

Invasive ductal carcinoma of the mammary like glands of vulva-with sentinel lymph node identification

Vulvanın meme benzeri bezlerinin invaziv duktal karsinomu-sentinel lenf düğümü incelemesi

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Abstract

Breast like carcinomas of the vulva was thought to be originated from ectopic breast tissue or metastasis from breast. It is now accepted that, as mammary-like glands are present in the vulva, any pathology that is found in the breast can be seen at these glands. Adenocarcinoma of the vulva is rare and the breast-like adenocarcinomas are presented as case reports in the literature. We present a primary breast-like carcinoma of the vulva in a 70 year old patient, who had complaint of vulvar mass and bleeding from it. The incisional biopsy was reported as vulvar adenocarcinoma. The intraoperative diagnosis of sentinel lymph node was adenocarcinoma metastasis and hemivulvectomy together with bilateral inguinal lymphadenectomy was carried out. Adenocarcinomas of vulva are very rare and breast like carcinoma must be kept in mind in these cases, as the prognosis and treatment is similar to primary breast carcinomas.

Keywords: Vulva, vulvar neoplasms, breast, carcinoma, ductal

Özet

Vulvanın meme benzeri karsinomlarının ektopik meme dokusundan kaynaklandığı veya meme karsinomu metastazı olduğu düşünülmüştür. Ancak, şu anda, vulvada meme-benzeri bezler olduğu için, memede görülen herhangi bir patolojinin bu bezlerde görülebileceği kabul edilmektedir. Vulvanın adenokarsinomu nadir olup, meme-benzeri adenokarsinomları literatürde olgu sunumları şeklinde yer almaktadır. Vulvar kitle ve kanama şikayetiyle başvuran 70 yaşındaki hastada tanımladığımız vulvanın primer meme benzeri karsinomu olgusunu sunuyoruz. İnsizyonel biyopsi vulvar adenokarsinom olarak raporlandı. İntraoperatif sentinel lenf düğümü incelemesi adenokarsinom metastazı olan hastaya hemivulvektomi ve bilateral inguinal lenfadenektomi uygulandı. Vulvanın adenokarsinomları nadir olup, prognoz ve tedavi primer meme kanseri ile benzerlik gösterdiği için bu olgularda meme benzeri karsinom akılda tutulmalıdır.

Anahtar sözcükler: Vulva, vulvar tümörler, meme, karsinoma, duktal

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Introduction

Mammary glands can be found as ectopic tissue in various sites in the milk ridge. Mammary tissue is described in anogenital region in 1872 by Hartung [1]. Its presence was accepted as ectopic for a long time, and presented as case reports in the literature [2-4]. In 1991 van der Putte [5] described ‘anogenital sweat glands’ with forming lobulus like structures in occasion. After then, in their review in 1994 [6] they made the explanation of ‘mammary like glands of vulva’ to be presented in the interlabial sulcus and stated that the ectopic breast tissue in the vulva is a ‘myth’ as there is no observational finding. Mammary like glands of anogenital region are normally not seen unless a pathology is present [1]. Most common pathology is sclerosing adenosis although any breast pathology can be seen [1, 5, 7]. The clues for the tumor to be primary in the vulva are presence of in situ component [2, 8, 9]; but it is advised to search for any breast pathology as there can be synchronous or metastatic breast tumor [1]. The treatment regimens differ according to stage of the disease, with wide resection or chemotherapy and be similar to breast carcinoma treatment [2, 4, 8-12].

Case report

A 70 year old, gravid 4, para 2 woman had applied to the gynecology clinic, with the complaint of vulvar mass and bleeding from it. On physical examination a tumoral mass of 2 cm in diameter on left labium majus was found. The CT scan, chest x-ray and tumor markers were all in normal ranges. The incisional biopsy was done and the result was vulvar adenocarcinoma. Left modified radical hemivulvectomy with sentinel lymph node evaluation was carried out and pathologic evaluation of sentinel nodes was positive for metastasis. Toluidin blue method was used for sentinel lymph node mapping. Right inguinal lymph nodes were also palpable and contralateral lymph nodes were resected although primary tumor is unilateral and lateral to center line. At the pathological examination, there was a mass with ulceration on the skin, of 4 cm in diameter, with a white colored cut section. The microscopic findings (Figure 1) showed an infiltrating carcinoma with cords and tubules and focal areas of solid growth. The morphology was very similar to invasive ductal carcinoma of the breast. The patient had mammography 5 months ago and was normal (Figure 2). The entire specimen was examined and foci of in situ ductal carcinoma were seen. The epidermis was normal morphologically, and with PAS staining. The immunohistochemical examination showed cytokeratin (CK) 7, carcinoembryonic antigen (CEA), estrogen receptor (ER), progesterone receptor (PR) and E-cadherin (Figure 3) positivity. Gross cystic disease fluid protein-15 (GCDFFP-15) was focally and weakly positive.

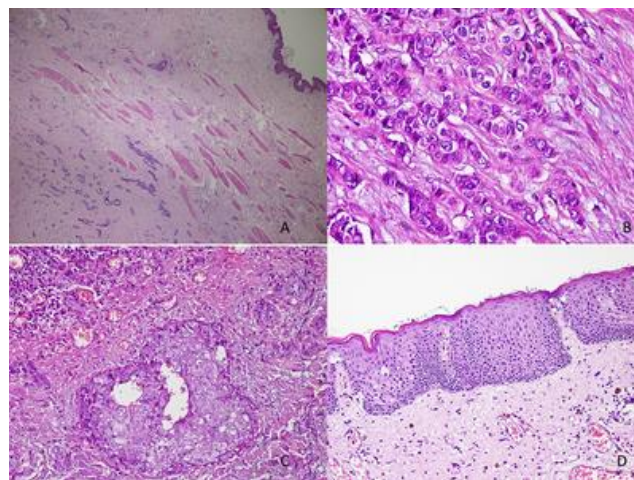


Figure 1A, B, C, D. A; infiltrative tumor in a desmoplastic stroma under epidermis (HE; X40), B; at higher magnification, ductular structures are seen (HE; X400), C; in situ ductal carcinoma adjacent to invasive tumor (HE; X200), D; epidermis showed no pathology (HE; X200).



Figure 2. Mammography showing no evidence for breast carcinoma.

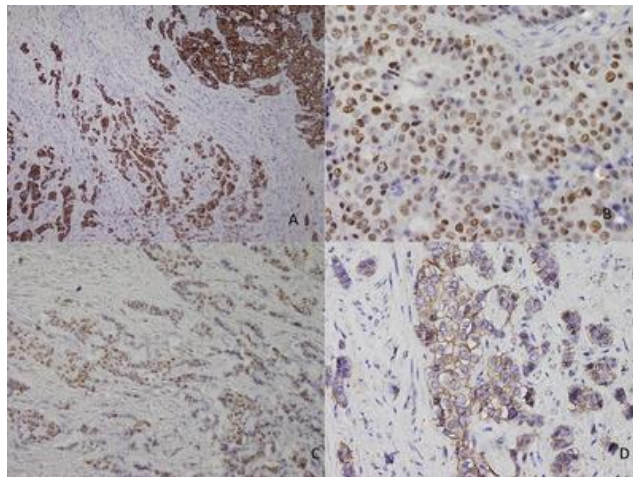


Figure 3. Immunohistochemical findings showing. A; CK7, B; ER, C; PR, D; E-Cadherin positivity (CK7;x200, ER;x200, PR;x200, e-Cadherin;x400 respectively).

Discussion

Vulvar adenocarcinomas are rare entities with most of them to be present with Paget's disease [7, 13] and a small group resembling breast carcinoma is described [1, 5]. For long years these tumors was considered to be originated from ectopic tissue or metastasis from breast carcinoma [2, 3]. van der Putte [6] was the first to describe 'mammary-like gland of the vulva' and their disorders. In their review in 2011, Kazakov et al. [1], stated the sclerosing adenosis to be the most common pathology and fewer than 25 cases were reported as carcinoma of 'mammary-like glands of vulva'. Our case was an adenocarcinoma with morphological resemblance to ductal carcinoma of the breast. The presence of in situ ductal carcinoma can be evidence for the primary [9, 14-16], as it was in our case, and the cases reported to be presented in ectopic breast tissue is based on the absence of this finding [2, 3, 8]. The majority of the reported cases are ductal type as in our case, but lobular or other types of carcinomas such as tubular, mucinous or adenoid cystic types were also reported [1, 12] although exceptional.

Immunohistochemistry was helpful in our case with ER, PR, GCDFP-15, E-cadherin positivity; and is reported to be useful by means of positivity with ER, PR [2, 8, 11], guinea pig milk-fat globule protein [2], GCDFP-15 [8]. There was no concomitant

Paget's disease in our case which is reported to be seen [4, 7, 17]. Aggressive prognosis with lymph node metastasis [2, 3, 7-8, 16] is frequent and bone metastasis has also been reported [10]. There was lymph node metastasis in our case but no distant metastasis is detected in a time of 8 months after hemivulvectomy. Treatment regimens are not standardized as there are few cases reported, but most of them are treated by surgery, followed by breast carcinoma like treatment modalities, such as chemotherapy [10] or tamoxifen [2, 8, 11]. Our case was operated due to vulvar carcinoma surgical therapy principles. Sentinel lymph node mapping using different techniques give the surgeon an opportunity to decide about the lymphadenectomy side and unilaterality as in our case. Usually gynecologic oncology surgery for vulvar carcinoma includes radical vulvectomy with bilateral inguinal lymphadenectomy. Lymphadenectomy may be unilateral if tumor is lateralized and 2 cm far from the central line of vulva. The treatment choice in our case was radical surgery followed by radiotherapy. Tumor was excised with a hemivulvectomy and ipsilateral lymphadenectomy was done. Contralateral lymph nodes were excised because of their enlargement. Mammary type carcinomas of vulva are speculated to be synchronous with breast carcinomas although it is very rare [1]. There was no concomitant breast carcinoma in our case.

In conclusion breast like carcinomas can be seen in vulvectomy specimens. The pathologist should be aware of their presence and, search for in situ component as evidence for primary carcinoma. Although there is no case series to standardize the treatment and set guidelines; wide radical excision should be completed with lymph node dissection due to high lymph node metastasis risk.

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