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Porokeratosis in chronic leg plaques in a patient with poor health literacy

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Abstract

Porokeratosis is a rare disease of the skin, characterized histopathologically by abnormal keratinization; it commonly affects photo exposed sites. Its exact etiology is unknown, but it may result from the interaction of genetic and environmental factors. However, this is an important disease to keep on the differential diagnosis of papules or plaques in the lower limbs, as it may progress to malignancy, most commonly squamous cell carcinoma. The patient is a 61 year old male with lower socioeconomic status.

Physical exam reveals chronic leg pain and plaques that present differently than expected from type 2 diabetes and fluid overload. There are multiple raised, keratotic papules on both lower extremities with mild peripheral edema. A punch biopsy was performed for the keratotic papules and revealed them to be porokeratosis. The patient was referred to outpatient dermatology for follow-up, but has poor medical compliance and did not follow with outpatient dermatology. The patient has returned to the hospital for dermatology follow-up times and is treated by the wound care team and by internists for his peripheral edema and then discharged where he then becomes non compliant with medication. The patient should follow with dermatology to have his condition treated and screenings for skin cancer but due to a multitude of factors such as poor health literacy, distance to office from home, financial issues, and required frequency of visits, he has not. Since the plaques present differently than diabetic ulcers it is believed some of them are a result of porokeratosis progressing to malignancy. Future research would include additional biopsies on the patient and possible medication formulations for porokeratosis such as 5-fluorouracil, lovastatin cream, or laser therapy.

Introduction

Porokeratosis is a disease characterized by abnormal keratinization [1]. In a swedish study, it was found that its prevalence was as high as 24.2 cases over 100,000 persons [2]. The etiology of this disease

is still unknown, but it is believed that it could result from the interaction of genetic and environmental factors, such as Ultraviolet (UV) exposure, trauma, drugs, infectious agents, and immunosuppression; especially among organ transplant patients [3]. It commonly presents on the limbs, usually as keratotic papules, or annular plaques with central atrophy [1]. Its histologic hallmark is cornoid lamella [1]. Porokeratosis is diagnosed by biopsy [1]. Up to this date, there are no guidelines on treatment standards, but some treatments used are imiquimod cream and topical or systemic retinoids [1]. 6.9 to 30% of the time, these lesions can progress to cancer; especially squamous cell carcinoma [1]. The main factors associated with malignancy include old lesions, lesions of great size, lesions on the limbs, and cases of linear porokeratosis [3]. We present a case of Porokeratosis in the context of a patient with no known risk factors, who was found to incidentally have the disease. Our main goal is to inform about this disease's presentation, so medical professionals could consider it in their differential diagnosis of dermatologic lesions.

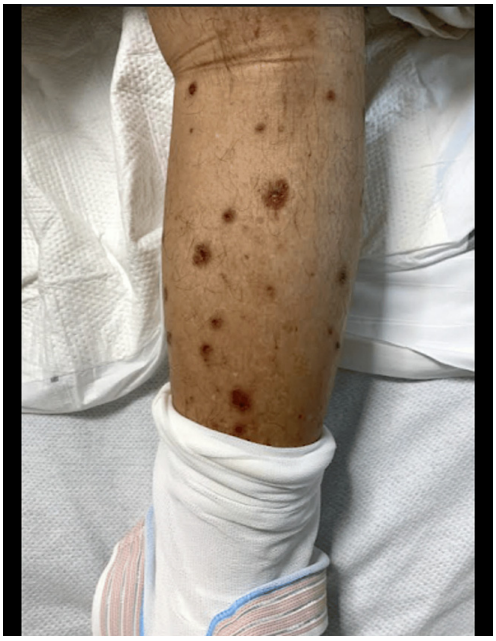

Figure 1: Multiple purple plaques at the left lower extremity.

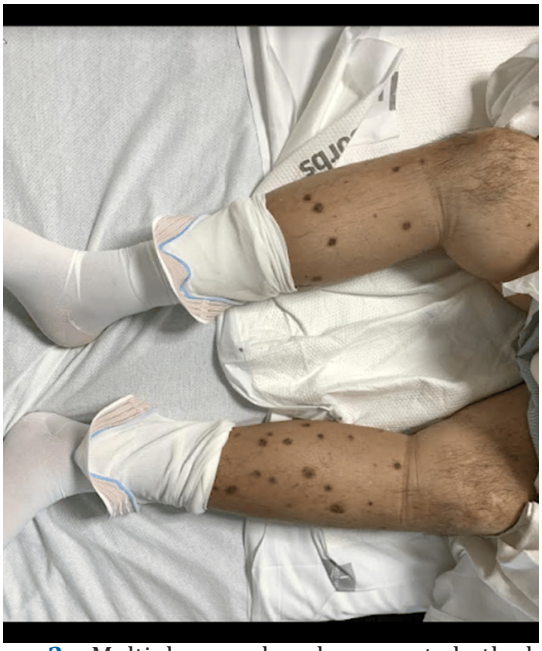

Figure 2: Multiple purple plaques at both lower extremities.

Case Presentation

We present a 61 year old male with a past medical history of type 2 diabetes, congestive heart failure, COPD and recurrent admissions for exacerbation of heart failure who presented with multiple raised plaques on his lower extremities with significant pruritus (Figure *2*). Social history included occasional smoking and drinking alcohol on weekends. Family history included a history of colon cancer in his mother who is deceased. Other social determinants of health include unstable living situations where he is in the care of his siblings , and noncompliance with medications. The patient reported that the lesions have been present for many years and are associated with stinging, pain, and itching exacerbated by exposure to sunlight. However, there was no mucosal involvement. He was then admitted for further workup and treatment, and Dermatology specialist's were consulted. Subsequently, a punch biopsy of the keratotic plaques was performed for histopathology characteristics of the lesion. The histological examination revealed cornoid lamellae of parakeratotic cells with an absent granular layer beneath them, and the superficial dermis

showed a dense perivascular lymphocytic infiltrate. Based on the clinical and histopathological findings, a definite diagnosis of porokeratosis was reported. Based on the Dermatology specialist's recommendations, the patient was instructed to follow up outpatient for additional treatment. As his presentation would be of particular interest due to being an uncommon disease, the patient was asked for consent to submit his case for publication, which he approved. Hospital course was uneventful and the patient was discharged with instruction to follow up with outpatient dermatology. However, on attempts to follow up with the patient, it was discovered that he had poor medical compliance and did not follow with outpatient dermatology.

Discussion

There are several factors associated with the development of porokeratosis. Some of the most important are the exposure to UV radiation and having previous autoimmune conditions [4]. In this specific case, our patient's main risk factor for the disease is that he has had the plaques several years before the diagnosis, which is consistent with the disease manifestation of around 50 years of age [1]. In regard of the this disease's etiology, there are indications that this could stem from a basophilic reaction involving Interleukin (IL-31), causing the pruritus and raised papules [5]. Furthermore, men appear to be more susceptible to porokeratosis; of the 22 cases reported by Takiguchi et al. (2010), 90% included men between the ages of 27 and 84 [6]. Importantly, porokeratosis can remain undiagnosed for many years, as there are other more common conditions that resemble it: plaques are usually misdiagnosed for actinic keratoses, lichen sclerosus, and lichen planus [4].

There have been numerous reports of basal cell carcinoma, squamous cell carcinoma, and melanoma developing from porokeratosis [4]. As the risk of malignancy is high, our patient ideally would follow with dermatology to have his condition treated, and for screenings of skin cancer to be performed. However, due to non compliance and poor health literacy the patient does not. Future workup would include additional biopsies on the patient and possible medication formulations for porokeratosis such as 5-fluorouracil, imiquimod cream and topical or systemic retinoids, depending on the subtype of porokeratosis [1,4].

It was shown that patients that were institutionalized in the past were more prone to comply with their medication [7]. However, this patient was already hospitalized several times for acute heart failure exacerbations, and even so he did not comply with his medications. Consequently, we consider his porokeratosis prognosis poor.

This case report highlights the importance of having a high index of suspicion regardless of symptomatology to prompt early recognition of such presentation and help readers to be aware of this disease with its potential malignancy transformation.

Conclusions

Healthcare professionals should be aware of porokeratosis as a differential diagnosis for a patient with multiple pruritic plaques. Although porokeratosis in itself is non-life threatening, its association with the development of skin malignancy warrants further investigation and regular skin exams for our patients.

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