

Phyllodes Tumor of the Vulva: Report of Two Cases

Vulvanın Fillods Tümörü: İki Olgu Sunumu

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ABSTRACT

Ectopic breast tissue can occur anywhere along the primitive embryonic milk line and can be the site of the same pathologic processes found in the normal breast. Phyllodes tumor is an extremely rare fibroepithelial neoplasm that occurs in ectopic breast tissue of the vulva. To date, only 8 cases of phyllodes tumor in the vulva have been reported in the literature. This paper presents two additional case of benign phyllodes tumor in the vulva. The first patient was a 43-year-old woman, presenting with a lesion on the left anterior mons pubis that had increased in size in the last three months. The second patient was a 50-year-old woman, presenting with a two-month history of a mass in the right labium majus. The patients underwent excisional biopsy. The histological examination of both specimens revealed a characteristic pattern of benign phyllodes tumor. In conclusion, the pathologists need to be aware that the lesion can occur in this location. Herein, the concepts about the histogenesis of phyllodes tumor in the vulva are discussed and a brief review of the previously reported cases is given.

Key Words: Vulva, Phyllodes tumor

ÖZ

Ektopik meme dokusu ilkel embriyonik sütyolu boyunca herhangi bir yerde ortaya çıkabilir ve normal memede izlenebilen aynı patolojik durumlar ektopik meme dokusunda da görülebilir. Fillods tümör vulvadaki ektopik meme dokusunda oluşan nadir bir fibroepitelyal neoplazidir. Literatürde bugüne kadar 8 olgu bildirilmiştir. Bu makalede literatüre ek olarak iki ayrı vulvar fillods tümör vakası sunulmuştur. İlk olgu, sol ön mons pubiste boyutu son üç ayda artış gösteren bir lezyon ile kliniğe başvurmuş 43 yaşında kadın hastadır. İkinci olgu 50 yaşında kadın hasta olup, iki aydır varolan sağ labium majusta kitle ile başvurmuştur. Mevcut lezyonlara eksizyonel biopsi yapılmış ve örneklerin patolojisi benign fillods tümör ile uyumlu rapor edilmiştir. Sonuç olarak, fillods tümörün vulva lokalizasyonunda da karşılaşılabilecek bir antite olduğu unutulmamalıdır. Burada, vulvar fillods tümörün histogenezi ile ilgili kavramlar tartışılmış ve olgularımız daha önce bildirilen olgular eşliğinde sunulmuştur.

Anahtar Sözcükler: Vulva, Fillods tümörü

INTRODUCTION

Phyllodes tumor is an uncommon fibroepithelial neoplasm that accounts for less than 1% of all breast tumors. It is also known to occur in various other anatomical sites, including the vulva, prostate and axilla (1-9). The tumor has epithelial and stromal components. It grows as leaf-like projections into the glandular lumens. The stroma is usually hypercellular, typically with periglandular condensation. The grade is defined by the stromal atypia as benign, low or high grade phyllodes tumor (10).

Ectopic breast tissue can occur anywhere along the primitive embryonic milk line bilaterally, extending from the axilla to the groin. Such ectopic breast tissue can be the site of the same physiological and pathological processes found in the normal breast. The occurrence of phyllodes

tumor arising within vulva is extremely rare (1-7). Herein, we report two cases of benign phyllodes tumor originating in ectopic breast tissue of the vulva.

CASE REPORTS

Case 1

A 43-year-old woman was referred for evaluation of a lesion on the left anterior mons pubis. The patient had noted the appearance of slightly raised, firm nodule a year ago and the lesion gradually began to increase in size in the last three months. She had no history of malignancy or breast disease. The patient underwent excisional biopsy of the lesion.

Case 2

A 50-year-old woman presented with a two-month history of a painless, non-tender subcutaneous mass in the right

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labium majus. She had been postmenopausal for about 1 year. There was no history of prior malignancy or breast disease. The lesion was excised surgically.

Pathologic Findings

The tumor of the first case was elastic, oval with a white-yellow cut surface and included cleft-like spaces. It measured 4.7x3.5x2.3 cm (Figure 1). The tumor of the second case was 3.2x2x1.2 cm in size. Macroscopically, it was oval and lobulated with a glistening white-yellow cut surface (Figure 2).

The histological examination of both specimens revealed biphasic tumor composed of bilayered ductal component and fibroblastic stroma (Figure 3A, B). The epithelium was hyperplastic in both tumors, and focal squamous metaplasia with keratinization was detected in the sections of the first case (Figure 3C). The stroma was prominent and interrupted by leaf-like spaces lined by epithelium. The stromal cellularity was marked in focal areas adjacent to the intratumoral ducts. No mitotic activity or cellular atypia were found. The entire specimens of both cases were sectioned and blocked, but no normal breast tissue was identified.

DISCUSSION

The breasts develop from ectoderm at about the fourth to sixth week of the embryogenic period. The thickened ectoderm bilaterally forms mammary ridges or “milk lines” on the ventral surface of the embryo, extending from the axilla region to the groin. The breast tissue continues to develop in the normal pectoral region of the milk line, with regression of the remainder of the mammary ridges. As a classical knowledge, it is accepted that failure of this regression leads to the development of ectopic breast tissue (11).

The true incidence of ectopic breast tissue is not known. It is found much more frequently in women. Although the tissue is present at birth, it does not become prominent until influenced by female sex hormones at puberty, pregnancy, or lactation. The axilla and vulva are the most frequent sites of presentation (11).

A variety of benign and malignant lesions have been reported in ectopic breast tissue. These include intraductal papilloma, fibroadenoma and adenocarcinoma (11). Phyllodes tumor is extremely rare in the vulva and only 8 cases have been previously reported in the literature. These patients have usually presented with a gradually enlarging mass, but also a long-standing stable mass and cyst were encountered. The most common anatomical sites were the

labium majus and minus, but the interlabial sulcus and mons pubis were reported once. They were usually solitary except the case reported by Tbakhi et al. (1). All the tumors were microscopically identical to its counterpart in the breast, with a characteristic biphasic phenotype of phyllodes tumor and leaf-like configuration (1-7). The differential diagnosis of phyllodes tumor of the vulva includes the

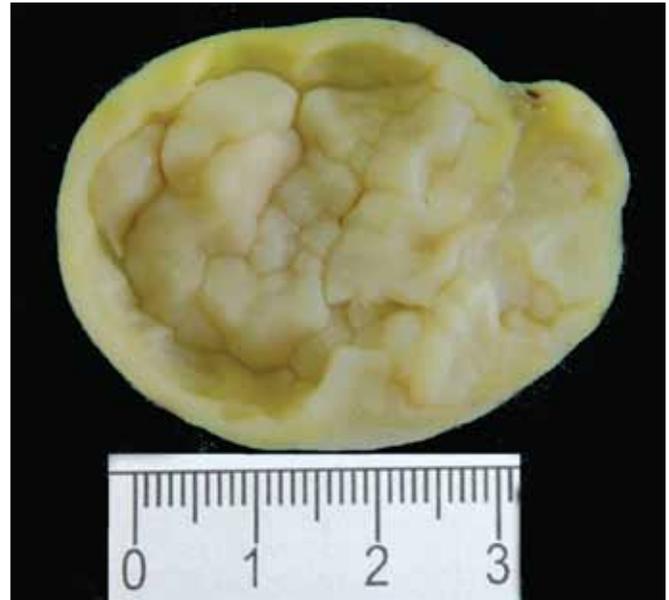


Figure 1: The tumor was oval in shape with a white-yellow cut surface and included cleft-like spaces.



Figure 2: The tumor was oval and lobulated with a glistening white-yellow cut surface.

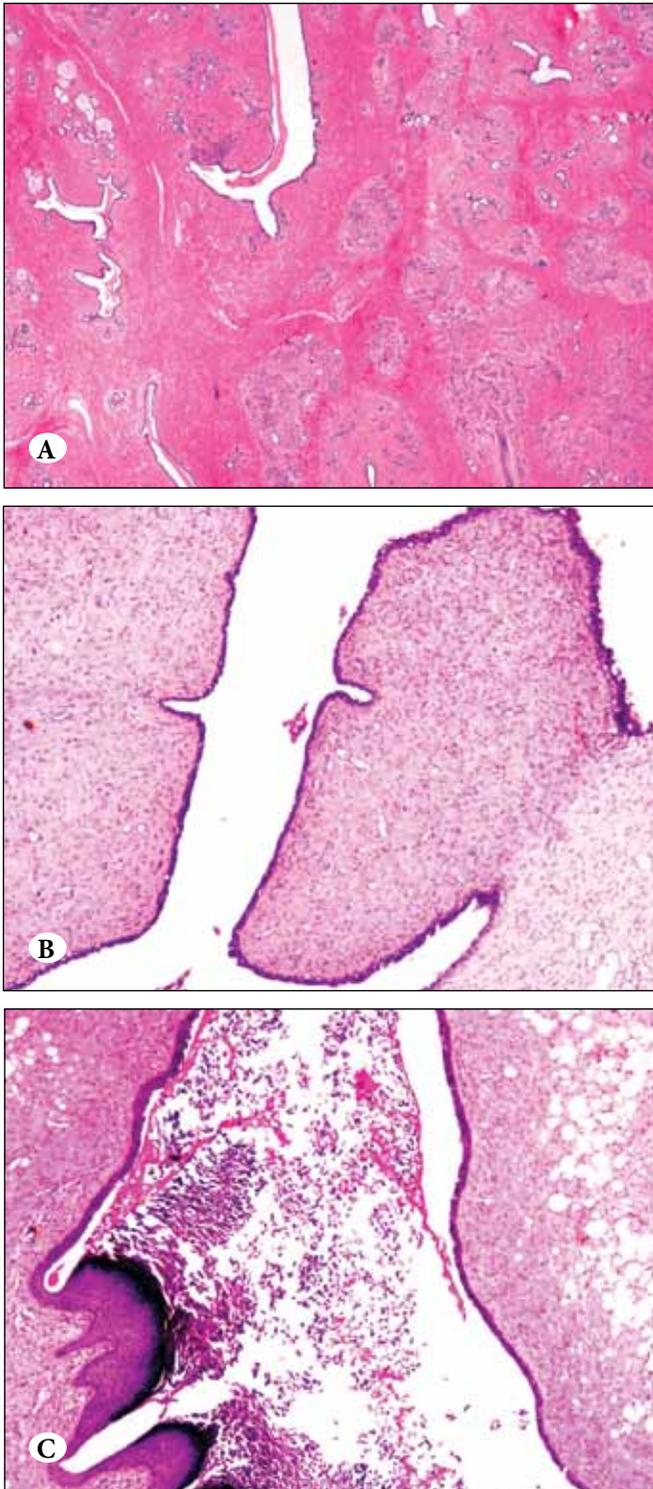


Figure 3: (A-B) The tumor was composed of fibroblastic stroma interrupted by leaf-like spaces lined by epithelium (A: H&E, x2, B: H&E x10), (C) The epithelium was hyperplastic and had focal areas of keratinizing squamous metaplasia (H&E, x10).

biphasic tumors such as chondroid syringoma and mixed Müllerian tumor (10). The behavior of phyllodes tumor of the vulva is difficult to predict by histology alone, as the existing data is limited. During follow-up, the reported cases were unremarkable except for one borderline case and two cases with recurrence (1-7).

Some authors believe that mammary ridges never reach the vulva during their embryogenesis. Van der Putte described a distinctive variant of cutaneous glands (12). In normal histology, the sebaceous glands are predominantly present on the medial surface of the labia majora with or without hair follicles. Apocrine and eccrine sweat glands are generally only found in hair-bearing areas (13). Van der Putte preferred to call these lesions mammary-like anogenital sweat glands rather than ectopic breast tissue that combine features of eccrine and apocrine glands (12). These glands showed the capacity to branch into lobuli and to form acini-like mammary glands. They were reported to take place in the interlabial sulcus, the paramedian area of the perineum and around the anus. Existence of normal breast tissue or mammary-like glands around the lesion may give some clues about histogenesis. Unfortunately, there was no non-neoplastic tissue around the lesions in our cases. Similarly, in the literature, many of this kind of lesions have been included into this category without the presence of co-existing normal glands.

In conclusion, histopathological findings could not help for determining the histogenesis of these lesions. Whatever the histogenesis, the present tumors were histologically identical to benign phyllodes tumor of the breast and only 8 cases of phyllodes tumor in the vulva have been reported in the literature.

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