

Pseudolymphoma versus lymphoma: An important diagnostic decision

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Abstract

Small innocuous growths on the face usually do not pose difficulty in diagnosis on histopathology. However, some benign inflammatory lesions might mimic malignancy and hence need further investigations for final diagnosis. The distinction between a benign/inflammatory/malignant lesion needs no emphasis as the treatment plan, prognosis and the patient's well-being depends on it. Lymphocytoma cutis, or Spiegler-Fendt Sarcoid, is classed as one of the pseudolymphomas, referring to inflammatory disorders in which the accumulation of lymphocytes on the skin resembles, clinically and histopathologically, cutaneous lymphomas. To obtain an accurate diagnosis, careful clinical evaluation, as well as histopathological and immunohistochemical examination is needed. One such case of an otherwise unassuming growth mimicking malignancy is being presented.

Key Words: Cutaneous lymphoma, polyclonality, pseudolymphoma, reactive lesion

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INTRODUCTION

Solitary nodules on the face can be challenging to diagnose. A host of differential diagnoses can be narrowed down majorly by the biopsy findings. However, at times the findings on hematoxylin and eosin (H and E) stained sections will not ascertain a definitive diagnosis, requiring additional investigations such as immunohistochemistry (IHC). Lymphocytoma cutis, a benign reactive lesion, is one such lesion which can present as a solitary nodule on the face mimicking lymphoma both clinically and histopathologically. By eliciting the polyclonality of the lymphocytes through IHC in such lesions, the suspicion of lymphomas can be eliminated, thereby establishing the inflammatory/reactive nature of the lesion in doubt. This differentiation needs to be emphasized, keeping

in view the treatment and prognosis of the patient. A case of single nodular swelling in an adult female patient, which was diagnosed as lymphocytoma cutis is being discussed here.

CASE REPORT

A 50-year-old female patient, reported with the complaint of a swelling on her forehead. The swelling was mildly itchy, started about 20 days back, slow growing and attained the present size of about 10 mm in diameter. On examination, a well-defined firm swelling was noted on the left side of the forehead near the hairline. The surface was smooth, with hair growing on it and the color was same as the adjacent skin. Patient did not give recent history of any triggering factors such as insect bite, drug intake, vaccination or body piercing. Clinically, since it

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was well defined and benign in nature, an excisional biopsy of the same was done and the tissue was sent for histopathological examination.

On gross examination, a solid round to ovoid mass of 10 mm diameter was seen. The surface was smooth with hairs growing on it. The cut surface was creamish white solid mass. The specimen was cut into two bits and both the bits were taken for processing.

H and E stained sections showed epidermis with its appendages and connective tissue stroma. The stroma was highly cellular, with a clear zone of connective tissue separating the cells in dermis from the epidermis [Figure 1]. The cells were predominantly lymphocytes with few plasma cells and histiocytes. It was arranged in a follicular pattern with germinal centers in few regions [Figure 2]. Based on the above findings a probable diagnosis of lymphocytoma cutis was made. However, to establish the polyclonality of the lymphocytes and thus rule out lymphoma, IHC with CD3, CD10, CD20 and leucocyte common antigen (LCA) was done. Results showed positivity with all the antibodies, ruling out lymphoma and arriving at the final diagnosis of lymphocytoma cutis [Figure 3]. Regular follow-up of the patient for over a year has been uneventful.

DISCUSSION

Lymphocytoma cutis, also known as Spiegler-Fendt Sarcoid or pseudolymphoma, is a rare lesion, presenting either as localized solitary nodule or as disseminated multiple shiny plaques.^[1] It is a pseudolymphoma of B-lymphocytes affecting young adults with a female predilection (3:1)^[2] and propensity for occurrence in caucasians.^[3] The localized type constituting 70% of the cases often occurs on the face, in middle-aged female patients.^[4] The disseminated type called as the miliary lymphocytoma cutis, is usually seen in midsection and extremities of elderly individuals.^[2] No data on the frequency and total number of lymphocytoma cutis cases are available. A discrete subset of lymphocytoma cutis, borreliolymphocytoma, occurs primarily in Europe, in areas in which the ricinus tick is endemic.^[5] Various factors such as acupuncture, body piercing, any drug intake, infection by the bacterium *Borrelia burgdorferi*, insect bite or tattoos are known to trigger this lesion. Many a times no such factor can be elicited and hence remains idiopathic.^[6,7] It can present as itchy solitary or multiple nodules or plaques with skin colored or erythematous or brownish to violaceous surface with a soft consistency.^[8] In the present case, a 50-year-old female patient reported with a very innocuous looking solitary nodule on the face. It was itchy and had the same consistency as the adjacent skin without any change in the surface color.

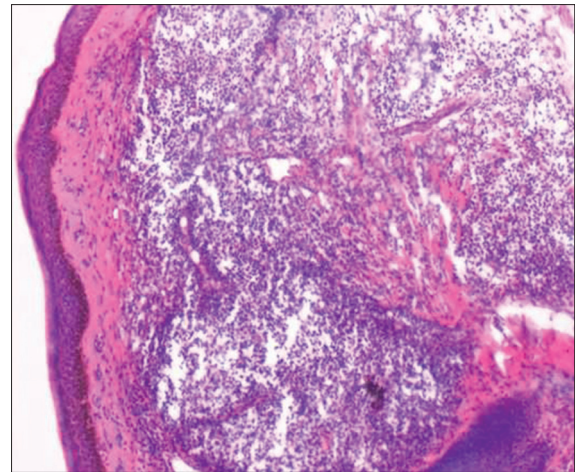


Figure 1: Photomicrograph reveals epidermis, highly cellular dermis and an acellular Grenz zone between the two (H&E stain, x40)

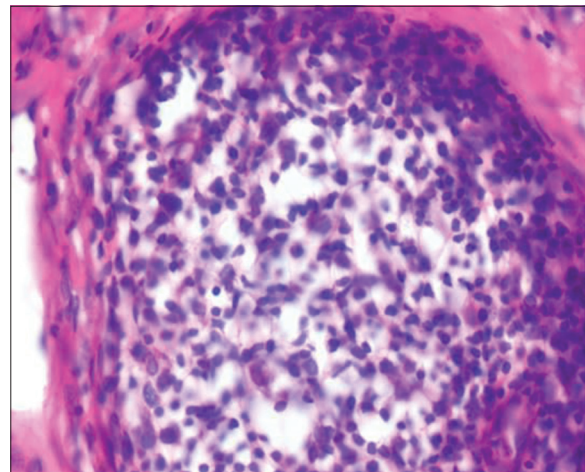


Figure 2: Photomicrograph showing follicular arrangement of cells, predominantly lymphocytes, few histiocytes and plasma cells (H&E stain, x400)

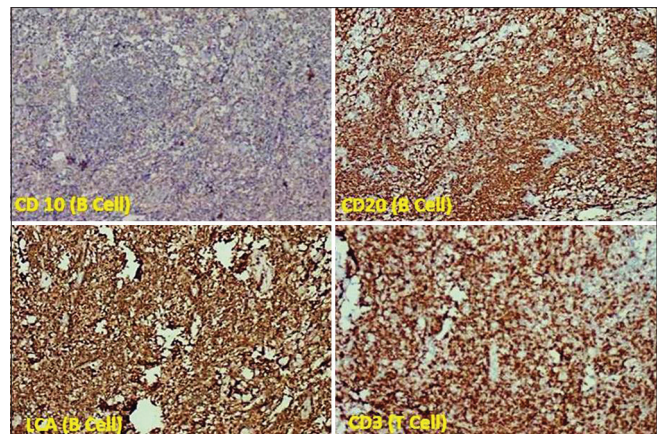


Figure 3: Photomicrograph showing strongly positive IHC staining for both B cell markers: a: CD10, b: CD20, c: Leucocyte Common Antigen and T-lymphocytes cell marker d: CD3, (IHC stain, x100) establishing the polyclonality

It was idiopathic as no etiologic factor could be elicited and was similar to the case previously published by Salvador *et al.*^[9]

It is a lymphoreticular and hyperplastic reaction to a known or unknown factor wherein the broad patches and plaques often mimic lymphoma, both clinically and histopathologically.^[10] The microscopic examination reveals the follicular arrangement of lymphocytes in the dermis with scattered histiocytes. Sometimes there is evidence of germinal centers with B-cells in the center, bordered by T-cells and the dermis may be separated by a thin area of collagen. However, usually there is no evidence of germinal center in lymphomas.^[2,3,8] By eliciting IHC positivity with a combination of the B-lymphocyte markers - CD20, CD79a, CD10, CD45/LCA and T-lymphocyte marker CD3, the polyclonality of the lymphocytes can be established, which rules out the possibility of lymphoma.^[8] In our case, even though the features of follicular arrangement with germinal centers and presence of few plasma cells and histiocytes suggested lymphocytoma cutis, the nodular arrangement of the predominant lymphocytes mimicked lymphoma. In view of the totally differing treatment modalities and prognosis, IHC with CD3, CD10, CD20 and LCA was done. Results showed positivity with all the antibodies, ruling out lymphoma and arriving at the final diagnosis of a reactive lymphocytoma cutis.

Many of these lesions involute on their own. Various treatment modalities include surgical excision, topical or intralesional steroid, cryosurgery, or photodynamics.^[8] The excisional biopsy seems sufficient in this case, as follow-up of more than a year has been uneventful.

CONCLUSION

Establishing a definitive diagnosis needs no emphasis, since the treatment of any condition greatly depends on it. In most cases, histopathological examination of clinically similar presenting lesions helps in arriving at a final

diagnosis. However in few, histopathology only narrows down the diagnosis, further requiring diagnostic molecular investigations such as IHC. This is of great importance in lesions where benign and malignant lesions need to be differentiated, since the treatment planning and prognosis differs significantly.

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Conflicts of interest

There are no conflicts of interest.

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