VULVAR PAGET'S DISEASE - A CLINICAL CASE

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Introduction
Paget's disease of the vulva is a rare epithelial lesion located in the epidermis, representing 1% to 5% of vulvar neoplasies. It affects predominantly caucasian women at the age of menopause. Common presenting symptoms are itchiness and vulvar discomfort. Macroscopically, the lesion presents itself as an eczema and begins from the vulvar areas covered with hair. In 10% to 20% of the cases, it is associated with an underlying invasive carcinoma, especially adenocarcinomas. The treatment of Paget's disease can be local surgical excision or extended vulvectomy, according to the histological findings and the extent of disease. Inguinal lymphadenectomy should be performed in cases of underlying vulvar adenocarcinoma. The rate of recurrence is high and can occur years after treatment.

Objectives
Description of a clinical case of Paget's disease of the vulva.

Materials
Retrospective revision of the patient's clinical profile.

Methods
Retrospective revision of the patient's clinical profile.

Results
An 82 years old woman presents to the outpatient clinic with a complaint of an itchy lesion in the vulva for the last three months. Vulvoscopy was performed and showed a well-demarcated eczematosus lesion, with slightly raised edges and a background of unpolished ulcerations and opacities, in the inferior half of the right labia majora. Biopsy was realized revealing an Extramammary Paget disease in the epidermis without stromal invasion.

The patient underwent a radical vulvectomy with skin grafting and lymph node dissection. Histological results revealed Vulvar Paget's disease without metastases in the regional lymph nodes.

Two years later, on a follow-up consultation, she complained of vulvar itchiness with scanty bleeding over the operated site. On gynecological examination, the skin over the vulva region was inflamed. Biopsy of the vulva was carried revealing images consistent with Paget's disease in the epidermis. The patient was then subjected to a simple vulvectomy with transposition of skin grafting. Histopathology confirmed intra-epidermal Paget's disease of the vulva without stromal invasion or glandular neoplasia in the depth of the graft, with the tumor positioned 4 mm away from the surgical plane. She was discharged from the hospital and is under surveillance by the outpatient gynecology service.

Conclusions
Paget's disease of the vulva a rare neoplasy. It affects predominantly postmenopausal caucasian women. Common presenting symptoms are pruritus and vulvar discomfort. Clinically, the lesion presents itself as an eczema and starts from the hair covered vulvar areas. According to histological findings and extent of disease, the treatment of Paget's disease can be either local surgical excision or extended vulvectomy. Inguinal lymphadenectomy should be performed in all cases of underlying vulvar adenocarcinoma. The rate of recurrence is high and can occur years after treatment.

References: