Fibroadenoma of Ectopic Mammary Tissue in the Vulva –
A Case Report

Maha M. Arafah, MD, and Shaesta N. Zaidi, MD

Department of Histopathology
King Khalid University Hospital, King Saud University
Riyadh, Saudi Arabia
marafah@hotmail.com

Abstract. Aberrant breast tissue can occur anywhere along the embryonic mammary streak. Its presence in the vulva is an uncommon clinical or pathological finding. The ectopic breast tissue can be the site of the same physiological or pathological process found in the normal breast. The developing of fibroadenoma is a rare entity and has rarely been reported in the literature. A case report of 46-years-old female was presented with an unknown progressively increasing mass in the vulva. Excision of the mass was performed, which on histology, it demonstrated the fibroadenoma with mammary tissue surrounding it, and with positive hormone receptors.

Keywords: Ectopic breast, Vulva, Fibroepithelial proliferation.

Introduction

Ectopic mammary tissue may occur anywhere along the primal embryonic milk line, which develops in the fifth week old human embryo and extends from the axilla to the groin, bilaterally. Incomplete regression of this band, results in accessory regions of the mammary tissue, and is found in 1% to 2% of the general population in form of supernumerary breast or accessory mammary tissue[1,2]. The tissue can be stretched from little focus parenchyma to a complete structure that includes the areola and breast nipple. Until now most report about
Accessory breast were in the axillary region\textsuperscript{[3,4]}. It can be seen outside the milk streaks at the back, in the thigh, face, hips, upper part of the arms and shoulders\textsuperscript{[4,5]}. Such ectopic tissue may undergo the same physiological and pathological processes found in the normal breast\textsuperscript{[6-9]}. Most of the reported cases in literature involve malignancy arising from ectopic breast tissue located in the vulva\textsuperscript{[10]}. There are few reports about the benign pathologies in vulva mammary gland. A report based on an additional case of fibroadenoma, developing in the vulvar accessory breast in an adult female is presented.

**Case Report**

A 46-years-old Saudi female, gravida 2; para 2, was presented to our institution with four months history of an off and on discharge in the vagina, and a vulvar mass of progressive growth for the past 8 months. The previous medical and family history was not contributory to the present illness. Physical examination revealed a fungating-ulcerated mass measuring 4 cm, located in the right labia majora. The mass was soft and movable, but not adhered to the skin or other structures. Fine-needle aspiration of the mass was performed, which was inconclusive. Radiological investigations reported no mass or retroperitoneal lymph node enlargement.

The patient underwent complete surgical excision of the mass under local anesthesia.

Grossly, a well-delimited, multilobular mass, partially covered by mucosa with focal ulceration, measuring 4.2 x 4 x 4 cm was acknowledged.

The cut section showed a lobulated white firm mass without necrosis and hemorrhage. Microscopically, a fibro-epithelial proliferation was seen with a well-defined border, surrounded by a rim of a normal mammary tissue. The proliferative epithelial structures were composed of slit like spaces (Fig. 1), which were lined with bilayers of inner secretory epithelial as well as outer myoepithelial cells. In certain places, the inner layer showed apocrine changes with many apical snouts and eosinophilic cytoplasm (Fig. 2). No evidence of epithelial hyperplasia was seen. The fibromyxoid stroma consisted of spindle cells with elongated nuclei, small nucleoli and little cytoplasm. However, there
was no evidence of atypia or mitosis in the stroma or glands. The surface was lined by keratinizing stratified squamous epithelium with extensive inflammation. Immunohistochemical study showed a diffuse positivity for cytokeratin-7, estrogen and progesterone receptor proteins in the epithelial cells (Fig. 3). The myoepithelial cells showed CD10, S-100 and SMA, positivity (Fig. 4).

The morphological appearance and immunohistochemical profile were consistent with the diagnosis of fibroadenoma of the ectopic breast tissue in vulva.

![Ectopic breast tissue histology. Proliferating dilated, branching glands surrounded by loose stroma (H&E, 20x).](image1)

![The glands lined by two cell layers, the inner layer showing apocrine changes (H&E, 40x).](image2)
Fig. 3. Estrogen receptor positivity in the epithelial cells (x10).

Fig. 4. CD10 positivity in myoepithelial cells (x10).

Discussion

Ectopic breast tissue has been found in multiple locations; along the milk line from the axilla to the vulva, although, the axilla is the most common area. In contrast, vulvar location is extremely rare, and to our knowledge there are only few cases reported in the literature[^1-^3^]. Most instances occur spontaneously; however, familiar cases have been reported in up to 10% of the affected population.
Ectopic breast tissue has been subjected to physiological and pathological changes just as the same as in the normally situated breast. Clinically, most of the lesions go undetected in prepubertal ages, as this tissue tends to remain quiescent until stimulated by hormones; especially the hormonal effects of puberty, pregnancy and lactation[14].

Histologically, aberrant breast consists of all three elements-paranchyma, areola and nipple, or a combination of all of them. Usually, there are large ducts without focally developed lobules or terminal ductules[11].

Ectopic breast (extramammary) lesions of the vulva, like fibroadenomas are rare and difficult to distinguish from other labial masses on physical examination, however they have been documented in the literature as far back as 1872[15]. For a long time, supernumerary mammary glands derived from rudiments of the embryonic milk lines in the vulva and were considered the possible etiology of these unusual tumors. Recently, there has been more focus on mammary-like anogenital glands as a possible source[16].

Since 1954, when Burger and Marcuse[17] reported their two cases of ectopic breast fibroadenoma of the vulva, more than 22 similar cases have been reported in the literature worldwide in the last 5 decades[7,8,16-22].

Several investigators have been reported of a higher incidence of urinary tract anomalies, such as hydronephrosis, polycystic kidneys, ureteric stenosis, and supernumerary kidneys, in patients with ectopic breast lesions[23,24]. The patient in this report also underwent a renal ultrasound examination, but no anomalies were revealed.

To conclude, this case of ectopic breast tissue in the vulva, which to our knowledge, is the first reported case experience in the Saudi population. Because of its rarity, when encountered as a solitary vulvar mass, it is not easy to consider the possibility of ectopic breast. However, physicians should be conscious of their existence when formulating the differential diagnosis and for that, a histopathological confirmation is crucial to reach the final diagnosis. A renal study should be considered once the diagnosis was achieved. Ectopic breast tissue can undergo malignant transformation, therefore, surgical excision is generally recommended, along with long-term follow-up of the patient.
References


Fibroadenoma of Ectopic Mammary Tissue in the Vulva


نمو غذوم ليزي من أنسجة ثنية منتذبة في المهبل،
تقرير حالة

ما محمد عرف، وشايستا نسيم زيدي
جامعة الملك سعود
الرياض – المملكة العربية السعودية

المستخلص. يمكن لالانسجة الثنية المنتذبة أن توجد في أي مكان
على طول خط الثدي الجنيني، ويندر وجود هذه الأنسجة في المهبل،
كما يتكرر تطورها إلى أورام حميدة. في هذه الحالة نقرر نمو غذوم
ليزي عند أومرة تناهز 46 عاماً من العمر، والتي كانت تعاني من
وجود كتلة في المهبل ازداد حجمها تدريجيًا، مما استدعى استئصاله
جراحيًا. الفحوص المجهرية النسيجي لهذا الورم، بين أنه ورم حميد
متكون من غذوم ليزي شبيه بالغذوم الليفي الذي يتكون في الثدي،
كما يبين أيضًا أن الخلايا بها مستقبلات هرمونية لمادة الإستروجين،
والبروجسترون كما في الثدي. وتمراجعة البحوث العلمية الموجودة
باللذينيات الإنجليزية تبين وجود حالات قليلة مشابهة لهذه الحالة.