Early stage malignant phyllodes tumor case report

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1. Introduction

Currently, breast cancer is the most common type of cancer in women in the United States besides skin cancers [11] and second leading cause of death in women. Among all types of breast cancers, phyllodes tumor of the breast is a rare type of tumor comprising approximately 0.5–1% of all breast cancers [7,15]. Although rare, phyllodes tumor is usually a benign non-cancerous fibroepithelial neoplasm that typically occurs in the third or fourth decades and usually does not involve lymph nodes. Malignant phyllodes tumor is an extremely rare malignant lesion that makes up about 5–10% of all phyllodes tumors.

As the name suggests, malignant phyllodes is an aggressive tumor that spreads hematogenously. Although aggressive, the clinical and radiographic presentation usually mimics that of benign lesions rendering it difficult to diagnose since it is very challenging and hard to distinguish by clinical features alone. Biopsy and histopathologic evaluations are required for definitive diagnosis and when confirmed, surgical resection with or without adjuvant chemotherapy and radiation therapy is the mainstay of treatment. Most common surgical management options are total mastectomy with or without lymph node dissection depending on lymph node involvement. Due to the scarcity of malignant phyllodes tumor cases, there is a paucity of literature and evidence for surgical management options.

In this case report, we discuss a case of a malignant phyllodes tumor without any metastasis that was successfully treated by lumpectomy.

2. Case report

This is a case of a 46-year-old female with family history of non-phyllodes breast cancer and no significantly contributing personal medical or surgical history. The patient’s mother was diagnosed with kidney cancer at age 61, her two sisters and her maternal aunt deceased secondary to breast cancer that was diagnosed at young ages. There was no genetic testing performed in the family. Besides depression for which the patient took Cymbalta, the patient had no other significant medical condition and did not take any illicit drugs. In December 2016, the patient felt a lump in her left breast a month prior to the presentation without any changes in the breast appearance or any breast discomfort. During this visit, physical examination revealed a small lump on the lateral quadrant at 3 O’clock of the left breast but failed to demonstrate any discharges, skin dimpling, discoloration, or abnormal disfiguration.

The patient has had routine mammogram screenings. Five years ago, a 1.0-cm lesion in the left breast was found on the mammogram, presumed to be benign at the time, with a BI-RADS score of 3 suggesting a short interval follow-up, but did not prompt a tissue biopsy due to two percent chance of malignancy with a BI-RADS score of 3. Until presentation, annual follow-up mammograms from 2013 to 2016 consistently suggested benign lesion with a BI-RADS score of 2 without changes in size, nodularity, calcifica-
Fig. 1. CC view mammogram of the left breast showing 3.4 × 2.7 cm lesion right adjacent to dense breast tissue.
Fig. 2. A) Ultrasound of the left breast mass at 3 o’clock – Sagittal view. B) Ultrasound of the left breast mass at 3 o’clock – Transverse view.
tion, or architectural distortion. In 2017, follow-up mammogram and ultrasound were conducted which revealed this same lesion to have changed to a macrolobulated heterogeneous hypoechoic mass measuring approximately 2.9 cm with a BI-RADS score of 4 raising suspicion of malignancy and therefore, required a biopsy (Figs 1, 2). An ultrasound-guided needle biopsy of the left breast lesion was performed with satisfactory biopsy clip placement. The pathology report suggested phyllodes tumor bordering between a borderline and malignant phyllodes tumor which prompted surgical excision of the lesion (Fig. 3).

The patient was informed and given available surgical options, potential risks, and complications. After a thorough discussion of the pathology and the disease process with the patient, the decision was made to proceed with left breast lumpectomy. During the operation in February of 2017 performed by a board-certified breast surgeon, a specimen was excised and sent for histopathologic evaluation. The patient tolerated the operation well without any significant complications. The pathology evaluation showed a 7.0-cm malignant phyllodes tumor demonstrating a leaf-like architecture composed of deep epithelial-lined clefts surrounded by a pleomorphic stroma of which the cells are cytologically malignant with strikingly abnormal mitotic figures and some atypical ductal hyperplasia.

A chest CT scan was performed that showed no evidence of distant metastasis. The case was discussed with the medical and radiation oncology teams at a multidisciplinary tumor board, and the decision was made not to proceed with adjuvant chemotherapy or radiation therapy. The consensus was that there is little to no evidence supporting the role of adjuvant therapies for malignant phyllodes tumors. Although there was no lymph node involvement and the margins were clean of malignant cells, the closest distance to the margin was found to be 0.2 cm which prompted additional excision to minimize the risk of recurrence.

Re-excision lumpectomy was performed a month after the initial lumpectomy and successfully confirmed to be free of any residual malignancy. The patient happily continued to comply and attend her follow-up appointments with the oncologist. At this point, she is symptom-free and there is no evidence of residual disease confirmed by monthly serial imaging.

3. Discussion

Phyllodes tumor of the breast is a rare condition and usually presents as a benign unilateral breast lump occurring in third or fourth decades. A very small portion of already rare phyllodes tumor is classified as malignant which can grow rapidly, and be highly aggressive spreading hematogenously. Due to its rarity, the management guideline of malignant phyllodes tumor is not deeply structured despite the fact that it is a well-studied condition [19]. According to the current NCCN guidelines for breast cancer, the management of phyllodes tumors >3.0-cm is surgical excision with clean margins >1.0 cm without axillary staging whether the tumor is benign, borderline, or malignant [13]. Many other studies have supported that when the margins are less than 1.0 cm, there is a higher recurrence rate ranging from 16.7% to 40% [5,9,10,14,17,18,21]. The guideline suggests that when possible, lumpectomy is a better option over mastectomy since it preserves the integrity and the overall architecture of the breast. For lumpectomy to be a potential management option, clinicians presume an important role in diagnosing the tumor when it is still small.

The clinical presentation and the radiographic findings of malignant phyllodes are strikingly similar to that of benign lesions such as fibroadenoma or even benign phyllodes tumor; thus, making it quite challenging for the clinicians to diagnose or even to suspect the disease at an earlier stage. Although routine breast biopsy may not be warranted, it is crucial for clinicians to consider and include phyllodes tumor in their differential diagnosis. Moreover, it is evident that the clinicians also cannot completely rely on radiographic findings. Sonography cannot distinguish between malignant, borderline, and benign phyllodes tumors [6]. In this case report, for
instance, the largest diameter of the tumor size on radiographs was 3.4-cm when in fact, the actual tumor size according to pathology was 7.0-cm.

Although the features of phyllodes tumor may be difficult to distinguish from those of fibroadenomas, especially in dense breast tissues, there are several distinct radiographic features that may suggest a higher likelihood of a phyllodes tumor than a fibroadenoma. A phyllodes tumor may demonstrate higher-density mass than surrounding fibroglandular breast tissue on a mammogram. In addition, phyllodes tumor on ultrasound is more likely to appear round and lobulated with a marked posterior acoustic enhancement that is less likely to be associated with fibroadenoma [22].

Because there have not been many malignant cases of phyllodes tumor, there is not sufficient data to fully calculate the statistics of the survival rate. Past case reports and studies have suggested that the prognosis of malignant phyllodes tumor of the breast is usually poor [8,23] while overall prognosis of benign phyllodes is good. Indeed, the overall survival rate of those patients with malignant phyllodes tumor is low when the patient is diagnosed with the disease after that it had already metastasized [2,10,15] to distant sites such as lungs, bone, liver, brain, tonsil, and kidney. It is pivotal for early diagnosis and staging not only to improve the overall outcome of the disease after treatment but also to better the quality of life of the patient causing less disfigurement.

There is currently no consensus regarding radiotherapy, hormonal therapy and systemic chemotherapy recommended for malignant phyllodes tumors. To this date, there has not been any double-blinded, multicenter study on this topic. Most case reports and studies treated these tumors with only wide local excision according to the current NCCN guidelines. Barth et al. demonstrated that perioperative radiotherapy increased 10-year local control rate from 59% to 86% for both benign and malignant tumors [3,4]. Since phyllodes tumor is more considered as a soft tissue sarcoma, adjuvant chemotherapy with doxorubicin plus dacarbazine could provide some benefits to the patients with large (>5.0-cm), high-risk tumors [12,20]. A deeper look into adding adjuvant therapy for large aggressive malignant cases of phyllodes tumor may prove to be fruitful [16].

In addition to the current NCCN guideline for phyllodes tumor management, there is a need to define the terms of early diagnosis. While the breast cancer screening guideline suggests that women over 40 years of age should begin routine mammograms to detect the presence of breast cancers, phyllodes tumor can occur a decade before the minimum screening age since it occurs during the third or fourth decades of life. Moreover, when the patient suspects that the lesion is growing in size within 6 months to a year, it should be considered for further workup. We strongly urge the surgical community to be cognizant of phyllodes tumors and consider them in the differential diagnosis should the clinical suspicion is raised; if the radiographic finding shows lobulation and heterogeneous hypoechoic internal echoes without calcifications with a BI-RADS score above or equal to 3 [5].

Conflicts of interest

None.

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Ethical approval

N/A. Since this is a case report, not a research study, ethical approval has been exempt/not required.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

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Quality control of data and algorithms: N/A
Data analysis and interpretation: N/A
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