Recurrent Extramammary Paget’s disease of Axilla: A case report

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ABSTRACT
Extramammary Paget’s disease of axilla is extremely rare and has not been well reported. It can be confused with eczema, superficial spreading melanoma or contact dermatitis. These patients can have an underlying malignancy. We report one such case in a 65 year old female who presented with recurrence of the lesion after 6 years.

Keywords: Extra-mammary Paget’s disease, Axilla, Recurrence, Eczema, Malignancy.

INTRODUCTION
Extramammary Paget’s disease is a rare, slow growing disease mostly observed in areas with numerous apocrine or eccrine glands. It presents as a well-demarcated, thickened, pruritic, erythematous, or white scaly plaque with irregular borders.2 It is usually an intraepithelial adenocarcinoma found outside of breast and characterized by a chronic eczema-like rash of the skin commonly seen around the anogenital regions like vulva, penis, and scrotum. Unlike Paget’s disease of breast, which is almost always associated with underlying in-situ invasive malignancy,2 extramammary Paget’s disease is associated with underlying invasive malignancy in about 10% of cases. Extramammary Paget’s disease of axilla has been reported only rarely. We are presenting one such case.

CASE REPORT
A 65 year old postmenopausal female reported with history of eczema and itching in her left axilla. It was associated with scancy serous discharge on and off. There was no other positive history related to this lesion. A local physician offered anti-allergics and calamine lotion with no relief. On examination there was a 7×8 cm hyperpigmented eczematous lesion. No breast lump or axillary lymph nodes were detected. Examination of anogenital region did not reveal any lesion. A punch biopsy from the lesion was taken and was associated with underlying in-situ invasive malignancy. The biopsy specimen revealed Paget’s disease. She underwent wide local excision of the lesion with primary closure. Histopathological evaluation of the excised specimen revealed epidermal acanthosis with atypical vacuolated cells in basal epidermis and adnexal epithelium with underlying dermal chronic inflammation (Figure 1a and 1b), suggestive of extra mammary Paget’s disease. Circumferential & deep resection margins were free. The slides were reviewed by a second pathologist who confirmed the same diagnosis.

On further history taking she recalled having a similar eczematous lesion at the same site which was excised more than six years ago. Tissue blocks and histopathological slides of this excision were retrieved which also revealed Paget’s disease. Mammography of both breasts was normal. Currently the patient is on follow-up and is disease free.

DISCUSSION
Extramammary Paget’s disease (EMPD) was first described by Crocker in 1889 involving scrotum & penis.16 EMPD is rare and occurs in apocrine-rich areas of skin and is extremely rare in the axillary region where it is not well documented. The origin of Paget’s disease is uncertain. It may be an intraepidermal malignancy with secondary extension to adjacent structures (intra-epidermal theory) or migration of tumor cells into the epidermis from an underlying carcinoma (epidermotropic theory). EMPD can be primary or secondary. The secondary type is usually associated with underlying carcinoma.39 In primary form of disease, skin adnexa (eccrine or apocrine glands), ectopic mammary glands, or pluripotential germinative cells in the epidermis and other structures have been implicated as a possible source of neoplastic cells. EMPD usually presents as an erythematous exudative dermatitis located on the vulva in women, the genitals in men, and the perianal area in both sexes. Extramammary Paget’s disease is associated with an internal malignancy in 50% of cases, usually carcinoma of the uterus, rectum, bladder, vagina, or prostate gland. Most of these cancers are usually related to the site of the dermatosis. The remaining is not associated with malignancy elsewhere, but has a high local recurrence rate and may become invasive. EMPD is seen most often in postmenopausal, older white women and is often mistakenly diagnosed as eczema, superficial spreading melanoma or contact dermatitis. Approximately 15% of the patients have
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an associated underlying adenocarcinoma of the apocrine or the bartholin’s glands. [3]

Axillary location of EMPD is extremely rare. Cheng et al. reported 7 cases of EMPD in axilla in a 20 year retrospective study. [6] It can occur as a solitary lesion or can be associated with involvement of anogenital region. Presence of an underlying carcinoma in axillary EMPD has been reported in 8 of the 23 cases (35%). EMPD has a propensity to recur locally and in our patient the lesion recurred six years after first excision.

The preferred treatment for Paget’s disease is wide local excision, with intraoperative microscopic control of margins. In view of frequent recurrences, multimodal approaches have been used. [7] Most commonly recurrences are treated with re-excision. In our patient, recurrence was treated with wide local excision with negative histological margins. Other less-established treatments for noninvasive Paget’s disease include topical chemotherapy, topical immune modifiers such as imiquimod, and photodynamic therapy. Radiation therapy and chemotherapy have also been used, but are generally reserved for patients with recurrent or invasive disease.

CONCLUSION

Patients with Extramammary Paget’s disease of axilla should be followed up closely after surgical excision for any recurrence or development of any malignancy.

ACKNOWLEDGEMENT

Nil.

CONFLICT OF INTEREST

Nil.

REFERENCES


Figure 1 (a): Photomicrograph (Low Power) showing epidermal acanthosis with atypical vacuolated cells in basal epidermis and adnexal epithelium with underlying dermal chronic inflammation.

Figure 1 (b): High Power of same.