

A Case of Extramammary Paget's Disease on the Scalp

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We treated a case of extramammary Paget's disease that affected the scalp of a 45-year-old female. It is extremely rare that the disease arises in areas other than the anogenital region and the axillae. The lesion was a round erythematous oozing crusted hairless patch. Histopathologically, many pagetoid cells were found within the epidermis and dermis. The cytoplasm of these cells stained with alcian blue at pH 2.5, CEA, EMA and low-molecular-weight-cytokeratin. The patient underwent a wide local excision. We used a mapping technique to reveal the distribution of microscopically involved lesions. (*Ann Dermatol* 11(3) 189-192, 1999).

Key Words : Extramammary Paget's disease, Mapping technique

Extramammary Paget's disease is an epithelial disorder usually observed in cutaneous apocrine gland-bearing regions. It most commonly affects the anogenital region and less frequently arises in the axillae. The occurrence of primary extramammary Paget's disease in other areas is exceptional. There are only a few reported cases in which the disease occurs at locations such as the buttock, lateral aspect of the back and lower portion of the chest¹.

CASE REPORT

A 45-year-old woman was first seen at our clinic in January 1996 because of oozing eczema on the scalp. About 5 months ago, she noticed a small round hairless patch at the site. The lesion had been hairless and eczematous and had gradually

increased in size. She was treated with antibiotics but the lesion did not improve. A physical examination revealed a round erythematous oozing crusted hairless patch measuring 5 cm in diameter (Fig. 1). No enlarged lymph nodes were found in her cervical, axillary, inguinal region. No lesions suggestive of extramammary Paget's disease were detected in her anogenital region or axillae. Her general condition was good and the results of routine laboratory investigations, including complete blood cell counts, blood chemistry studies and urinalysis, were within normal limits. No organisms were found on bacterial smears and culturing. The nuclei of the pagetoid cells were compressed to the periphery of cells, resembling so called signet ring cells. A histological examination of a punch biopsy of the lesion revealed many pagetoid cells within the epidermis and dermis (Fig. 2). The cytoplasm of these cells was stained with alcian blue at pH 2.5 (Fig. 3), carcinoembryonic antigen (CEA), epithelial membrane antigen and low-molecular-weight cytokeratin. With S-100, HMB-45 and vimentin, the cells were not stained. We considered the possibility of metastatic adenocarcinoma because of the signet ring cell appearance of the pagetoid cells and the dermal invasion of pagetoid cells and then practiced the following studies. The serum carcinoembryonic antigen, alpha-fetoprotein and carcinoma antigen-125 were within the

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Fig. 1. 5 × 6 cm sized oozing yellowish crusted hairless patch on the scalp.

Fig. 3. Paget cells are stained positively with alcian blue at pH 2.5.(× 100).

normal range. In mammography, gastroduodenoscopy, sigmoidoscopy and liver ultrasonography, no abnormal findings were detected. The patient underwent a wide local excision with a 2 cm resection margin by plastic surgery. We used a mapping technique to reveal the distribution of microscopically involved lesions(Fig. 4). She revisited our clinic 2 years later complaining of an oozing lesion at the margin of the previous lesion. A punch biopsy of the oozing scalp lesion confirmed the recurrence of the tumor.

Fig. 2. Numerous pale staining, large, rounded Paget cells nearly replacing the epidermis.(H & E, × 100).

Fig. 4. Diagram showing distribution of extramammary Paget's disease. The outline of the diagram is a resection margin of the lesion.

DISCUSSION

Lesions of extramammary Paget's disease are usually found in sites where there is a high density of apocrine glands. These include the vulva, penis, scrotum, anus, anal margin, perianal region and axillae. Other lesions where apocrine sweat glands (or modified apocrine sweat glands) are located have also been sites of extramammary Paget's disease. These include the eyelids and external ear canal¹. The term "ectopic" extramammary Paget's disease has been applied to those cases in which the disorder occurs at locations where apocrine glands are not usually found such as the buttock, lateral aspect of the back, or lower portion of the chest². In 1994, Sai *et al* reported a case of extramammary Paget's disease that occurred on the scalp of a 81-year-old Japanese

man³. Mammary Paget's disease is said to emanate from the "migration" of neoplastic cells into the epidermis from an underlying carcinoma of the breast, whereas extramammary Paget's disease is thought to originate within the epidermis by the proliferation of primitive native stem cells with the potential for glandular differentiation⁴.

Morphologically, lesions of extramammary Paget's disease present as sharply demarcated, infiltrated, erythematous to gray-white plaques, which are occasionally eczematoid, crusting, scaling, papillomatous, or, rarely, ulcerated. The lesions may be asymptomatic. However, pruritus, a burning sensation and pain may be noted⁵. Lesions are often treated with a variety of topical medicaments intended for more banal conditions. The duration of the lesion or symptoms from onset until the time of diagnosis may range from 1 month to 31 years⁴. In our case, the patient was treated for an infectious disease at first and then treated with oral and topical antibiotics for about 5 months at a local clinic. The diagnosis was not confirmed until the skin punch biopsy was taken from the two sites of the lesion. The diagnosis of extramammary Paget's disease should be considered when lesions showing eczematoid conditions do not heal within 1 month of topical therapy¹. A biopsy will confirm the diagnosis.

Histopathologically, the diagnosis of extramammary Paget's disease is confirmed by the presence of Paget cells on routine hematoxylin-eosin histological sections. Paget cells are round cells with abundant pale staining cytoplasm and a large, central, reticulated nucleus⁶. Extramammary Paget's disease must be differentiated from Bowen's disease and from superficial spreading malignant melanoma. Although extramammary Paget's disease will stain positively for low-molecular-weight cytokeratins and CEA, it will be S-100 negative, Bowen's disease will stain positively for high-molecular-weight cytokeratins, and superficial spreading malignant melanoma will be S-100 positive and CEA negative⁷. The Paget's cell nucleus is often compressed to the periphery of the cell by the cytoplasm, giving a signet ring appearance. Paget cells may involve not only the pilary outer root sheath epithelium but be found in sebaceous glands and in structures of apocrine and eccrine glands as well. Paget cells can proliferate and disrupt the basement membrane and extend into the dermis. In

our case, Paget cells showed signet ring cell appearances and were found in the epidermis, pilary outer root sheaths, apocrine glands and dermis. The outline of histologically involved areas was highly irregular and multiple separate foci of disease were present. From serial sections of total excised specimens, we made a diagram showing irregular margin and skip areas between the Paget's lesions.

Extramammary Paget's disease remains in situ within the epidermis and adnexal epithelium in two-thirds of the cases. The treatment of choice for extramammary Paget's disease is surgical excision. However, the frequency of local recurrence after surgical excision is a significant problem in the treatment of this disease. Mohs micrographic surgery, which has been found to be quite effective, has the advantage of sparing uninvolved tissue by the mapping technique. The dissection of regional lymph nodes need not be undertaken if extramammary Paget's disease is confined to the epidermis and adnexal epithelium and if a search for underlying internal cancer is negative. When Paget's cells involve the dermis, however, metastases may and do occur, and some investigators advocate regional lymph node dissection in these cases⁴.

In conclusion, our case was extramammary Paget's disease that had developed on the scalp and was treated as infectious dermatitis for 5 months. The diagnosis of the extramammary Paget's disease should be considered when eczematous lesions are unresponsive to various medications prescribed elsewhere. Because the extent of histologically demonstrable disease is far greater than that of the visible lesion, a skin biopsy from more than two sites of the lesion is recommended.

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