INTRODUCTION

Ectopic breast tissue is very rare and found most commonly in the axilla; however, the vulva is the second most common location.\(^1\) It is estimated that ectopic breast tissue occurs in 2% to 6% of women in the general population, with the vulva considered a rare site.\(^2\) In 2012, there were 10 reported cases of ectopic breast tissue on vulva worldwide.\(^3\) This tissue can arise anywhere along ectodermal primitive milk streaks as a result of incomplete involution during embryonic development.\(^4\) While the cause of a new presenting vulvar mass may be unclear, a previous case report suggested that ectopic breast tissue in the vulva can be related to pregnancy, and should be suspected in a patient presenting with an enlarging mass in the postpartum period.\(^2\)

Other causes of breast tissue in the vulva are thought to be a normal part of the anogenital area, as these lesions can look very similar to mammary glands, but there is very limited literature as most masses have been reported as ectopic breast tissue.\(^5\) Since ectopic breast tissue functions in the same way as normal breast tissue, the ectopic tissue is subject to hormonal stimulation, thus often appearing in pregnancy, with lactation, or with menstrual cycles.\(^2\)

Imaging, such as magnetic resonance imaging, has been used in other cases for characterizing mass lesions,\(^6\) however, in this case report, the mass originally was suspected to be a lipoma based on physical exam. To confirm the diagnosis, the tissue must be biopsied and assessed histologically.\(^5\) While there are other case reports of breast tissue in the vulva, it is extremely rare with various presentations, so further discussion is warranted. Breast cancer and fibroadenomas also have been reported on the vulva, so benign and malignant neoplastic processes should be on the differential for masses discovered in the vulvar region.\(^1,6-9\)

This report described a unique case of a unilateral vulvar mass found to be breast tissue. While vulvar masses associated with pregnancy and recurrence have been reported previously in the literature, this case was unique as the cause of this vulvar mass is unknown and completely random, with no associated conditions or correlation with pregnancy, lactation, or menstrual cycles or previous history of malignancy.\(^1,2,10,11\)

CASE REPORT

A 44-year-old woman presented to the gynecology clinic after a mass that had been present for two years had become increasingly uncomfortable. The patient reported itchiness and she also had concerns that the mass was increasing in size. She denied any purulent discharge or erythema around the area. The patient had no significant medical or family history.

The patient presented to a family medicine clinic for evaluation of the mass. Due to its size, she was referred for evaluation by a gynecologist. On physical exam, a 2 x 3 cm rubbery, mobile, nontender mass was present on the external genitalia, 3 cm lateral to the clitoris and near the mons. The remainder of her genital exam was normal.

Due to the size of the mass and discomfort the patient was experiencing, the patient underwent an uncomplicated excisional biopsy completed under spinal anesthesia. A 2 cm incision was made on the inferior portion of the mass and it was resected away from normal skin with Metzenbaum scissors. This was continued until the base was reached. The right labial 2.4 x 2.4 x 1.1 cm pink-tan nodule was removed in whole and sent to pathology in formalin. The specimen was sectioned serially to reveal pink-tan fibrous cut surfaces. The final pathology report was consistent with benign fibroadenoma, and the surgical pathologist reported that there was no normal breast tissue, which implies that this is likely a fibroadenoma arising from anogenital mammary-like glands rather than arising from ectopic breast tissue (Figures 1 and 2). At her follow up visit two weeks post-surgery, the patient reported no concerns. Her biopsy site was healing well with no signs of infection. No other imaging or diagnostic assessments were performed.

Figure 1. The biphasic lesion is composed of large, irregularly branching ducts and smaller glands in a lobular arrangement surrounded by hypocellular, dense collagenous stroma. The periphery of the lesion appears well-circumscribed but unencapsulated. (Image 4x)

Figure 2. Both the stromal and epithelial components are cytologically bland and lack mitoses. Scattered myoepithelial cells with clear cytoplasm easily are identified surrounding the ducts and glands. The histologic appearance is identical to fibroadenoma of the breast. (Image 20x)
Ectopic breast tissue is a benign finding that can present as a vulvar mass. Clinicians providing routine gynecologic care may encounter this rare finding in the course of their career.

Ectopic tissue in women most frequently can be found in the axilla, but also anywhere along the milk line as well as beyond it. The ectopic tissue can become tender with menstrual cycles and experience pathology, similar to what is expected of normally located breast tissue. Other cases of ectopic breast tissue in post-menopausal women have been reported, indicating that the glands can remain dormant for many years, then present after menopause has occurred.12

Common pathologies that can be found in ectopic breast tissue include fibrocystic changes, mastitis, fibroadenoma, and carcinoma.1,6-9 Specifically, pathologic cases of ectopic breast tissue with adenocarcinoma, intraductal papilloma, and fibroadenoma have been reported. Vulvar fibroadenomas initially may be misdiagnosed as an epidermal cyst, follicular cyst, Bartholin’s gland-duct cyst, or lipoma and can be confirmed only with biopsy.13

Determining the type of ectopic breast tissue present is important because management will differ. There were cases of malignant ectopic breast tissue that were managed with immunohistochemical staining and sentinel lymph node biopsy, and hormone therapy.14-18 Due to the rarity of these conditions, there are no clear guidelines on management. As in the case presented here, fibroadenomas can arise from anogenital mammary-like glands. Many causes of ectopic breast tissue have been reevaluated due to the recognition of mammary-like anogenital glands (MLAG), which are a normal constituent of the vulva.19 Further evidence that this tissue is most likely due to MLAG, as opposed to ectopic breast tissue, is that vulvar MLAG cannot be derived from the mammary ridges or milk lines since the breast and vulva are separated widely by time and space.19 Since MLAG tissue can contain features consistent with eccrine or apocrine tissue, these glands are most likely the cause of masses presenting in the anogenital region.

The diagnosis of breast tissue in the vulva can be done with fine needle aspiration cytology or excisional and incisional biopsy.4 For symptomatic masses, the breast tissue should be excised completely to prevent recurrence of symptoms. In this case, the patient presented with an enlarging mass associated with pruritis with complete resolution of her symptoms after surgical removal. To our knowledge, there is no previous case report on a mass with pruritis as a presenting symptom. There is limited literature describing ectopic breast tissue in the vulva presenting as an enlarging mass over time.2,4,10,13,19 One report specifically described vulvar ectopic breast tissue presenting as an abscess with pain and discharge, as well as vulvar swelling that had increased in size over the course of two years.10

In our case, the patient has not had any recurrence of symptoms to date; however, recurrent vulvar breast fibroadenoma has been reported in the literature and estimated to be around 3%.3 Based on this report, masses that were greater than 2 cm or masses with incomplete removal were associated with higher risks of recurrence, so further surveillance and close follow-up is warranted for larger masses and those with concerning features. An asymptomatic nodule in the vulva consistent with fibroadenoma of the breast also has been reported.13 Vulvar masses can present at any age with various symptoms, and both benign and malignant masses should be considered. It would be reasonable to include ectopic tissue, as well as mammary-like glands of the vulva, on the differential for vulvar pruritis when no other obvious pathology is identified, as pruritis can be a preceding sign of vulvar masses that are too small to be detected. A misdiagnosis of vulvar masses as ectopic breast tissue instead of mammary-like glands which are normal in the vulva should be considered in other cases.

DISCUSSION

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