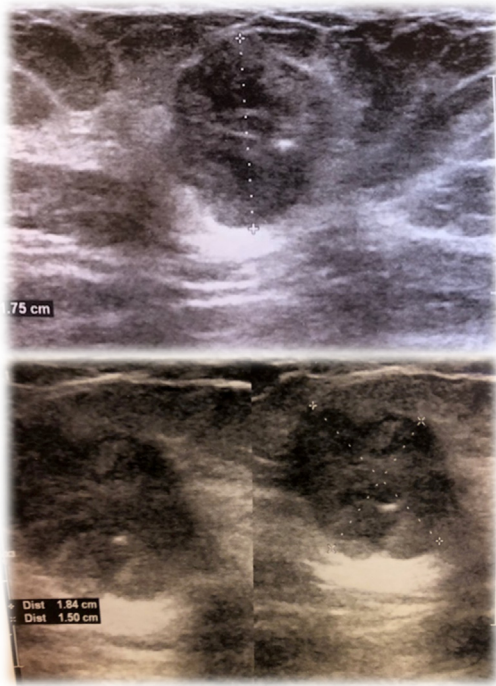


Fig. 1. An ultrasound image of the right breast showing a heterogeneous soft tissue mass, measuring 18 × 17 × 15 mm, with irregular borders, and no obvious vascularization.

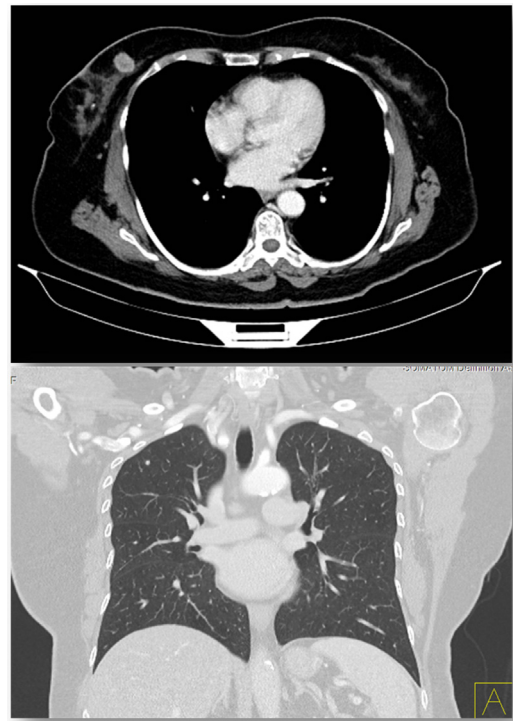


Fig. 2. A chest CT scan showing a right breast nodule, with small sub-centimetric para-tracheal nodule, and multiple micronodules, largest being 4 mm in the right upper lung field that appear to be inflammatory in origin.

node dissection because the methylene blue dye or technetium were not injected preoperatively due to low suspicion in malignancy.

The patient had a smooth post op course and was discharged home on post-operative day 4, after removal of the surgical drains, in a stable medical condition.

The final pathology reported an Adenomyoepithelioma. Microscopically, the tumor composed of multiple nodules, surrounding a sclerotic core. Proliferating small acinar structures are seen, associated in areas to myoepithelial cells, forming a trabecular pattern, and small nests. A discontinuous myoepithelial layer was identified, and nuclei are mildly enlarged with weak mitotic activ-

ity. (Ki67 mitotic index <5%). The intervening stroma is fibrotic and collagenized in central areas, entrapping small proliferating acini. Immuno-histochemical stains P63, ACTIN, and Ki67). Axillary lymph nodes were negative for malignant cells. The patient is to be followed up in 6 months.

3. Discussion

Breast adenomyoepithelioma (AME) is a rare breast tumor, with the first case reported in 1970 by Hamperl [1]. Several years later, in 1983, the second case of AME was reported by Zarbo [2]. A number

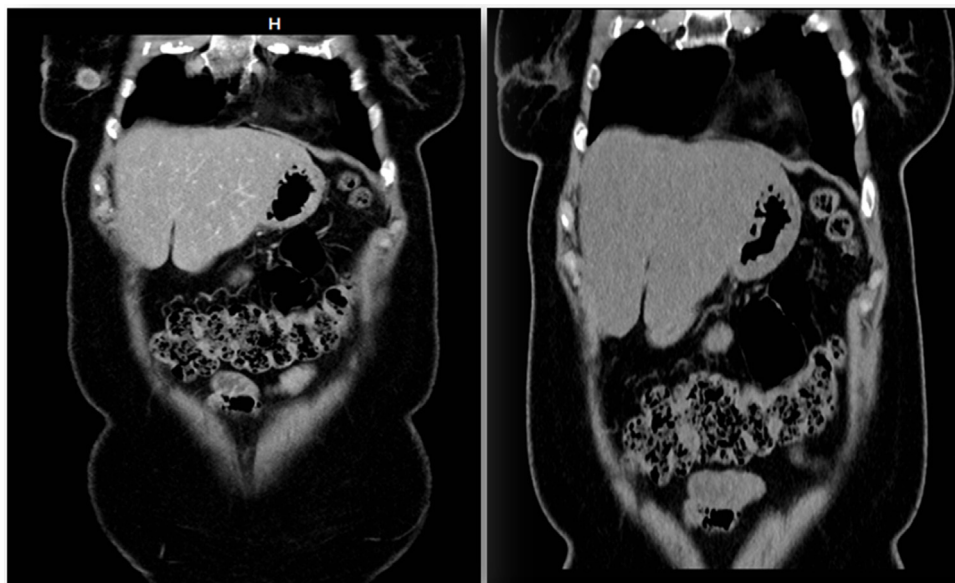


Fig. 3. Abdominal CT scan showing no hepatic focal lesions, yet an enlarged liver. No visible pelvic masses, absent uterus.

of case reports has been published after that. By 2013, only 150 cases have been reported [3].

This tumor has a female predominance; and only two male cases were reported so far [5,6]. The majority of the patients reported were middle aged and elderly over 40 years of age. However, the age distribution is wide, with the youngest benign case being 16 years of age [7], and the oldest being 86 years of age [8].

The majority of the patients present with a symptomatic breast mass; however, few patients are asymptomatic and have their masses detected by screening studies or by autopalpation. Other patients have presented with breast mass or masses, and was first diagnosed with benign lesions, before seeking advice again, as in our patient. (Probably due to misdiagnosis or sampling error).

These lesions are unilateral in the majority of cases, however, a case of bilateral adenomyoepithelioma was reported in a young female patient, 16 years of age, by Bajpai et al. in the journal of cancer research and therapeutics in 2013 [7], so far being the only case of tumor bilaterality reported.

These lesions tend to present as a single mass; however, Suzanne et al. reported a case of two adjacent breast masses that were detected by screening mammography, and were both diagnosed as adenomyoepithelioma [9]. Imaging techniques are non conclusive; mammographic features and MRI are non specific and rarely show microcalcification. Ultrasound features show solid oval hypoechoic mass with irregular borders [3].

It was reported that these tumors tend to occur centrally within the breast, more precisely in the areola area. This can be explained by the fact that myoepithelial cells are located between the ducts and the basal layers of the acinus, and the major breast ducts aggregate in the areola area within abundant myoepithelial cells [10].

Breast AME are uncommon neoplasms, and are composed of two population of cells: luminal epithelial cells and myoepithelial cells. Pathologically, they are characterized by a biphasic proliferation of both cell populations. There is a spectrum of histological patterns depending on the distribution of proliferating glandular cells, prominence of the papillary and fibrotic components, and the morphology of the myoepithelial cells [9]; thus, defining the tumor as papillary, lobulated, tubular, or with mixed architectural pattern [11]. Biologically, the majority of the tumors are benign, however, either the epithelial or the myoepithelial component or both, might undergo malignant transformation. AME can be associated with other breast lesions, including collagenous spherulosis, low-grade adenosquamous carcinoma, or adenoid carcinoma [12].

Breast AME are classified into three categories: (a) benign, (b) malignant, (c) atypical. In a study conducted by Erza et al. the tumor is reported as “AME with malignant transformation”, or “malignant AME” if the tumor exhibited overgrowth of either component, severe atypia, infiltrative growth pattern, increased mitotic activity, and/or necrosis. Tumors that do not possess all features were classified as “Atypical AME” [11].

So far, 40 cases of malignancy were described in literature. The metastatic potential is not well studied, but it appears to be a hematogenous spread rather than a lymphatic spread, and occurs with tumors of greater than 2 cm in size [1,15]. Distant metastasis is extremely rare, and some authors described metastasis to the thyroid gland, lungs, liver and brain [3]. Maffini et al. described a rare case of metastasis to the lung, from malignant AME of the breast in a 40 year-old lady, 1 year after being operated for her malignancy [13].

Given the fact that these lesions are pathologically heterogeneous, limited tissue sampling can sometimes lead to misdiagnosis, as possibly in our case whose initial FNA results were reported as Fibroadenoma. Because the tumor is hypercellular, diagnosis by cytology is difficult [16], and its best achieved by a core biopsy, providing a more accurate diagnosis. If cytology was chosen, and a

benign lesion was diagnosed, a total excision of the lesion with safe surgical margins is recommended [14].

Concerning the surgical treatment, both benign and malignant tumors should be removed. Once the cytopathological diagnosis is clear, some authors have suggested simple mastectomy with negative margins, and given the potential of local recurrence, or even malignant transformation, an extended resection or breast conserving surgery, with surgically negative margins is also an option [10]. Local recurrences are managed with re-excision, and several re-excisions were reported in literature. Radiotherapy was used in few cases with local recurrence.

With respect to the spread to the regional lymph nodes distant metastasis, as mentioned earlier, several cases including lung and brain metastasis were reported, in the setting of malignant AME. Peri et al. studied five cases of malignant AME, all of which received axillary dissection, but the axilla was reported as negative for malignancy [17]. The tumor has the potential for local recurrence, even after the index operation, but lymph nodes can be excluded from the disease. Given these data, and because there is no definite conclusion in literature concerning the management of the axilla, some authors think that it is preferable to combine sentinel lymph node dissection (SLND), with simple mastectomy, and act accordingly [10].

Is there a role for adjuvant systemic treatment in malignant AME?

With respect to malignant AME, the hematogenous spread is the more common means of spread, and several cases of hematogenous metastasis were reported including lung, brain, bone, and even thyroid metastasis [18,19]. As reported by Bult et al. one third of the patients with malignant AME tend to have local recurrence or distant spread within 4 months to 2–3 years after the diagnosis. Because of this potential, the scholars recommended adjuvant chemotherapy for malignant AME cases. Concerning the endocrine therapy, it somehow has no role in malignant AME, as these tumors tend to be ER and PR negative, unless associated with or combined with a receptor positive infiltrating carcinoma [20].

In conclusion, adenomyoepithelioma is a rare entity, which both pathologists, surgeons, and clinicians need to be aware of as part of the differential for symptomatic and asymptomatic breast masses. The variable spectrum of behavior ranging from benign to malignant, with tendency to local recurrence, makes that tumor interesting, and should be kept in mind while diagnosing any breast lesion.

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This article was exempt from ethical approval.

Availability of data and materials

Not applicable.

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This article has no conflict of interest with any parties.

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Ethical approval

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Consent

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A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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