Pigmented Plaque on Perineum

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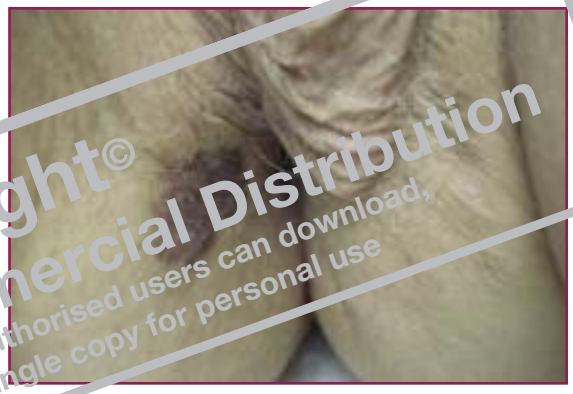
A 60-year-old female is referred to the dermatology clinic for assessment of a gradually enlarging lesion of two to three years duration, located on the perineum. She is otherwise healthy and is not taking any medications.



What is your diagnosis?

a. Superficial spreading melanoma
b. Psoriasis
c. Pigmented basal cell carcinoma
d. Pigmented Bowen's disease
e. Extramammary Paget's disease

Answer: Pigmented Bowen's Disease

Bowen's disease (BD) refers to cutaneous squamous cell carcinoma (SCC) in situ, a malignant tumour of keratinocytes.[1] It was first described in 1912 by John T. Bowen, an American dermatologist.[1] BD presents as a sharply demarcated, scaly plaque.

It is especially important to consider the diagnosis of BD for patients with risk factors such as significant sun exposure, HPV infection in their genital skin, and individuals with immune deficiency, especially transplant recipients and those with HIV infection. Other risk factors include arsenic, tar, and chronic radiation dermatitis.[1] BD affects males and females equally. It is found much more frequently in Caucasians than in black persons. The highest incidence is found in patients over 60-years-of-age.[1] BD can be located anywhere on the body. In males, lesions are more commonly found on the head and neck, while in females the lower limbs are the most common site. BD located on the glans penis or the mucosa of the labia is sometimes referred to as erythroplasia of Queyrat.[1]

The pigmented variant of Bowen's disease (PBD) is especially important, as it can often be confused with a seborrheic keratosis or psoriasis and therefore remains incorrectly diagnosed.[2] PBD is characterized by a hyperpigmented, sharply demarcated plaque with a velvety, verrucous, or flat surface. This variant is most likely to show up at perianal, flexural and, subungual sites.[1]

The rate of transformation of BD to invasive SCC is estimated to be between 3 and 5%. Of these, up to one-third may metastasize.[1] The risk of progression is higher on genital sites. The presence of pigment does not enhance malignant potential; therefore, PBD is treated similarly to non-pigmented BD.[3]

BD is often misdiagnosed due to morphological similarities it shares with a number of other skin diseases, including psoriasis, nummular eczema, and tinea corporis. It is important to keep in mind the possibility of BD in any isolated, psoriasis-like,

scaly plaque that fails to heal with topical therapy, especially one that develops for the first time in an older individual. Patients with actual psoriasis will often have evidence of psoriasis elsewhere, such as the elbows, scalp or nails.

The pigmented Bowen's variant is often confused with pigmented basal cell carcinoma, but can be distinguished clinically by the absence of arborized vessels, and a pearly translucent surface with rolled borders. Extramammary Paget's disease, a superficial cancer of apocrine sweat glands, may be hyperpigmented and commonly occurs in the perineum. Paget's disease differs, however, in that it will typically be moist and very red and may contain superficial erosions.

Melanocytic lesions such as congenital nevi and melanoma can present similarly to PBD. As opposed to PBD, malignant melanoma typically does not produce much scale. On examination with a dermatoscope, a hand-held instrument used in most dermatology offices, a pigment network with melanocytic lesions can be seen.[4] A definitive diagnosis made by biopsy is recommended.[5]

Treatment

Treatment of BD includes topical and surgical options. Topical chemotherapeutic agents such as 5-fluorouracil and imiquimod are effective options. Five percent topical fluorouracil applied b.i.d. for eight to nine weeks has shown clearance rates up to 92%.[6] A few small, randomized controlled trials have assessed the efficacy and safety of 5% imiquimod in the treatment of BD and erythroplasia of Queyrat.[6] One proposed dosing schedule of 5% imiquimod is application three times weekly for four weeks, followed by a gradual decrease in dosing frequency to complete an eight-week course of treatment.[6] Cryotherapy is another treatment option. Surgical excision may also be considered, but may be precluded by location or patient comorbidities. Surgery should be performed in cases where invasive SCC cannot be excluded.

In summary, it is important for the clinician to have an index of suspicion for BD in the presence of a persistent, psoriasis-like, scaly plaque. Early diagnosis and therapy can prevent the evolution into invasive SCC. Dx

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