Multicentric pigmented Bowen's disease

A report of 2 cases

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Summary

Multicentric pigmented Bowen's disease is a rare form of intra-epidermal carcinoma characterised by its pigmented appearance as well as its confinement to the perineal region. Two examples of this disease are presented, illustrating the classic features of this disorder.

Bowen's disease is an intra-epidermal carcinoma often found in white patients and presents as a slowly enlarging erythematous plaque which may be found anywhere on the body. A rare variant is multicentric pigmented Bowen's disease (MPBD). This, unlike classic Bowen's disease, is characterised by its pigmented appearance as well as its confinement to the genitocutaneous region. Two cases are presented which apparently reflect the first report of this condition in southern Africa and its first appearance in black patients.

Case 1

A 28-year-old coloured woman presented with a pigmented lesion immediately posterior to the anus. The lesion had first appeared 5 years previously as two small, warty plaques measuring about 1 cm in diameter. A diagnosis of condyloma acuminata was made, and podophyllin applied. The lesion was, however, still present 2 months later, but was now indented. Infective dermatitis was then diagnosed, and a topical corticosteroid as well as an oral antibiotic prescribed. The condition nevertheless persisted for 4 years until the patient was referred to a dermatologist for biopsy of a 'possible malignant melanoma'. On examination, the patient had a pigmented, non-indurated plaque in the anal crease posteriorly (Fig. 1). The lesion consisted of two coalescing plaques measuring about 2 x 4 cm. It was red with numerous areas of black and brown pigment and had a velvety, slightly verrucous surface. There were no overt signs of internal malignant disease. Histological examination revealed the features of Bowen's disease — intra-epidermal carcinoma with numerous atypical periodic-acid-Schiff (PAS)-positive, diastase-labile cells showing loss of normal polarity. In addition, unlike classic Bowen's disease, a moderate amount of pigmentation was present within the epidermis and in the upper dermis. This confirmed the clinical diagnosis of MPBD.

Case 2

A 34-year-old black woman presented with a 2-year history of a pigmented asymptomatic lesion in the anal cleft. There was no history of arsenic exposure. The lesion consisted of a large plaque 2 cm in diameter and two smaller satellite lesions. The surface was slightly warty, and was strikingly and variably pigmented. The patient had no other cutaneous or systemic disorders. Histological examination of the excised lesion (Fig. 2) showed the classic features of MPBD — intra-epidermal carcinoma containing numerous atypical PAS-positive, diastase-labile cells and keratinocytes lying in total disarray, and heavy deposits of pigment within the epidermis as well as in the papillary and reticular dermis.

Discussion

MPBD was first described by Lloyd in a 22-year-old Yemenite male. By 1977, 13 cases had been reported, of which 10 were in females. These 13 cases included 7 Japanese, 1 Yemenite, 1 Lebanese, and 2 white Americans. Since its first description, the condition has been reported under several titles — multicentric Bowen's disease of the genitalia, multicentric Bowenoid acanthoma, reversible vulvar atypia, Bowenoid atypia of the vulva, and Bowenoid lesions with spontaneous regression. MPBD is characterised by: (i) a
Fig. 1. Lesion in gluteal fold showing pigmented, velvety lesion.

younger age presentation than classic Bowen's disease; (ii) symmetrical lesions confined to the genitocrural region; (iii) black or brown-black flat or papular lesions; (iv) slow progression or occasional spontaneous regression; and (v) characteristic histological features.

Our 2 cases are typical examples of MPBD, and are further notable for their occurrence in black subjects. The classic non-pigmented form of Bowen's disease has been described as occurring in blacks, but here too it is extremely rare. Thus in 1944 Herold and Cooper wrote that Bowen's disease had not yet been reported in blacks. And by 1982 only 7 cases of the classic non-pigmented Bowen's disease had been reported in black people.

The age incidence of classic non-pigmented Bowen's disease in whites has been established as 60 - 69 years, which is the same as that in blacks. This age incidence is thus considerably higher than the 15 - 37-year age incidence of MPBD.

Case 1 clearly illustrates how easily MPBD may be misdiagnosed. Furthermore, because of their rarity in blacks, both classic Bowen's disease and MPBD are usually not even considered in the differential diagnosis.

Fig. 2. Photomicrograph showing intra-epidermal carcinoma with melanin pigmentation in epidermis and upper dermis.

REFERENCES