An unusual secondary extramammary Paget’s disease of vulva

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Abstract

Extramammary Paget's disease (EMPD) of the vulva is a rare disease that occurs mainly in postmenopausal women. It can be primary or secondary. Prognosis of primary EMPD is good, but when secondary, we need to find the underlying malignancy as the prognosis depends on it. Very few cases of secondary EMPD of the vulva have been reported till now. We hereby report a case of a 65-year-old postmenopausal woman with EMPD secondary to invasive adenocarcinoma with neuroendocrine features in the dermis and she was treated surgically followed by radiotherapy.

Key words: Extramammary Paget’s disease, genital malignancy, vulval carcinoma

INTRODUCTION

Extramammary Paget's disease (EMPD) is a rare neoplastic condition accounting for 6.5% of Paget’s disease. It predominantly involves the apocrine-rich areas such as penis, scrotum, vulva, perianal area and axilla, while vulva being the most common site.[1] It mainly occurs in elderly postmenopausal women. It can present as primary or secondary disease. Primary being intraepithelial adenocarcinoma arising within epidermis and secondary EMPD due to intraepithelial spread of adnexal carcinomas or visceral carcinoma.[2]

CASE REPORT

A 65-year-old, married, postmenopausal woman without any comorbidity presented to us with a slow-growing pruritic reddish lesion over vulval skin for 5 years. It started as a small pea-sized lesion which gradually enlarged to attain the present size over 5 years. No history of any bladder or bowel disturbances was noted.

On cutaneous examination, a single well-defined erythematous plaque measuring about 6 cm × 10 cm seen over the left labia majora extending posteriorly to perianal area with surface showing gray crusting and nodulation with serosanguinous discharge was found [Figures 1 and 2].

There was no bleeding or induration. There was no regional lymphadenopathy. Per rectal and vaginal per speculum examination was normal.

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Incisional biopsy was done from the vulval lesion, and on histopathological examination, there was full-thickness dysplasia of the epidermis with the presence of polygonal-to-oval cells with ground-glass cytoplasm, pleomorphic vesicular nucleus, and prominent nucleoli called Paget’s cell in the epidermis [Figures 3 and 4]. A diagnosis of EMPD was made.

She was further investigated to find out the presence of any underlying malignancy. Cervical smear, chest X-ray, and ultrasonography of the abdomen and pelvis were normal. Computed tomography scan of the abdomen and pelvis showed a heterogeneously enhancing lesion in the left vulva with metastatic left inguinal and external iliac lymph node. She was referred to surgical oncology where radical left vulvectomy with left groin dissection, laparoscopic left inguinal lymph node dissection, and perineal V-Y plasty repair were performed.

Histopathological examination of the excised specimen showed the presence of an invasive adenocarcinoma in the dermis with neuroendocrine features. A diagnosis of secondary EMPD of stage T<sub>1b</sub>N<sub>2c</sub>M<sub>0</sub> was finally made. Adjuvant radiotherapy was given after wound healing.

**DISCUSSION**

EMPD is a marginated plaque resembling Paget’s disease clinically and histologically but occurs in the apocrine-rich areas such as penis, scrotum, vulva, perianal area and axilla. It occurs more commonly in women starting at the fifth decade of life or thereafter. Two types of EMPD have been identified: primary (intraepithelial adenocarcinoma) and...
secondary (intraepithelial spread of adnexal or visceral carcinomas).\cite{2}

The common presenting features include severely pruritic, bleeding, oozy, tender, reddish eczematous lesion. Most often, it is nonspecific and can mimic many other dermatoses, such as Bowen’s disease, tinea infections, eczema, mycosis fungoides, and psoriasis.\cite{4}

Diagnosis is made by histopathological demonstration of Paget’s cells, which are distinct intraepithelial mucus-secreting neoplastic proliferative cells with abundant pale staining cytoplasm and large atypical nuclei. Sialomucin, a nonsulfated acid mucopolysaccharide, is found in the cytoplasm of Paget’s cells that stains with periodic acid–Schiff, colloidal iron, Alcian blue, and mucicarmine E and diastase resistant. Immunohistochemistry shows Paget’s cells positive for cytokeratin (CK) 7, carcinoembryonic antigen (CEA), and CK20 and negative for S100.\cite{3,5}

Treatment of choice for EMPD is surgery with recurrence rate of up to 44% with wide local excision. Other modalities include Mohs micrographic surgical excision, radiotherapy, topical agents, including 5-fluorouracil, bleomycin, imiquimod, and photodynamic therapy.\cite{1,6}

Primary EMPD has good prognosis, but the chances of recurrence are high. In secondary EMPD, prognosis depends on the underlying carcinoma. Clinical evidence of nodulations, lymph node enlargement, and dermal invasion indicate poor prognosis.\cite{2}

The significance of this disease lies in the fact that the cutaneous lesion may be an indicator of underlying malignancy.

Any elderly female presenting with nonspecific perineal lesion not responding to routine treatment, EMPD should also be kept in mind and should be confirmed by biopsy and immunohistochemistry. Further, underlying carcinoma if present has to be identified and treated accordingly.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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