

An unusual case of mammary gland-like carcinoma of vulva: case report and review of literature

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Summary

Introduction: Accessory breast tissue is a very rare finding in the general population with an incidence of one to two percent. An even rarer occurrence is accessory mammary-like tissue which developed breast carcinoma. The authors present a case of aggressive and metastatic carcinoma of vulvar originating from mammary-like tissue. **Case:** A 73-year-old Caucasian female presented with a lesion in her left vulva. The lesion was ulcerated and fragile. A dermatologist had evaluated the lesion and took a punch biopsy. Result was vulvar carcinoma. She was admitted to the gynecologic oncology clinic then after and was operated. After a radical vulvectomy and bilateral inguinal lymphadenectomy she received adjuvant radiotherapy because of lymph node metastasis. One year after the finish of radiotherapy patient was found to have lung and femur metastasis. She began to receive systemic chemotherapy for metastasis. **Conclusion:** Primary mammary-like adenocarcinoma of the vulva is exceedingly rare. There is no consensus about the diagnosis, treatment, and follow up of these patients in literature. However, given that histological data confirms these cancers are behaving like breast cancers instead of known patterns of vulva cancer, the best treatment practices for breast cancer may be applied to treat these vulvar carcinoma patients.

Key words: Vulvar carcinoma; Ectopic breast tissue; Prognosis; Treatment.

Introduction

Ectopic breast tissue may occur anywhere along the primitive milk line from the axilla to vulva [1, 2]. These type of accessory breast tissue is a very rare finding with an incidence of one to two percent [3]. Usually these glands are silent and may only be active during pregnancy and lactation. Ectopic breast tissue may include all the normal components of normal located breast tissue; areola, nipple, and parenchyma [3]. This ectopic tissue responds to all physiological and hormonal changes like the normal one. Malignant changes can be also seen in these abnormal located mammary tissue. Vulvar primary carcinoma of ectopic mammary tissue is an exceptionally rare disease and only 26 cases being reported in the English literature between 1872 and 2012 [4, 5]. Here the authors described vulvar carcinoma arising from mammary-like glands of vulva.

Case Report

A 73-year-old G4P2 Caucasian female presented with a bleeding mass to an urban hospital dermatology clinic. Her general health was good and had a history of hysterectomy together with bilateral oophorectomy for endometrial hyperplasia 17 years ago. A topical steroid treatment was given for three weeks and a biopsy was carried out after that treatment period. Pathologic evaluation of the biopsy revealed that lesion was adenocarcinoma

of the vulva with squamous differentiation areas. Patient was referred to the present gynecologic oncology clinic then after. There was a nearly two-cm mass in the left labium minus including the posterior forchette. She had a normal mammography result six months before the presentation. An inguinal lymph node survey was negative for masses. Speculum evaluation of the vaginal cuff was normal.

Computed tomography (CT) scan of the pelvis and abdomen with IV contrast showed no intra-abdominal metastasis. All pre-operative blood tests were in normal ranges. Radical vulvectomy and bilateral inguinal lymphadenectomy were carried out in the present clinic. Final pathology revealed that it was a vulvar mammary-like gland adenocarcinoma of 3 x 2 x 3 cm. Histologic grade was 2 and there were two metastatic lymph nodes in every side. Immunohistochemical analysis of tumoral tissue was found as: GCDFP-1 weak positive, P63 negative, ER positive, PR positive, CK8/18, CK7, CK14, and E-cadherin all positive.

Patient was discharged after four days of hospital stay after operation and oncology board decided to give her an adjuvant radiation therapy. Between October 19, 2011 and December 1, 2011 vulvar tumor bed, bilateral inguinal and pelvic lymph nodes were treated curatively using volumetric modulated arc radiotherapy. Total dose of 6,000 cGy and 5,000 cGy was given in 30 fractions to the vulvar tumor bed, bilateral inguinal lymph nodes, and pelvic lymph nodes using simultaneous integrated boost technique. Six MV photons and daily kV-kV image guidance was used during treatment. Grade III dermatitis and grade II diarrhea occurred during treatment.

Eighteen months later after the first surgery, pulmonary lesions were found in pulmonary CT scan and positron emission tomog-

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raphy computed tomography (PET-CT) confirmed that these lesions were metastasis. Two more metastatic lesions in left femur and L3 vertebra was also diagnosed by this PET-CT scan. Patient had no complaint from pulmonary system and no finding in the vulvar operation area during this time period. She had only moderate degree bone pain in left humerus.

An oncology board consultation was done and it was decided to begin an aromatase inhibitor and zoledronic acid. A second palliative radiotherapy application to bone metastasis was also planned. In February 2013, total palliative dose of 2,000 cGy was given in five fractions to the L3 vertebra and left 1/3 upper humerus using 18 - 6 MV x-rays. Three months after beginning of this therapy regimen all the pulmonary lesions were found to be decreased in size and number and bone metastasis totally cured. Patient is still under same chemotherapy regimen without any complaint.

Conclusion

The first reported case of ectopic mammary gland adenocarcinoma located in vulva was reported by Hartung in 1872 [6]. There are nearly 26 cases worldwide in English literature to date [5]. This rare tumor type seems to be growing slowly for years in the vulvar area without complaints. The only way to correctly diagnose and treat this tumor type requires a good and detailed pathological evaluation of both biopsy specimen and surgically resected tissue. Adjuvant treatment regimens may include breast carcinoma experiences of medical oncologists and hormonal therapies like tamoxifen usage or aromatase inhibitors must be kept in mind. Management of patients

must be directed like breast carcinoma in every step, although lesion seems to be a gynecologic oncology issue.

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