Pruritus Ani in an Elderly Man

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REPORT OF A CASE

A 78-year-old man presented with a 6-month history of perianal pruritus. His history was noncontributory. His father died of colon carcinoma. Previously, at another hospital, the diagnosis of eczema with possible fungal infection was suggested, and he was treated with topical corticosteroids and antifungals without significant improvement. The patient noted an indurated, damp, painless, and mildly pruritic lesion in the perianal area several weeks earlier.

There was an erythematous, slightly elevated, exudative, well-defined plaque with irregular borders (diameter, 4 cm), located in the intergluteal fold near the anus. Around that lesion a symmetrical, erythematous, and slightly desquamative plaque was apparent (Figure 1). Culture was positive for Candida albicans. Topical treatment with an imidazole was begun to limit the extension of the larger lesion. A biopsy specimen of the exudative plaque was stained with hematoxylineosin and is shown in Figure 2.

What is your diagnosis?

HISTOPATHOLOGIC FINDINGS AND CLINICAL COURSE

The epidermis appeared hyperkeratotic and slightly acanthotic. Numerous large cells with abundant palestaining cytoplasm, dispersed singly or in clusters, were present at all levels of the epidermis. These Paget's cells stained positively with periodic acid-Schiff and colloidal iron, indicating the presence of both neutral and acid mucopolysaccharides. Carcinoembryonic antigen was identified in tumor cells with avidin-biotin-peroxidase.

Fifteen days later, the patient was again seen in our hospital with remarkable improvement of the erythematous lesion located in the intergluteal sulcus, but the slightly elevated plaque and two small well-delineated lesions extending to the perineum and scrotum persisted.

The digital rectal examination was normal; the prostate was slightly indurated. The prostate-specific antigen level was 3.2 µg/µL (normal, 0 to 2 µg/µL). Colonoscopy was performed and two tubular adenomatous polyps were resected. Diverticula were present

in the sigmoid and descending colon. The appearance of the rectal mucosa was normal. An intravenous pyelogram was without pathology.

Five weeks after his first visit to our center, he underwent resection of all cutaneous lesions with a 1-cm margin of apparently healthy skin and reconstruction of the defect by local flap. The resection margins were histologically negative.

DISCUSSION

Extramammary Paget's disease is an extremely rare condition, usually presenting between the sixth and eighth decades, with a slight female predominance. The disease occurs most frequently in the anogenital area, but has been found in the axilla, oral cavity, external auditory meatus, cheek, eyelid margin, and umbilicus. There is a high incidence of associated cancers with extramammary Paget's disease. A recent review of the literature reported a 29% incidence of associated internal malignant disease.[1] In several studies, the most common underlying malignant lesion found was transitional cell carcinoma of the bladder.[2,3]

The pathogenesis of extramammary Paget's disease is variable. In some cases, it represents an in situ malignant transformation of the intraepidermal component of the sweat duct.[4] It is suggested that the Paget cell in the epidermis originates from embryologically placed ectodermal apocrine-type secretory cells that become altered and visible under carcinogenic stimulus.[5] Often, Paget's disease appears to be an epidermotrophic metastasis from an associated sweat gland carcinoma.

Paget's disease of the eyelid is associated with carcinoma of Moll's gland, and Paget's disease of the external auditory meatus is associated with ceruminous gland carcinoma.[6] Occasionally, the lesion is due to epidermotrophism from a distant malignant neoplasm, such as a carcinoma of the rectum or endocervix. In the case of anal involvement, the tumor can originate from mucussecreting cells of the rectum. Perianal Paget's disease is associated with carcinoma in as many as 73% of cases.[7,8] In the literature, data are variable; Helm et al3 found that only one of eight patients with perirectal extramammary Paget's disease had an underlying adenocarcinoma. In our patient, after clinical and endoscopie explorations, the diagnosis of the associated adenocarcinoma was dropped.

The following clinical subtypes can be distinguished in extramammary Paget's disease, besides purely intraepithelial disease: colorectal carcinoma, urinary bladder carcinoma, prostatic carcinoma, and endometrial and cervical carcinoma. Surgery is indicated. Intraoperative histologie examination of margins is ideal.

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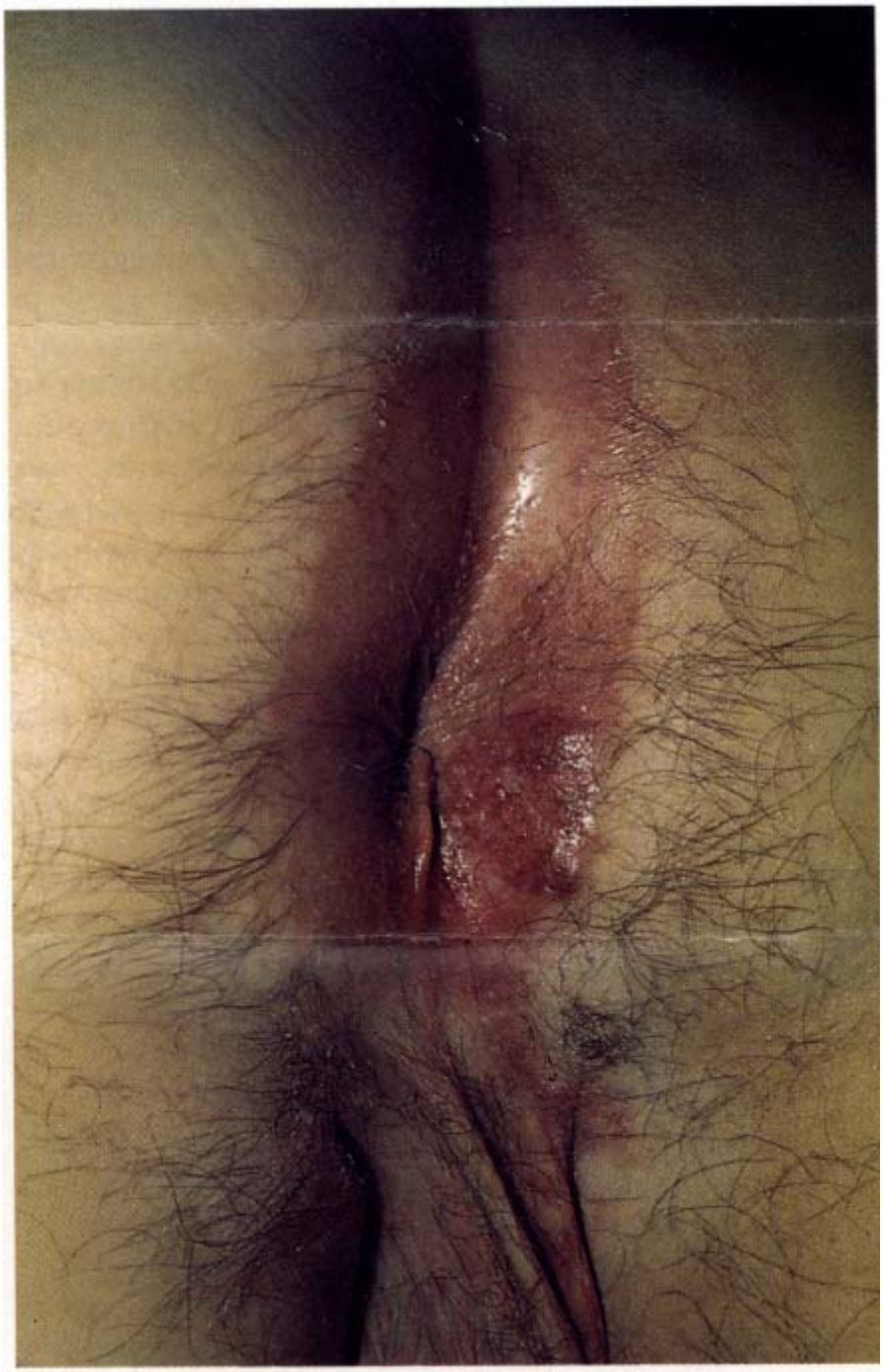

Figure 1

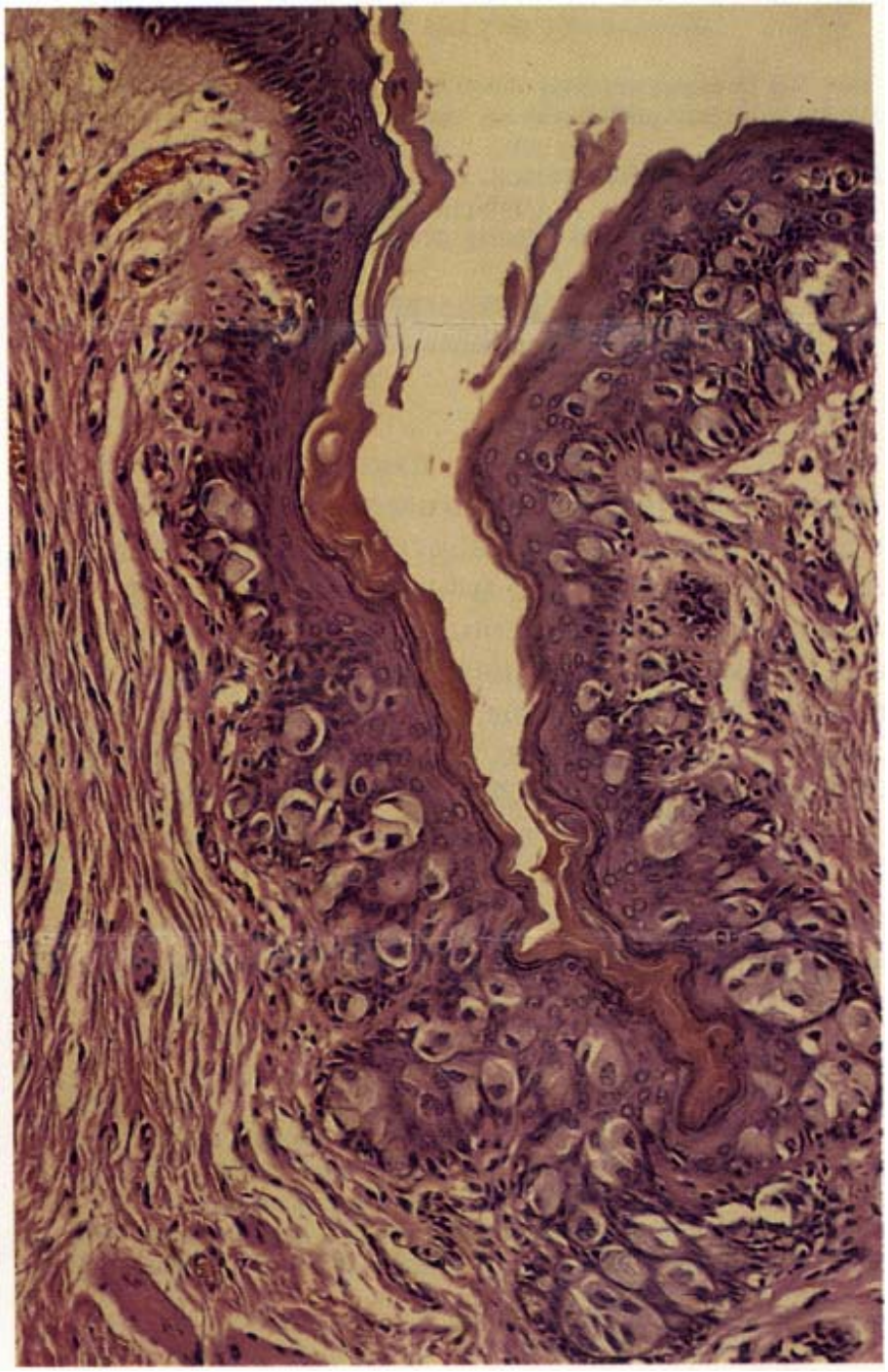

Figure 2