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Infiltration of abdominal striae distensae by Hodgkin's Lymphoma.

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Conflicts of interest:

The authors declare not to have any interest conflicts.

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Abstract

Background: Cutaneous Hodgkin's lymphoma (HL) is generally secondary to dissemination from a nodal HL; it tends to occur late in the course of the disease, and is associated with a poor prognosis. This paper reports on a female patient with nodular Hodgkin's lymphoma, initially diagnosed due to cutaneous involvement affecting abdominal *striae distensae*.

Methods and results: A 54-year-old woman consulted the Dermatology Unit with thickening and intense itching in abdominal *striae distensae* associated with previous pregnancies; for the last three months, striae had been hard and whitish in colour. Microscopic examination of a skin biopsy revealed dermal and hypodermal nodular infiltration by large CD30+ and CD117+ Sternberg-like cells, together with inflammatory cells (lymphocytes, plasma cells, eosinophils and neutrophils) and focal collagen sclerosis. These findings were consistent with a diagnosis of infiltration by Hodgkin's lymphoma. Lymph-node ultrasonography revealed multiple adenopathy. Examination of a resected lymph-node confirmed the diagnosis of nodular sclerosis HL.

Conclusions: Cutaneous involvement is reported in 0.5-3.4% of cases. This paper describes a case of nodular sclerosis HL diagnosed on the basis of a skin biopsy performed due to thickening and intense pruritus in abdominal *striae distensae*. To the authors' knowledge, no similar case has hitherto been reported. This highly-unusual presentation may be linked to Wolf's isotopic response.

Introduction

Cutaneous Hodgkin's lymphoma (HL) is generally secondary to dissemination from a nodal HL[1,2]; it tends to occur late in the course of the disease, and is associated with a poor prognosis[3,4]. It often manifests first as painless erythematous nodules and papules in the chest area[4,5]. Primary cutaneous HL is extremely uncommon, and only a few cases have been reported[5].

Three mechanisms have been suggested to account for the spread of systemic HL to the skin: hematogenous dissemination; direct extension from an underlying nodal focus; and retrograde lymphatic spread, this being the most common[2].

Clinically, skin invasion by HL may mimic infectious diseases such as scrofuloderma, sporotrichosis, actinomycosis and tularemia[6].

This paper reports on a female patient with nodular Hodgkin's lymphoma, diagnosed due to cutaneous involvement affecting abdominal *striae distensae*.

Materials and Methods

A 54-year-old woman with no known drug allergies, a smoker with a personal history of urticaria attributed to metamizol, chronic obstructive pulmonary disease and chronic gastritis. The patient was under examination by the Internal Medicine Unit for constitutional syndrome, accompanied by hard, painless, non-adhering, enlarged axillary and inguinal lymph nodes and lower-limb non-pitting edema. She was referred to the Dermatology Unit for with thickening and intense pruritus in abdominal *striae distensae* associated with previous pregnancies; for the last three months, striae had been hard on palpation, whitish in colour and shiny in appearance (Fig.1).

Clinical suspicion of scar sarcoidosis led to a skin biopsy which revealed dermal and hypodermal nodular infiltration by large CD30+ and CD117+ Sternberg-like cells, together with inflammatory cells (lymphocytes, plasma cells, eosinophils and neutrophils) and focal collagen sclerosis. These findings were consistent with a diagnosis of infiltration by Hodgkin's lymphoma (Fig. 2 and Fig. 3).

Blood tests showed leukocytosis (22,950) with neutrophilia (84.7%), platelet count 662,000 and ESR (1st hour) 56 mm; blood biochemistry showed IgA 40 mg/dl.

Lymph node ultrasonography revealed multiple adenopathy; affected nodes were rounded, ranged between 22 and 32 mm (major axis), and displayed abnormal flow patterns. Fine-needle aspiration biopsy (FNAB) was of little diagnostic value. Bone marrow biopsy showed no infiltration by tumor cells. Flow cytometry showed that T cells accounted for 8%, with a CD4/CD8 ratio of 1:1, and B cells for 1.6% with no clonal proliferation. CT scan of chest, abdomen and pelvis revealed large axillary, mediastinal, perivascular and retroperitoneal pelvic and inguinal lymph nodes. Nodules were detected in subcutaneous cell tissue around the chest wall and below both breasts, together with marked edema of the anterior abdominal wall, in the hypogastric region where cutaneous striae were located. Examination of a resected inguinal lymph node confirmed the diagnosis of nodular sclerosis LH (Fig.4).

Discussion

Skin invasion by Hodgkin's lymphoma is uncommon, and is associated with a poor prognosis[2,3]. It is reported in 0.5-3.4% of cases of HL, generally in patients with human immunodeficiency virus (HIV); it tends to appear late in the course of the disease and is clinically aggressive, posing a major challenge to the immune system[1]. Lesions tend to occur distal to involved lymph nodes due to retrograde lymphatic spread[7]. Introcaso *et al.*[2] report that the incidence of cutaneous LH has decreased in recent decades, owing to more effective treatment with chemotherapy, radiotherapy and stem cell transplantation

In the right histological context, CD30 positivity in Hodgkin lymphoma cells is the main immunohistochemical marker for diagnosis in a skin biopsy[8]. However, CD30+ expression may also be observed in a wide range of cutaneous lymphoid tumors (lymphomatoid papulosis, mycosis fungoides, anaplastic lymphoma, large-cell B lymphoma) as well as in inflammatory processes[2,9,10,11]; immunohistochemical examination using other markers, including CD117 (cKIT)[2,5,12], is required, and a correct clinical-pathological correlation is essential. Here, the patient presented with multiple elevated white and reddish abdominal striae, which were inflammatory in appearance and clinically suggestive of sarcoidosis. Microscopic analysis revealed lymphoid nodules containing CD30+ and CD117+ Reed-Sternberg cells bound by dense collagen bundles, prompting a preliminary diagnosis of cutaneous HL which was later confirmed by axillary lymph node biopsy.

Abdominal *striae distensae* tend to manifest as white and reddish lines; at histological examination, epidermal atrophy, a linear arrangement of collagen bundles and absence of dermal adnexa are common findings[13,14]. The etiology of this non-specific skin manifestation is unknown, but it has been associated with a combination of genetic, mechanical and endocrine factors[13], including growth spurts during adolescence, obesity, pregnancy, connective tissue abnormalities, Cushing's syndrome and corticosteroid drug treatment[15].

Macaya *et al*.[3] reported cutaneous granulomas as the first manifestation of Hodgkin's disease, while Jurisic *et al*.[1] described cutaneous spread of HL in the form of erythematous papules in the upper limbs and abdominal region.

The literature contains very few reports of primary cutaneous Hodgkin's lymphoma in the absence of systemic disease. Mukesh *et al*.[5] described a 59-year-old male patient with a painful, indurated erythematous lesion on the left flank which was diagnosed as primary cutaneous HL, and which regressed completely after six cycles of chemotherapy. Szur *et al*.[16] reported three cases of primary cutaneous Hodgkin's lymphoma confined to the skin, with no lymph node, liver or spleen involvement. Sioutos *et al*.[17] studied five patients with primary cutaneous Hodgkin's lymphoma, all of whom presented with skin lesions without evidence of systemic HD at the time of diagnosis. Two patients developed systemic HL 2 months and 6 years, respectively, after the initial diagnosis of primary cutaneous HL. The remaining three patients had a benign course without systemic disease with up to 20 years of follow-up. Some authors[18,19] argue that primary cutaneous HL, without concomitant lymph-node involvement, is exceptional, and that most published cases are in fact lymphomatoid papulosis or large cell anaplastic lymphoma.

To conclude, this paper reports on a case of nodular sclerosis Hodgkin's lymphoma diagnosed on the basis of a skin biopsy taken from thickened and itching abdominal *striae distensae*. To the authors' knowledge this is the first published case of this unusual presentation, which may be linked to Wolf's isotopic response (appearance of a new dermatosis at the site of a previous dermatosis)[20].

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Legends:

Figure 1.

Abdominal *striae distensae*: hard on palpation, whitish in colour and shiny in appearance.

Figure 2.

Dermal and hypodermal nodular lymphocyte infiltration (A, HE 1x, and B, HE 10x), with large Sternberg-like cells and accompanying inflammatory cellularity (C, HE 20x).

Figure 3.

CD30-positive Reed-Sternberg cells (40x).

Figure 4.

Lymph node affected by nodular sclerosis Hodgkin's lymphoma (A, HE 1x, and B, HE 10x) with Reed Sternberg cells (C, HE 20x) which were CD30 positive (D, HE 20x).

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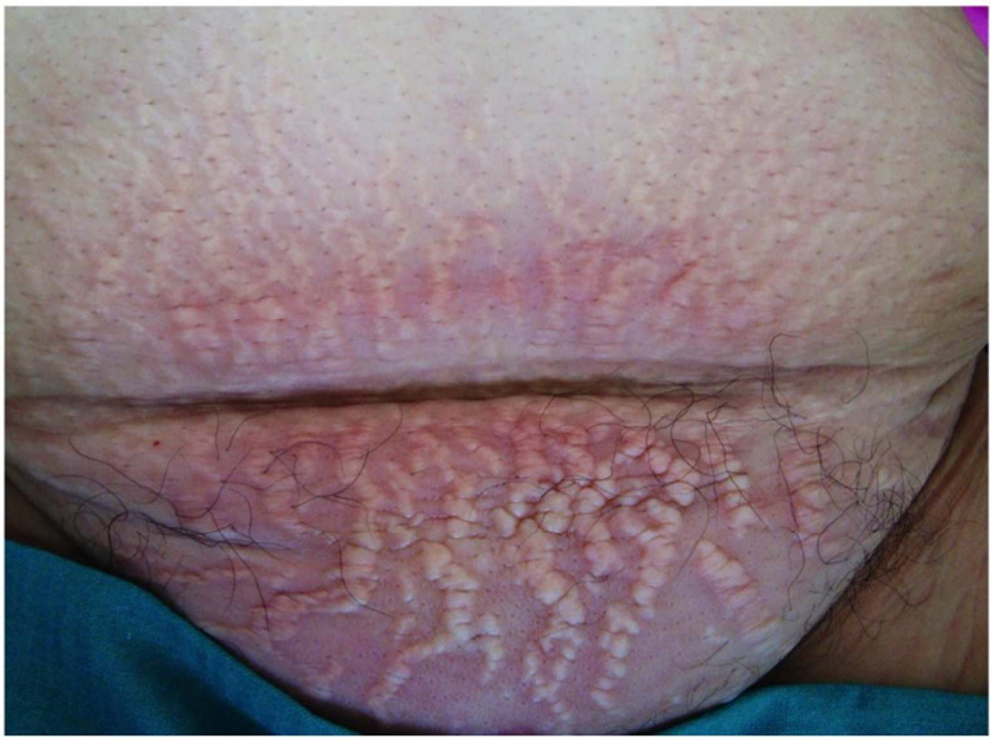

Abdominal striae distensae: hard on palpation, whitish in colour and shiny in appearance.

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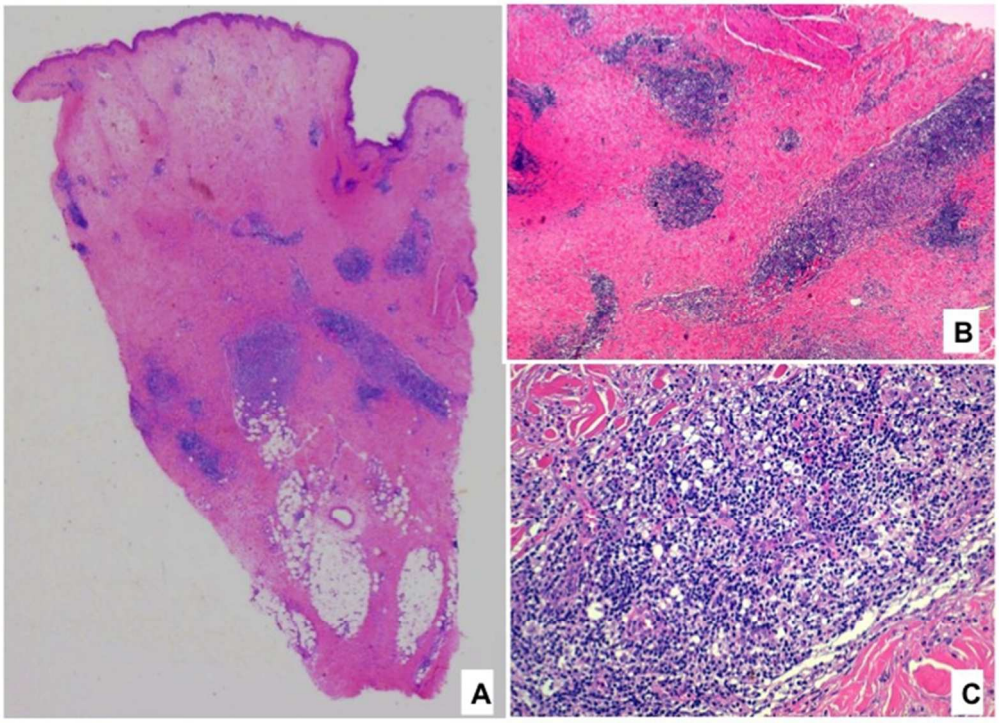

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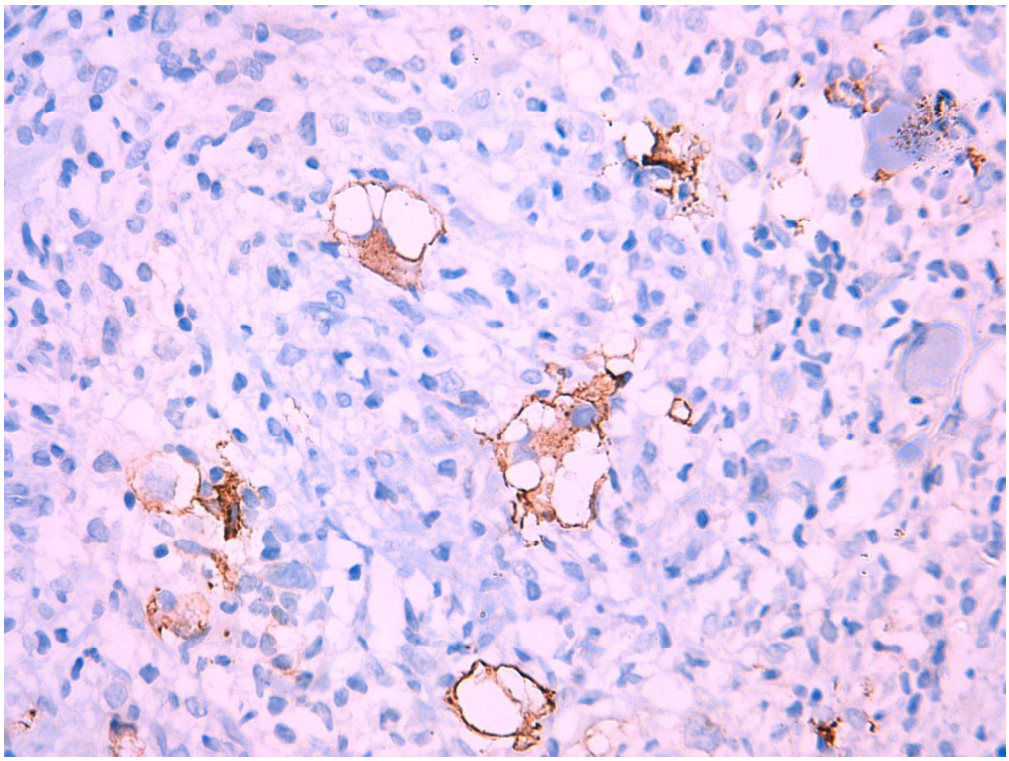
249x180mm (72 x 72 DPI)



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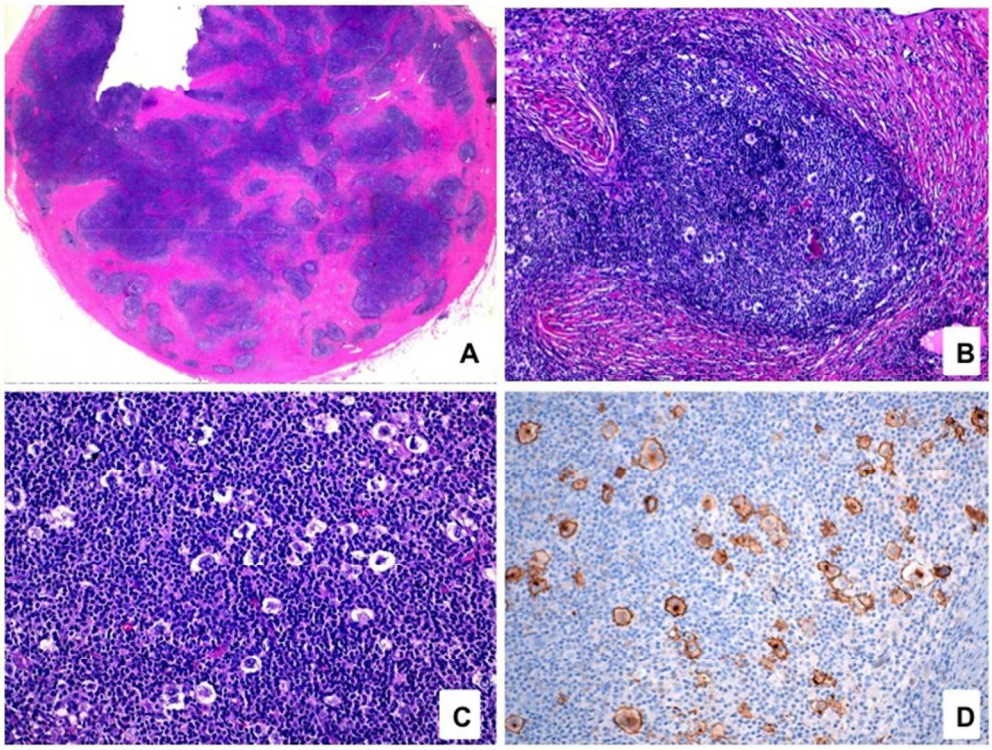

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