

Single Case

Chronic Prurigo: An Unusual Presentation of Hodgkin Lymphoma

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Keywords

Prurigo nodularis · Hodgkin lymphoma · Psychogenic pruritus · Diagnostic approach

Abstract

Background: Prurigo nodularis is a condition of unknown origin defined by papulonodular eruption and intense pruritus. Hodgkin lymphoma often presents nonspecific initial symptoms. An association between systemic malignancy and cutaneous manifestations has long been documented. We report a case of prurigo nodularis as a first presentation of Hodgkin lymphoma. **Case:** A 35-year-old woman presented with a 2-year history of pruritus. Previously diagnosed with bedbugs, the pruritus persisted even after insect eradication, with the appearance of papulonodular lesions consistent with chronic prurigo. The pruritus and the pain were refractory to all treatments. She had no past medical history or clinical, radiological, or laboratory findings. A lymphadenopathy was revealed 2 years after onset of the symptoms. Lymph node biopsy showed a nodular sclerosis Hodgkin lymphoma. The patient was initiated on chemotherapy and the skin lesions decreased. **Conclusion:** This case report of chronic prurigo as the first manifestation of a systemic malignancy reminds us of the importance of a systematic diagnostic approach to this kind of patients initially and throughout time, especially if the symptoms do not respond to treatment. Our case may question the role of imaging examinations in the management and follow-up of a persistent prurigo nodularis.

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Background

Prurigo nodularis (PN) is defined by the presence of numerous, symmetrically distributed hyperkeratotic or erosive nodules, especially in areas accessible to scratching, intense pruritus being the dominant symptom [1, 2]. The etiology can be related to multiple diseases ranging from dermatological (atopic dermatitis or cutaneous mycobacterial infection) and systemic (HIV infection, anemia, diabetes, or *Helicobacter pylori* infection) to psychiatric disorders (depression or anxiety) [3]. A direct association with itch is not always shown and the time to diagnosis can be long [4]. PN is widely known to be a sign of malignancy, in particular of malignant lymphoproliferative disorders including leukemia and Hodgkin disease [3, 5]. Hodgkin lymphoma (HL) represents approximately 10% of lymphomas and the pathway to diagnosis is often difficult with a long delay before diagnosis. Howell et al. [6] reported 87 days from help-seeking to HL diagnosis and 158 days from time of symptom onset to HL diagnosis.

We report the case of a patient with no past medical history presenting with PN as an isolated prodromal symptom of Hodgkin disease.

Case Presentation

A 35-year-old woman presented with a 2-year history of intense pruritus with pain, followed 14 months after onset of the symptoms by the eruption of papulonodular lesions (Fig. 1). The patient had no past medical history and was not undergoing any treatment. Initially, bedbugs had been identified at her house, but the pruritus had persisted even after eradication by a professional pest control company. Initially, the laboratory investigation only showed a high IgE rate (272 kU/L; normal <100 kU/L) and no other abnormal findings on the clinical examination or on the chest X-ray. The patient consulted her gynecologist, who did not find anything in particular. Treatment with antihistamines, phototherapy, topical corticoids, and tricyclic antidepressants did not achieve any symptom remission. Only whole-body cryotherapy sessions led to a small improvement of the pruritus.

About 14 months after onset of the symptoms, the patient began to have papulonodular cutaneous lesions. A skin biopsy showed hyperkeratosis with epidermal acanthosis, focal hypergranulosis, and mononuclear infiltrate in the upper dermis, consistent with a chronic nodular prurigo. Patch tests were performed to investigate for cutaneous allergy, but the results could not explain the extensive dermatosis of our patient.

Six months later, she revisited her general practitioner with intractable pruritus, chills, and nagging pain. There was no history of fever, night sweats, or weight loss, but the patient complained of severe tiredness. A physical examination revealed firm and shaped cutaneous nodules from 0.3 to 2 cm in size involving the extensor aspect of the limbs, back, and buttocks. Excoriations and hyperchromic scars from previous nodules were present. A clinical examination did not reveal any adenopathy or hepatosplenomegaly.

Treatment with dexamethasone at 6 mg/day was introduced, and after the corticosteroid therapy the symptoms worsened, with the appearance of a palpable supraclavicular lymphadenopathy. A chest X-ray showed a mediastinal paratracheal mass confirmed by a CT scan, which also revealed several anterior mediastinal, right paratracheal, and right hilar lymphadenopathies.

The laboratory test results were as follows: IgE rate 130.0 kU/L (normal <100), C-reactive protein 13.38 mg/L (normal <5), leukocytes 11.9 G/L (normal 4–10), and immunoglobulin M 3.71 (normal 0.40–2.30). Protein electrophoresis showed no anomalies. Serologic tests ruled

out hepatitis, HIV, toxoplasmosis, parvovirus B19, syphilis, Epstein-Barr virus, bartonellosis, rickettsioses, Q fever, brucellosis, and tuberculosis infection. A lymph node biopsy revealed a polymorphic lymphoid infiltrate with fibrous bands, as well as the presence of Reed-Sternberg cells, neoplastic cells (CD30, MUM1, PD-L1, and CD20 positive; CD15 negative) and T cells (CD3 positive). These findings were consistent with a nodular sclerosis HL. The patient was initiated on chemotherapy with 12–14 sessions of Adriamycin-bleomycin-vinblastine-dacarbazine. She was relieved of the pruritus and pain after the first session and the skin lesions decreased.

Discussion

Chronic pruritus has a strong influence on quality of life and presents a true challenge to diagnosis and treatment. Our case illustrates the complexity of PN, for which a single etiological diagnosis is not always easy to find. Symptomatic treatment is thus often attempted, but this can be disappointing in the long term. Our patient presented with pruritus which had persisted even after insect eradication and after having visited several practitioners for reevaluation. Considering that there were no other relevant findings, bedbugs remained the first diagnosis as a direct cause of the itching or a factor that generated psychological anxiety maintaining the pruritus. The pruritus had thus first been misdiagnosed as having a psychogenic cause; then, finally, the diagnosis of HL was established, 2 years after onset of the symptoms. However, the usual investigations had not detected any consistent signs.

Our case illustrates the normal steps of clinical follow-up and highlights the lack of precocious markers for some lymphomas. The delay in diagnosis has several negative implications as patients' complaints might be ignored while the pruritus is in fact symptomatic of a serious disease. In addition, some patients could feel neglected and lose trust in medical care, resulting in an inadequate follow-up. It has already been reported that generalized PN without any primary dermatosis requires examination to exclude a systemic disease, including paraneoplastic itch [7]. Even with a suspicion of a psychological origin with no other evident etiologies and despite extensive investigations, physicians should keep in mind a relatively broad range of differential diagnoses and heed any new clinical signs such as the appearance of an adenopathy. Lymphadenopathy has been reported to be the most frequent clinical feature in HL [8], and cutaneous paraneoplastic manifestations occur in 17–53% of patients with HL [9]. The role of imaging examinations in the diagnosis and management of lymphoma has already been discussed [10, 11]; however, and considering epidemiological factors and the clinical context, our case may question their utility when faced with persistent PN.

Conclusions

This case provides further evidence for an association between PN and HL. Thus, it is important to remember that chronic pruritus always requires a systematic diagnostic approach initially and throughout time, particularly when a patient does not respond to treatment. Several factors such as the history of the patient and epidemiological factors should help to redefine the role of imaging examinations in the diagnostic approach to nodular prurigo.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Disclosure Statement

The authors declare that they have no competing interests.

Author Contributions

M.P. was involved in direct management of the patient. S.D. drafted the first manuscript and reviewed the literature. L.T.T. supervised the manuscript drafting. All authors read and approved the final manuscript.

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Fig. 1. Prurigo papules with scratch marks and pigmentation seen on the legs.