Perianal Paget’s Disease *

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STOUT believes that extramammary disease is a peculiar skin lesion that is clinically and microscopically similar to Paget’s disease of the nipple. Microscopic examination of the lesion reveals large, rounded, vacuolated, eosinophilic “pagetoid” cells in the epithelium, extending from carcinoma of the underlying glands. The underlying tumor is adenocarcinoma of the apocrine glands. Owing to the diversity of hypotheses regarding the nature of extramammary Paget’s disease, it is a controversial subject and apparently it will remain controversial until the histogenesis of the Paget cell is solved. At present there are two schools of thought regarding the origin of the disease. Some maintain that it is not associated with deep cancerous tumors but that it begins in the epithelium and invades the underlying tissues. According to Montgomery, the epidermal lesion and the Paget cells are typical, can be recognized easily and there is not the slightest involvement of the corium. He claims that it presents a widespread multicentric malignant process entirely limited to the epidermis and believes the cells develop in situ and are precancerous. Other authorities believe that extramammary Paget’s disease is a panepidermotropic cancer migrating into the epidermis from the deeper layers with no transition between normal epithelial cells and the Paget cells.

Extramammary Paget’s disease may be difficult to diagnose clinically because of the varying duration and degree of secondary infection presenting an innocuous appearance similar to various forms of eczema. The diagnosis can be established correctly by adequate biopsy, preferably by excision of the involved skin with its apocrine glands such as those of the axillae, scrotum, perianal tissues, perineum and vulva. By histopathologic study Paget’s disease may be differentiated from hyperkeratosis, hyperplasia of the basal cell layer of the skin with chronic dermatitis, leukoplakia, Bowen’s disease and invasive carcinoma.

According to Willis, Paget’s and Bowen’s disease may resemble each other microscopically especially since the round, pale or vacuolated cell of Bowen’s disease is indistinguishable from the Paget cell. He also reports that Paget’s long-standing balanitis ending in cancer, Paget’s disease of the genitalia and Bowen’s disease of the same regions are no more than variants of the same disease. He feels that these should be considered one entity without different names and should be accepted as intraepithelial cancer and not precancerous lesions. The terminology of carcinoma of the apocrine glands of the anal region and other intra-epidermal tumors in situ should be changed. The term extramammary should be abandoned and Paget’s disease of the anus, scrotum, penis and vulva should be used instead.

In 1937, a review of 58 cases supposed to be extramammary Paget’s disease was reduced to 14 by Weiner and two years later this number was again reduced to nine by Pinkus and Gould.

The earliest case on record involved the perianal tissues, the perineum and the

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scrotum and was reported by Darier and Couillaud\(^3\) in 1893. In 1949 the author\(^6\) reported a case in which the perianal tissues and the sacral, coccygeal and gluteal areas were diseased (Fig. 1). Microscopic examination revealed no involvement of the transitional epithelium lining the walls of the anal canal. It was of "status simplex epiderma" origin in which there was a downward growth of overlying epidermis and superficial invasion by squamous cells exhibiting many masses of Paget cells. This condition could be classified as Bowen's dermatose which Sequeira and Turnbull\(^9\) believe to be a variety of Paget's disease (Fig. 2, 3, 4).

**Case Report**

A 56-year-old woman with a negative medical and surgical history was examined in 1946. She complained of mild itching in the perianal area beginning on the inner surface of the right buttock at the level of the coccyx and in five months it had extended to the opposite side. There was a serosanguineous exudate. The involved area measured 14 by 8 cm., was circinated with sharply demarcated advancing borders of a light amber hue which were elevated about 2 mm. The excoriated areas of the intergluteal surfaces were clearly outlined and contained moist, granular, scarlet verrucae which were ulcerated and elevated about 4 mm. above the skin level. Flat-fingerlike projections having dry pink and flaky surfaces extended peripherally from the edges of the excoriated areas. These projections tapered gradually and blended to within one half to 1 cm. of the advancing border of the entire skin lesion. The perianal border of the lesion encircled the posterior half of the external anal margin. Sigmoidoscopic examination was negative. Nine months after onset a radical excision was performed. The skin of the posterior half of

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Fig. 1. Paget's disease of the perianal areas.

Fig. 2. Low-power magnification of lesion in Figure 1.

Fig. 3. Medium-power magnification of lesion in Figure 1.

Fig. 4. High-power magnification of lesion in Figure 1.