Enigmatic Diagnosis: Idiopathic Cutaneous Pseudolymphoma - A Case Report

Case Report

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ABSTRACT

Introduction pseudolymphomas are benign reactive lymphoid proliferations that clinically, histologically, or both clinically and histologically resemble cutaneous lymphomas. They arise from known or unknown stimuli. Traditionally, cutaneous pseudolymphomas have been classified based on their histological and immunophenotypic characteristics or in relation to the lymphoma they mimic.

ObservationThis is a 60-year-old female patient with no significant medical history or medication use. She was admitted for management of a swelling at the root of her nose that had been gradually increasing in size over 2 years.

Facial CT scan showed a tissue mass causing multi-fragmentary destruction of the nasal bones and reaching the inner corners of the orbits, measuring 25x12x25 mm, with no cervical lymphadenopathy noted.

Based on these findings, the patient underwent complete excision of the nasal root mass; the excised specimen was sent for histopathological examination. Immunohistochemical confirmed a B-type pseudolymphoma.

For this patient, a somewhat aggressive treatment approach was chosen, starting with complete excision of the lesion followed by treatment with potent dermocorticoids, resulting in a somewhat satisfactory outcome after a 6-month follow-up period.

Discussion Pseudolymphoma present a diagnostic challenge. A judicious panel of immunohistochemistry is sufficient to diagnose pseudolymphomas. In many cases, an appropriate etiology may not be identified, leading to labeling these cases as idiopathic. A follow-up of at least 5 years is necessary to exclude the risk of cutaneous lymphomas. This observation highlights the difficulties in diagnosing benign cutaneous lymphoid hyperplasia as well as the delicacy of management.

Key Words: pseudolymphomas, idiopathic, lymphomas, immunohistochemistry

Received: 14 June 2024, **Accepted:** 5 July 2024.

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ISSN: 2090-097X, October 2024, Vol. 15, No. 4

INTRODUCTION

pseudolymphomas are benign reactive lymphoid proliferations that clinically, histologically, or both clinically and histologically resemble cutaneous lymphomas. They arise from known or unknown stimuli such as insect bites, vaccination, trauma, folliculitis, medications, jewelry, and contact agents. Traditionally, cutaneous pseudolymphomas have been classified based on their histological and immunophenotypic characteristics or in relation to the lymphoma they mimic. In other words, they are categorized as cutaneous B-cell pseudolymphomas or cutaneous T-cell pseudolymphomas, depending on the predominant lymphocytic component. It is crucial to differentiate pseudolymphomas from malignant lymphomas, which requires clinical, histological, and immunohistochemical evidence, as even benign lesions may exhibit a malignant histological pattern.

OBSERVATION

This is a 60-year-old female patient with no significant medical history or medication use. She was admitted for management of a swelling at the root of her nose that had been gradually increasing in size over 2 years.

Clinically, examination revealed a firm, non-tender swelling at the nasal root, fixed in the deep plane, with smooth, shiny overlying skin (figure 1); lymph node areas were unremarkable, and the rest of the physical examination was normal.

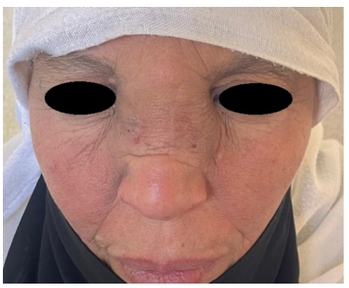

Figure 1: Clinical image showing swelling at the base of the nose with inflamed shiny skin nearby.

 DOI: 10.21608/omx.2024.297599.1253

Facial CT scan showed a tissue mass causing multi-fragmentary destruction of the nasal bones and reaching the inner corners of the orbits, measuring 25x12x25 mm, with no cervical lymphadenopathy noted.

Based on these findings, the patient underwent complete excision of the nasal root mass; the excised specimen was sent for histopathological examination which revealed fibroadipose and striated muscle tissue separated by a dense lymphoid infiltrate predominantly arranged in variable-sized follicles, sometimes centered around a germinal center. The lymphocytes appeared small, resembling reactive lymphocytes, with rare eosinophils noted.

Immunohistochemical analysis showed that the infiltrate was predominantly of B-cell phenotype (CD20 positive) with staining of germinal centers for Bcl6, CD10, and Ki-67, confirming a B-type pseudolymphoma.

For this patient, a somewhat aggressive treatment approach was chosen, starting with complete excision of the lesion followed by treatment with potent dermocorticoids, resulting in a somewhat satisfactory outcome after a 6-month follow-up period (figure 2).

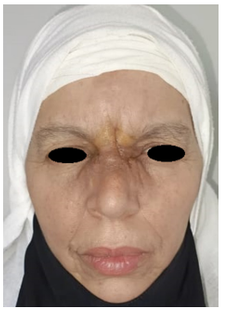

Figure 2: Postoperative control image after 6 months.

DISCUSSION

The pseudolymphoma, also known as cutaneous lymphoid hyperplasia, constitutes a heterogeneous group of conditions with a benign course, clinically and histologically resembling primary cutaneous lymphomas [1]. The concept of pseudolymphoma was initially reported under the term "sarcoma cutis" by Kaposi in 1891 [2], and later described as "cutaneous lymphoid hyperplasia" by Caro and Helwig in 1969 [3].

Cutaneous pseudolymphomas typically affect adults, although they may occur at any age.

Epidemiological data on cutaneous pseudolymphomas are rare, although B-cell pseudolymphomas seem to be more common than their T-cell counterparts, and they are also more frequent in female patients, as reported in this work.

A contributing factor proposed for the pathogenesis of cutaneous pseudolymphoma is the proliferation of lymphoid tissue associated with the skin, analogous to mucosa-associated lymphoid tissue, following antigenic stimulation [4]. Based on these data, cutaneous pseudolymphoma could potentially evolve into a true cutaneous lymphoma with permanent antigenic stimulation, as occurs in the gastric mucosa in the presence of persistent Helicobacter pylori infection [5].
However, true progression from a correctly diagnosed pseudolymphoma to lymphoma is very rare, if not impossible, despite some reported cases of progression to cutaneous lymphoma [6,7].
Nodular lesions resemble B-cell lymphomas, while plaque forms resemble T-cell lymphomas. It is essential to differentiate pseudolymphomas from malignant lymphomas, which involves clinical, histological, and immunohistochemical evidence, as even in benign lesions, a malignant histological pattern may be observed.

Immunohistochemical staining allows the histological diagnosis of lymphocytic proliferations to categorize cutaneous pseudolymphomas into type T and type B. In other words, they are classified as cutaneous B-cell pseudolymphomas or cutaneous T-cell pseudolymphomas, depending on the predominant lymphocytic component.
The concept of pseudolymphoma is a source of ambiguities and confusion due to its heterogeneity.

Cutaneous pseudolymphomas encompass entities where no cause can be identified, termed idiopathic, and others where a cause has been identified, and removal usually leads to recovery [8,9]. Pseudolymphomas result from known or unknown stimuli such as insect bites, vaccinations, trauma, folliculitis, medications, jewelry, and contact agents, leading to the accumulation of inflammatory cells that not only histologically but also clinically mimic lymphoma. Only the course of the disease will ultimately distinguish between a benign entity and a cutaneous lymphoma because sometimes it is impossible to differentiate between pseudolymphoma and cutaneous lymphoma, whether clinically, histologically, or through molecular biology techniques.
These ambiguous situations may be classified into a third category of lymphoproliferations of uncertain or indeterminate prognostic significance, similar to Monoclonal Gammapathy of Unknown Significance [10,11].

It is also worth noting that several entities initially considered cutaneous pseudolymphomas have since been reclassified as low-grade lymphomas based on clinical and pathological findings, molecular biology studies, and follow-up data.
To diagnose cutaneous pseudolymphoma, contrasting clinical and histological results are necessary, evaluating the architecture and composition of the inflammatory infiltrate, and complementing these results with immunohistochemistry and genetic rearrangement studies.

Diagnosis poses a challenge, as the mixed infiltrate pattern including histiocytes, eosinophils, and plasma cells is significant. Polymorphic infiltrates, the lack of atypical lymphocytes, and dominant lymphocytic clones are highly suggestive of pseudolymphomas [12]. The infiltrate tends to be "dense" in pseudolymphomas, whereas in most lymphomas, it is centered in the deep dermis. LCA also serves as a major differentiation marker, being negative in pseudolymphomas and strongly positive in lymphomas.

For any suspected hematolymphoid lesion, minimal immunohistochemistry includes CD10, LCA, CD20, and CD3. CD20 is used for B-cell expression, CD3 for T-cell expression and T-cell germinal centers, and CD10 for B-cell germinal centers.

CONCLUSION

In conclusion, pseudolymphomas present a diagnostic challenge. A judicious panel of immunohistochemistry is sufficient to diagnose pseudolymphomas. In many cases, an appropriate etiology may not be identified, leading to labeling these cases as idiopathic. A follow-up of at least 5 years is necessary to exclude the risk of cutaneous lymphomas. This observation highlights the difficulties in diagnosing benign cutaneous lymphoid hyperplasia as well as the delicacy of management.

FUNDING ACKNOWLEDGEMENT

The authors declare that no funding was received.

CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest.

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