

CASE REPORT CASO CLÍNICO

1. Department of Obstetrics and Gynecology,
Lehigh Valley Health Network, Allentown,
Pennsylvania, U.S.A.

- a. Attending Physician
- b. Obstetrics and Gynecology Resident

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Correspondence:

Guillermo Antonio De La Vega

143 Nursery Street, Suite 202, Fogelsville,
PA 18051

+1 (856) 397-5536

guillermo_a.delavega@lvhn.org

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Active ectopic breast tissue in the vulva of a postpartum patient. A case report Tejido mamario ectópico activo en vulva en paciente posparto. Reporte de un caso

Guillermo Antonio De La Vega^{1,a}, Emma Jean Qureshey^{1,b}

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ABSTRACT

Ectopic breast tissue (EBT) occurs in 0.3-6% of females and is typically located in the axilla, but it can also be found in the vulva. Diagnosis often occurs when there are hormonal changes that influence growth and development of the tissue. Due to symptomatology and malignant potential of this ectopic tissue, excision is typically recommended. We present the case of a 31-year-old woman, gravida 1 para 1001, attended four weeks after a normal vaginal delivery for a painless vulvar mass. Incision, drainage and biopsy were performed, revealing ectopic breast tissue with lactational changes. Simple partial vulvectomy revealed fibrocystic and lactational changes in mammary tissue of the vulva without atypia or malignancy. Our case displays a rare finding of aberrant, lactating breast tissue presenting in a postpartum patient.

Key words: Breast tissue, ectopic, Vulva, Postpartum.

RESUMEN

El tejido mamario ectópico (TME) ocurre en 0,3 a 6% de las mujeres y generalmente se localiza en la axila, pero también puede ser encontrado en la vulva. El diagnóstico a menudo ocurre cuando hay cambios hormonales que influyen en el crecimiento y desarrollo del tejido mamario. Generalmente, se recomienda la escisión debido a la sintomatología y al potencial maligno de este tejido ectópico. Presentamos el caso de una mujer de 31 años de edad, grávida 1 para 1001 que fue evaluada por una masa vulvar indolora cuatro semanas después de un parto vaginal. Se realizó incisión, drenaje y biopsia de la masa, obteniendo 30 mL de líquido lechoso. El resultado de la biopsia reveló tejido mamario ectópico con cambios de lactancia. La paciente fue sometida a vulvotomía parcial simple que mostró cambios fibroquísticos y de lactancia en el tejido mamario de la vulva, sin atipia ni malignidad. Nuestro caso es un hallazgo raro de tejido mamario aberrante en una paciente posparto.

Palabras clave: Tejido mamario, ectópico, Vulva, Posparto.



INTRODUCTION

Breast tissue begins to develop during the sixth week of gestation as bilateral bands of ectodermal thickenings called milk lines. This tissue runs from the axilla to the inguinal region. The location of persistent tissue varies among species, with humans having the majority of mammary tissue in the pectoral region⁽¹⁾. Ectopic breast tissue (EBT) occurs when there is an incomplete involution of this embryologic cellular line and is the most common congenital breast abnormality. This ectopic tissue can be unilateral or bilateral and is most commonly found in the axilla, followed by the vulva. EBT can be categorized as supernumerary breast which includes a communication to the outside skin via a nipple or an areola, or aberrant breast tissue with no communication to the overlying skin⁽²⁾. Tissue is present at birth; however, it does not become evident until puberty, pregnancy or lactation when under the influence of female sex hormones⁽³⁾. Due to the possibility of malignant changes in this tissue, complete preventive excision is recommended⁽⁴⁾.

CASE REPORT

A 31-year-old woman, gravida 1 para 1001 presented for her routine postpartum visit four weeks following a normal spontaneous vaginal delivery. Labor was induced at 37 2/7 weeks secondary to preeclampsia without severe features. Her past medical history was significant for asthma, rheumatoid arthritis, Sjogren's syndrome and reduction mammoplasty at age 22. At 18 weeks gestation she underwent an ultrasound-guided core biopsy of 2.9 cm left breast mass. Biopsy was consistent with a fibroadenoma and follow up-ultrasound at 6 months revealed an interval decrease in size.

On evaluation, she was complaining of an enlarged, non-tender, left vulvar mass for two weeks postpartum. The mass was gradually increasing in size and was not associated with any drainage. The patient was not breastfeeding. Physical examination revealed a normotensive, afebrile, well appearing woman. Local examination revealed a 5 cm x 3 cm x 3 cm non-tender and non-erythematous single cystic mass on the left labia majora. There was no tenderness to palpation, no warmth, lymphadenopathy or drainage that would suggest focal infection. Incision and drainage were performed yielding 30 mL of milky fluid and a biopsy was taken of the cyst wall.

Fluid culture revealed no aerobic or anaerobic growth and histopathology examination displayed features of vulvar mammary type tissue with fibrocystic changes, with one of the cystic areas showing intrapolyoid projections of hypocellular, mitotically inactive stroma into the gland lumina. Differential diagnosis included benign mammary tissue, partially sampled papilloma or partially sampled fibroadenoma. Complete excision was recommended to perform further evaluation. Immuno stains for estrogen receptor alpha (ER- α) and progesterone receptor alpha (PR- α) and beta (PR- β) were positive for 5% and 2% of nuclei respectively⁽⁵⁾.

The patient was taken to the operating room for excision due to concern for engorgement, pain, infection and potential for future malignancy. Prior to the procedure, local examination revealed a 2 cm x 2 cm non-tender, non-erythematous, non-draining cyst. She underwent a simple partial vulvectomy which was uncomplicated. Final pathology was consistent with the initial cyst wall biopsy of ectopic breast tissue of the vulva. It revealed fibrocystic and lactational changes in mammary like glands of the vulva (Figure 1). There was no atypia or malignancy identified (Figure 2).

DISCUSSION

As the most common congenital breast abnormality, up to 6% of women are thought to have EBT though it is difficult to diagnose. It is not typically recognized until there is pathology present whether that is benign or malignant tumors, fibrocystic changes, lactation, mastitis or abscess. Surgical excision should be strongly considered

FIGURE 1. ECTOPIC BREAST TISSUE IN THE VULVA SHOWING LOBULAR UNIT WITH DUCT AND ACINI AT 10X MAGNIFICATION.

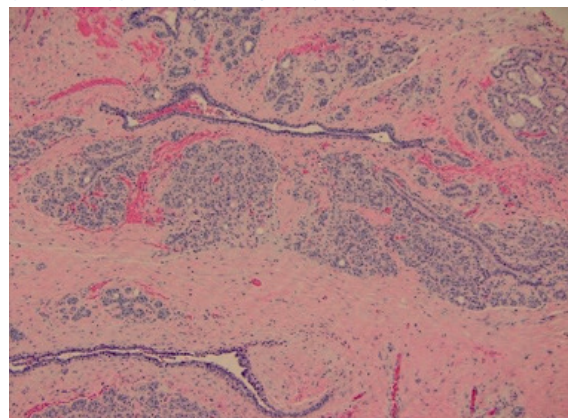
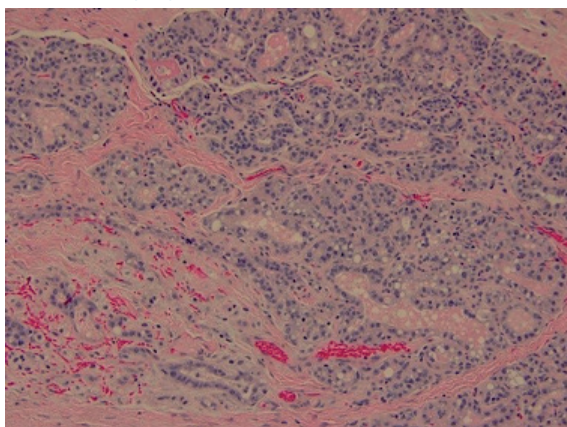




FIGURE 2. LACTATIONAL CHANGES IN THE VULVAR MAMMARY TISSUE WITH DILATED ACINI CONTAINING SECRETIONS AND VACUOLATED LUMINAL EPITHELIAL CELLS.



to reduce the potential for complications. During pregnancy, hormonal changes including increasing amounts of progesterone, prolactin and placental lactogen increase the development of breast tissue. In the postpartum period, prolactin and oxytocin control the synthesis and secretion of milk in mammary tissue. These changes occur along the milk lines wherever there is remaining active mammary tissue. In this patient, the continued production of milk leading to engorgement and deformity as well as need for full histologic evaluation were indications for removal. There is also the potential, albeit extremely low, risk of malignant transformation^(4,6).

Though a rare condition, it is important to consider this diagnosis as it carries the potential for malignancy. When malignancy is diagnosed in EBT it is most commonly adenocarcinoma or ductal carcinoma found in normal appearing breast tissue. Only 0.2-0.6% of all breast cancers are found in ectopic breast tissue making this an extremely rare diagnosis and even fewer (only 4% of those) are located in the vulva⁽⁷⁾. Other than cancer of the skin, breast cancer is the most common cancer diagnosed in women and is the second leading cause of cancer death in this population. In considering EBT for malignant potential, histopathologic evaluation is required. In addition, determining for expression of hormonal receptors, including estrogen receptors (ER), is helpful. Normal breast tissue expresses both ER- α and ER- β to varying degrees; ER- α is felt to have the majority of estrogenic effects in the breast. Studies have shown that expression of ER- β is linked to inhibition of tumor cell formation in response to estradiol and this may have a protective effect⁽⁸⁾.

Badejo⁽⁹⁾ reported that the incidence of malignant changes in accessory breast tissue was 14%. Cheong et al. reported that only 4 cases (0.28%) of breast cancer of ectopic breast tissue in the axilla were found in a total of 1 430 cases of breast cancer operated.

While primary vulvar breast cancer is extremely rare, it has been increasingly described. A recent review of the literature identified approximately 28 cases of primary mammary adenocarcinoma of the vulva in the English literature⁽¹⁰⁾. A population study in the Netherlands identified 5 cases of this type among 108 primary malignant neoplasms of the vulva from 2000 to 2015⁽¹¹⁾, but the true incidence of this type of breast cancer is unclear. In the absence of symptoms or concern of malignancy, it may be reasonable to consider expectant management.

Although a rare occurrence, EBT should be included on the differential diagnosis of a woman who presents either during pregnancy or in the postpartum period with complaints of enlarging vulvar mass with or without drainage. The majority of previously reported EBT cases have involved cancer and therefore treatment considered complete excision. Very little information is available regarding the management of benign EBT. In the absence of symptoms or concern for malignancy it may be reasonable to consider expectant management. Development of breast cancer remains a concern and further investigation is warranted to determine appropriate surveillance of EBT.

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