

EXTRAMAMMARY PAGET'S DISEASE OF THE INGUINUM: A CASE REPORT

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Extramammary Paget's disease (EMPD) is a rare intra-epithelial malignancy that is occasionally associated with an invasive adenocarcinoma component as well as other secondary cancers. The diagnosis is confirmed by the presence of Paget's cells on histopathological examination of a tissue specimen. The standard treatment method of inguinal EMPD, as well as EMPD in other areas, is surgical resection. We present a patient with extra-mammary Paget's disease, who developed the erythematous eruption in the right inguinal region.

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Key words: *Extramammary Paget's disease, inguinum, diagnosis, treatment*

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Introduction

Extramammary Paget's disease (EMPD) is a rare intra-epithelial malignancy that is occasionally associated with an invasive adenocarcinoma component as well as other secondary cancers (1). EMPD can generate from all areas characterized by a high density of apocrine glands, as the axilla, inguinum, anus and perianal region, vulva in women, penis and scrotum in men. The lesion is located deep within the dermal to epidermal layer, and underlying carcinoma of the sweat glands has been suggested as a possible etiology (2).

EMPD should be differentiated from benign papulosquamous diseases, squamous cell carcinoma, and melanoma. Very often, it is mistaken for eczema or contact dermatitis. The diagnosis is confirmed by the presence of Paget's cells on histopathological examination of a tissue specimen. The standard treatment method of inguinal EMPD, as well as EMPD in other areas, is surgical resection (1).

We present a patient with extramammary Paget's disease, who developed an erythematous eruption in the right inguinal region.

Case report

A 75-year-old male presented with a 8-month history of recurrent, itchy, eczematous erosion, and indurated patchy lesion in right inguinal area (Figure 1a). The patient had been initially treated for an erythematous, pruritic lesion in the inguinum with topical corticosteroids. He had been treated with the steroids for 7 months. On physical examination, there was a centrally eroded, scaly erythematous lesion, 9 x 11 cm in diameter, on his right inguinal area, spreading to the scrotum and base of the penis.



Figure 1a. Skin lesions in the right inguinal area

No lesions were seen in the other apocrine-bearing regions, including breasts, perianal area, or external auditory canals. Superficial lymph nodes were not palpable in the inguinal regions. The results

of complete blood counts and blood chemistry examinations, including carcinoembryonic antigen (CEA), were normal. There was no evidence of internal malignancy on clinical, X-ray, abdominal and pelvic MSCT examinations.

The patient underwent wide excision of the skin lesion with a 2-cm margin to the macroscopic normal tissue, and primary closure. Surgical margins were histopathologically negative and the wound healed without complications. (Figure 1b).



Figure 1b. Final result of the previous case

Histopathological examination of a biopsy specimen of the skin lesion revealed an infiltration of the epidermal-dermal junction by Paget cells, that are atypical glandular-type cells, larger than the adjacent keratinocytes and with fine granular amphophilic to basophilic cytoplasm. The cytoplasm is paler than that of adjacent keratinocytes. The nucleus is oval with one or more prominent nucleoli. (Figure 2).

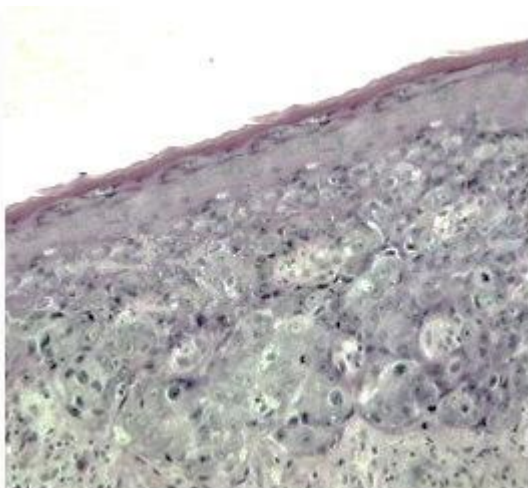


Figure 2. Histology of the excised specimen

Although the surgical margins were negative for malignancy, lymph nodes from the inguinal, external iliac, internal iliac and obturator regions were negative for malignancy by ultrasound (US) and MSCT examination, and postoperative radiotherapy and chemotherapy were not recommended.

During the one-year follow up, local recurrence and metastasis on US and MSCT examinations were not present.

Discussion

Extramammary Paget's Disease is a rare neoplastic lesion. The lesion generally appears as eczema and the most frequently reported symptom is the itch. Because of these poor clinical features, there is usually a delay in its diagnosis, based on the typical biopsy histological pattern. (1).

The most common site of EMPD is the vulva, and more than 200 cases have been reported. The second most common site is the perianal region, with more than 86 cases documented (3). Other locations include the perineum, scrotum, axilla, and eye lids. We present a patient with rare inguinal EMPD. EMPD generally occurs between the ages of 50 and 80, most frequently in Caucasians. In Japan, men are affected with extra-mammary Paget's disease twice as often as women (4), whereas women are predominantly affected in western countries (2).

The symptoms of the disease are not specific. Most patients report itching, burning, and soreness. A small number of patients may be asymptomatic. The presence of inguinal pain, bleeding, and tumor formation are reported to be more common in patients affected by invasive disease. The signs and symptoms are skin lesions, often mistaken for eczema, that may be itchy or painful (1). Our patient also had non specific symptoms and had been initially treated for an erythematous, pruritic lesion in the inguinum with topical corticosteroids for 7 months.

Extramammary Paget's disease is usually seen in isolation and is associated with an underlying invasive malignancy in about 12% of the cases. It is associated with an underlying adnexal malignancy in about 24% of the cases (5). It has a good prognosis in absence of malignancy, but may result in a poor quality of life because of frequent recurrences with the necessity of ablative therapies and anxiety about possible cancerization. Rarely, EMPD can be invasive or associated with adenocarcinoma or other kinds of cancer (6). We did not find any underlying malignancy after abdominal MSCT examination.

In conclusion, inguinal EMPD is a rare disease, as has been described in the currently available literature. It is usually associated with adnexal and visceral malignancies and generally has a poor prognosis due to its progressive course if not treated early. Early biopsy is very important for correct diagnosis in patients who fail to respond to conventional topical therapy. The standard treatment of inguinal EMPD is surgical resection.

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Prikaz bolesnika

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EKSTRAMAMARNA PAGETOVA BOLEST INGVINUMA

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Ekstramamarna Pagetova bolest (EMPB) predstavlja retki intraepitelni maligni tumor koji je povezan sa invazivnim adenokarcinomima, kao i sa drugim sekundarnim karcinomima. Dijagnoza se potvrđuje prisustvom Pagetovih ćelija na histopatološkom ispitivanju uzoraka tkiva. Standardna metoda lečenja za preponsku EMPB, kao i za EMPB u drugim područjima, je hirurška resekcija. Predstavljamo bolesnika s ekstramamarnom Pagetovom bolešću, koji je razvio eritematozne erupcije u desnoj ingvinalnoj regiji.

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Ključne reči: ekstramamarna Pagetova bolest, ingvinum, dijagnoza, tretman