

Pigmented plaque on the perineum

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CLINICAL FINDINGS

A 62 year-old female patient presented with a solitary lesion on the perineum (Fig. 1) for one year duration. The lesion started as a flat asymptomatic small-sized lesion which gradually enlarged and became more palpable. During the last few months, the lesion started to be irritable. There was no past history of similar condition. The patient had a history of diabetes mellitus for the last 12 years and hypertension for the last 7 years. On examination, there was a solitary motley pigmented, well defined plaque that measured about 1.6 x 1.4 cm (Fig. 2). The surface was mostly verrucous while it was partially eroded and scaly on one side. There was no tenderness, induration or paralesional infiltration. Laboratory investigations revealed elevated blood sugar, high serum cholesterol and triglycerides in addition to low serum iron.

What is your clinical differential diagnosis?

Condyloma accuminata (genital wart), lichen planus, bowen's disease, bowenoid papulosis, basal cell carcinoma (superficial pigmented), extramammary Paget's disease, and malignant melanoma.

PATHOLOGICAL FINDINGS

The lesion was totally excised with 2 millimeter safety margins. The histological examination showed epidermal acanthosis, hyperkeratosis and



Fig. 1 A 62 year-old female patient presented with a solitary lesion on the perineum.



Fig. 2 A solitary motley pigmented, well-defined plaque, measured about 1.6 x 1.4 cm. surface is mostly verrucous while it was partially eroded and scaly on one side.

parakeratosis (Fig. 3). There were focal areas with disarrangement of keratinocytes in addition to the presence of numerous atypical keratinocytes arranged in disorderly fashion throughout the epidermis (Fig. 4). The dermis showed superficial perivascular mixed inflammatory infiltrate with melanin incontinence and a considerable number of melanophages.

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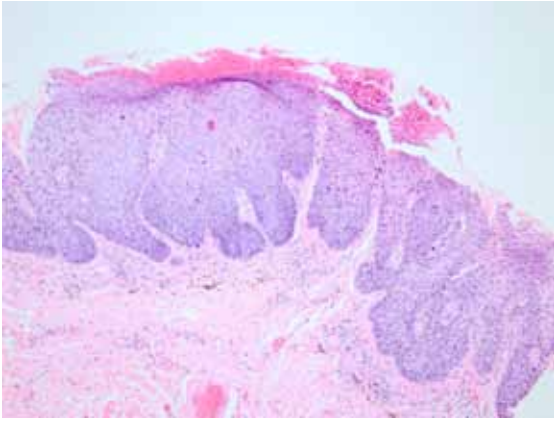


Fig. 3 Epidermal acanthosis, hyperkeratosis and parakeratosis with mild superficial perivascular lymphohistiocytic infiltrate admixed with numerous melanophages (H&E x40).

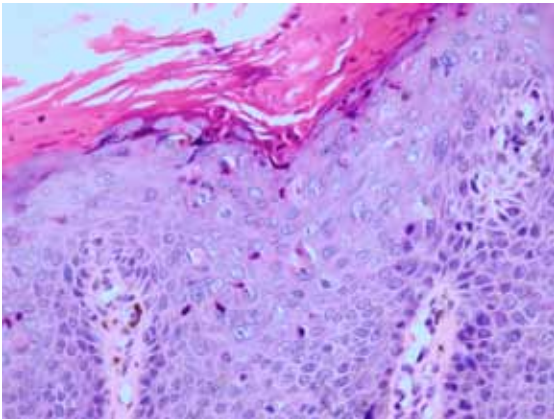


Fig. 4 Focal areas with disarrangement of keratinocytes in addition to the presence of numerous atypical keratinocytes arranged in disorderly fashion throughout the epidermis (H&E x400).

DIAGNOSIS

Bowen's disease.

COMMENT

Bowen's disease (BD) was firstly described by JT Bowen in 1912 as an in situ squamous cell carcinoma. The incidence of BD is rarely reported but the annual average rate among Americans was markedly variable from 14.9 to 142 cases per 100,000. Although BD could be transformed into invasive SCC, this is usually rare and not exceeding 5% but this incidence are higher (up to 30%)

in genital lesions. It is widely accepted that BD has no significant correlation with internal malignancies and it is more favored not to be classified as a paraneoplastic condition.¹⁻³

BD equally affects males and females with higher incidence among older age group more than 60 years. It is usually located on sun-exposed areas of white races but genital lesions may be higher in darker-skin individuals. Clinically, BD is usually presented with asymptomatic slowly enlarging erythematous patch or plaque which is characterized by a well demarcated borders and scaly or crusted surface. The lesion is usually solitary and located more on the lower limbs and head and neck. Atypical lesions of BD include atypical location such as periungual, subungual, palmar, genital and perianal forms, atypical number such as multiple form and atypical color such as pigmented form.^{4,5}

The etiology of BD is not yet clear, but several factors have been incriminated in the development of the lesions such as; radiotherapy, arsenic exposure, immunosuppression status (eg, after organ transplantation and AIDS) and viral infection. It has been reported that BD has a significant correlation with Human Papilloma virus (HPV) especially those lesions located on the perianal and genital lesion. Moreover, genital BD shows a higher risk of progression into an invasive carcinoma (10%) compared with extragenital lesions (3-5%).^{6,7}

The differential diagnosis of genital BD includes condyloma accuminata (genital wart), lichen planus, psoriasis, nummular eczema, tinea corporis, extramammary Paget's disease and malignant melanoma while BD in sun-exposed area could be differentiated from superficial or pigmented BCC, seborrheic keratosis and actinic keratosis (listed

The clinicopathological challenges of Bowen's disease

Diagnosis	Clinical	Pathological
Lichen planus	<ul style="list-style-type: none"> Presented with flat-topped violaceous papules covered by fine white lines Affect the flexor surface of the extremities, the trunk, and the genitalia May involve mucous membranes and nails 	<ul style="list-style-type: none"> Band-like lymphocytic infiltrate admixed with melanophages Vacuolar alteration of the basal layer Acanthosis with irregular elongation of rete ridges with 'saw tooth' appearance Wedge-shaped hypergranulosis Hyperkeratosis
Psoriasis	<ul style="list-style-type: none"> Presented with well-circumscribed erythematous patches with a silvery white scale Positive Auspitz's sign (bleeding points) More involvement of the extensor surfaces of the extremities 	<ul style="list-style-type: none"> Acanthosis with regular elongation of rete ridges Thin supra-papillary plate Diminished granular layer Mounds of parakeratosis Intracorneal collections of neutrophils (Munro micro-abscesses)
Condyloma accuminata	<ul style="list-style-type: none"> Fleshy exophytic lesion on the anogenital region May occur on the penis, vulva, and cervix 	<ul style="list-style-type: none"> Marked acanthosis, hyperkeratosis and papillomatosis Vacuolated cells and coarse keratohyaline granules in the granular cells
Actinic keratosis	<ul style="list-style-type: none"> Affect elderly, fair-skinned, sun-sensitive persons Arise in sun-exposed areas Presented as circumscribed red, scaly lesions May be hyperkeratotic or pigmented 	<ul style="list-style-type: none"> Basal dysplasia and architectural disorders of the epidermis Atypical keratinocytes with variable size and shape in the lower epidermis Thin granular layer, hyperkeratosis and focal parakeratosis
Extramammary Paget's disease	<ul style="list-style-type: none"> Presents as an erythematous, eczematoid, slowly spreading plaque Affects mainly anogenital region and less commonly the axilla There is a predilection for older individuals 	<ul style="list-style-type: none"> Epidermal hyperplasia with hyperkeratosis and parakeratosis The tumor cells have abundant pale cytoplasm and large pleomorphic nuclei The cells are arranged singly or in small groups, in the basal and parabasal regions
BCC	<ul style="list-style-type: none"> Common in sun-exposed areas of fair-skinned individuals More in elderly men Commonly presented with papulo-nodular lesion with telangiectasia May be ulcerative, pigmented or presented as atrophic plaque 	<ul style="list-style-type: none"> Nests of basaloid cells with peripheral palisade and attached to the undersurface of the epidermis. The tumor cells have a hyperchromatic nucleus with poorly defined cytoplasm There are numerous mitotic figures Peri-tumoral clefts are common

in the table). The characteristic clinical sign of BD is the asymptomatic lesion while the histological hallmark is the full-thickness atypia of the epidermis.⁸

The treatment of BD includes different modalities such as cryotherapy, curettage with cautery, surgical excision, 5-fluorouracil (5-FU), radiotherapy, laser, photodynamic therapy (PDT) and imiquimod in addition to other rare therapeutic options such as local hyperthermia and ultrasonic surgical aspirator. A new therapeutic option that was recently described in the treatment of BD was cyclooxygenase enzymes inhibitors (diclofenac 3%

gel) which showed successful result in the treatment of a few number of cases.⁹⁻¹¹

These different modalities suggested that there is no single definite treatment for BD and the treatment of choice should be guided by the efficacy, location and size of BD in addition to the number of lesions, availability of the therapy, the clinician expertise, patient factors (age, immune status and compliance), cosmetic outcome and the patient preference. Follow up for one year is recommended to evaluate the recurrence and protective measures such as sunscreens and photo-protective clothes should be stressed to these patients.¹²

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