

# Bowen's disease with compact orthokeratosis at perianal region: a case report

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**Abstract:** Bowen's Disease (BD) is an in situ squamous cell carcinoma (SCC) of epidermis, potentially malignant, which mainly affects sun-exposed areas. Nevertheless, the disease had an increased incidence in non-photo exposed areas, such as the anogenital region, correlating ambiental exposures, highlighting the Human Papillomavirus (HPV) infection as an important risk factor. BD has a positive prognosis according to its slow progress and prolonged time taken for full expression. In the anogenital region, presented in this case, the risk of invasive carcinoma is less than 6%. This case reports an unusual presentation of BD, describing a lesion with compact orthokeratosis in the anogenital region. The condition was diagnosed in a prevention routine of primary care attention, emphasizing the importance of premature diagnosis and satisfactory treatment, not just at BDs, but in a variety of conditions.

**Keywords:** Neoplasms; Squamous Cell Carcinoma; Bowen's disease; Primary Health Care.

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## 1. Introduction

Bowen's Disease (BD) was first described in 1912 by John Templeton Bowen and consists of a squamous cell carcinoma (SCC) in situ [1]. However, only 27 years later after his discovery, the BD was described in the perianal region [2]. BD is an unusual lesion that slowly grows and holds the pattern of preserving the basal membrane of epidermis [3]. It can encroach on any part of the human body but is usually located in photo exposed areas [4]. However, non-photo-exposed areas can be affected, such as its presentation at the anogenital region, which younger patients, between 30 and 40 years old, are most affected [5].

Among the challenges of BD early diagnosis includes its lenified progression and slow presentation behavior, scarce symptomatology and multiple macroscopic phenotypes which produces a vast number of differential diagnoses [6]. This case reports an atypical presentation of Bowen's Disease in the perianal region. Our primary objective consists of describing the disease presentation and highlighting the primary care health attention as the key to the construction of the health chain and effective health promotion.

## 2. Case Report

The presented case study describes a unique occurrence of Bowen's disease (BD) in a 42-year-old female's perianal region. The patient, a smoker with a history of 11 pack-years,

was otherwise healthy with negative results for HIV, VDRL, Hepatitis B and C, and a prior normal cytopathological exam of the cervix.

The suspicious lesion, on the perianal region, was characterized by a painless, friable, and rapidly growing nature over six months. On physical examination, the lesion was the size of 0.5 centimeters, located at 4 centimeters from the anal verge, hyperchromic, irregular limits, with no sign of inflammation or pain. This context prompted a prompt excision with a 1-centimeter safety margin, resulting in a fragment of skin measuring 1.6 x 0.9 centimeters in area and 0.6 centimeters in thickness, with orthokeratosis measuring 0.8 x 0.5 centimeters.

The subsequent histological analysis revealed a skin area with moderate to intense compact orthokeratosis with the BD lesion, identified as in situ squamous cell carcinoma (SCC). Associated to this, the presence of nonspecific chronic inflammation and notably, there wasn't neoplasia infiltration, and the surgical margins were abnormalities free. Following the procedure, the patient was referred for dermatological evaluation 3 months after the excision, revealing a 2-centimeter scar with no sign of new lesions. A subsequent assessment after 5 months showed good healing with no recurrence.

### 3. Discussion and conclusion

The discussion delves into the epidemiology of BD, highlighting its predominance in Caucasian women over 60 [7], with a major correlation to ultraviolet (UV) light exposure. The cumulative exposure to this radiation produces a DNA damage and immunosuppression, facilitating the clonal expansion of a p53 gene mutation [8].

At global locations with reduced daylight hours, the lesions are mainly associated with infections caused by different subtypes of Human Papillomavirus (HPV), highlighting the subtype 16, strongly correlated to vulvar BD [5]. Other correlations are also described such as winter immunosuppression, exposure to arsenic and other pathologies, like the inflammatory bowel diseases and colon neoplasia [8].

The morphological variations of BD lesions are briefly discussed, emphasizing the importance of dermoscopy for diagnosis, particularly in identifying vascular structures not visible to the naked eye [10]. BD is characterized as a precancerous disorder with the potential to progress to invasive SCC [11]. The time required for this progression is noted to be 2 to 40 years [2] and, in the anogenital region, the risk of invasive carcinoma is reported to be 2 to 6% [12].

BD presents as an asymptomatic disease in the vast majority of cases, however, symptoms such as itching, burning, local pain and bleeding can occur [6]. The appearance of the lesions on physical examination varies from erythematous-scaly plaques, with clear and irregular edges, to vegetative lesions and even ulcerated lesions, which may be associated with hypo or hyperpigmentation [3]. Slow growth, scarce symptoms and multiple macroscopic phenotypes impacts on a wide range of differential diagnoses, thus creating a challenge for early diagnosis [11].

As it presents with varied manifestations, a plurality of interventions can be chosen [5]. Surgical local resection is the consensus choice approach, with lower recurrence rates performed adhering wide margins and possibly associating skin grafts or flaps according to the size of the skin defects for primary closure or secondary intention [9]. However, we have less invasive alternatives such as photodynamic therapy, argon laser, chemotherapy, and cryotherapy [2]. These consist as options for retreatment in cases of recurrence.

Bowen's disease has a good prognosis, according to its slow growth and positive response to interventions [5]. It has only 3% potential of progression to invasive squamous cell carcinoma [7]. Some genetic changes, such as the mutation in the p53 tumor suppressor gene and the expression of the nuclear antigen Ki-67, elevates the progression tendency to invasive carcinoma. However, more studies need to be developed for a more assertive association [3]. The case presented, besides of revealing an unusual body region presentation, stands out the diagnostic effectiveness of the health primary care unit relied on the Brazilian public health service, reinforcing the importance of a complete physical

examination of the perineum at gynecological routine evaluation, bringing out the benefits for patients justified by eventual absence or frustrating symptoms that reflect a late or even a missing diagnosis [13].

The whole case shows that it's important to be aware of details, they will not always reveal the most common expected result. The doctor-patient relationship is deeply decisive to create a link that allows the patient to feel comfortable sharing whatever is. The primary health care service, in Brazil, is the pillar for this construction as it promotes longitudinal and integral follow-up of the patient. Therefore, reveals the importance of diagnosing and premature treating of diseases, such as this one described with an atypical presentation. This set of care, besides providing a better prognosis for his disease, turns the patient the main protagonist in promoting his own health.

In conclusion, this case study on Bowen's disease provides a valuable contribution to the literature, offering a rare insight into an atypical presentation in the perianal region with compact orthokeratosis. The meticulous documentation, insightful discussion on epidemiology, and thorough follow-up make it a noteworthy addition to dermatological knowledge. I strongly recommend the publication of this study as it not only enriches our understanding of Bowen's disease but also serves as a valuable reference for clinicians and researchers encountering similar cases.

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**Conflicts of Interest:** None.

**Supplementary Materials:** None.

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