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Case Report

Lupus vulgaris with Schaumann bodies – An atypical histopathological finding

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Abstract We report a case of lupus vulgaris with Schaumann bodies which is an atypical histopathological finding.

Key words
Lupus vulgaris, Schaumann bodies, sarcoidal granulomas.

Introduction

Lupus vulgaris is a progressive form of cutaneous tuberculosis occurring in a person with moderate to high degree of immunity. It develops as a result of direct inoculation, direct extension from underlying organ or by lymphatic spread.[1] Schaumann bodies are commonly seen in sarcoidal granulomas, but can be rarely seen in granulomas due to other causes especially tuberculosis. This is an example of unusual presentation of lupus vulgaris.[2]

Case report

Our case was an 11yrs old female with a single erythematous plaque over right knee since 3yrs.

She was having a positive contact with her grandfather having pulmonary tuberculosis 15 years back and he had completed full course of treatment. She was a term normal birth weight child born out of non-consanguineous marriage.

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On general examination child appeared normal.

On cutaneous examination single asymmetrical erythematous scaly plaque of size 8x6 cm **(Figure 1)** which initially started as small nodule after having trauma to knee and gradually multiple plaques developed on either side and coalesced and progressed to present size. It is not associated with any systemic symptoms. No other lesions over the body. Routine blood investigations and CXR were normal.

USG abdomen showed few non-specific mesenteric LN of size 13x7cm.

Mantoux test was positive (14TU PPD).

Histopathological confirmation The epidermis displayed irregular acanthosis associated with a dense lymphohistiocytic interface dermatitis **(Figure 2)**. Within the superficial and deep dermis there was a dense, granulomatous infiltrate composed of numerous lymphocytes, epithelioid histiocytes and Langerhans giant cells **(Figure 3)** which clinched the diagnosis of lupus vulgaris. The granulomas showed a tendency to confluence. Focally, Schaumann bodies were also seen **(Figure 4)**.

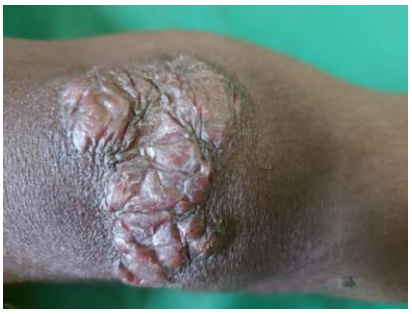

Figure 1 A single asymmetrical erythematous scaly plaque of size 8x6 cm over right knee.

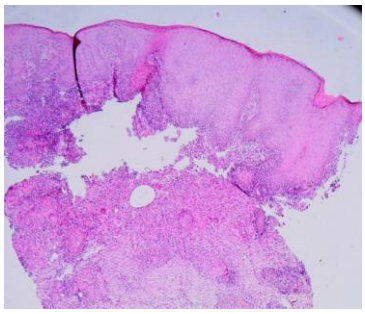

Figure 2 Epidermis with irregular acanthosis associated with a dense lymphohistiocytic interface dermatitis.

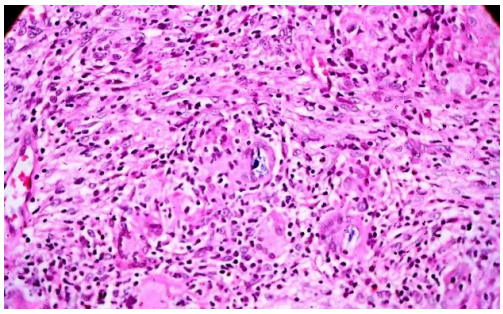

Figure 3 Reticular dermis shows marked histiocytic and lymphocytic infiltrate. Epitheloid granulomas with Langerhans type giant cells are seen.

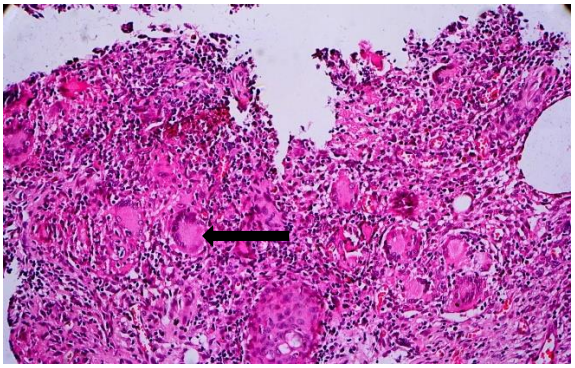

Figure 4 Langhans cells containing calcified material (Schaumann bodies in black arrow) in their cytoplasm.

Discussion

Schaumann bodies are commonly seen in sarcoidosis. Schaumann bodies also called as *conchoidal bodies* were first described by *Jorge Schaumann* in 1941. They are large concentric calcifications often containing refractile calcium oxalate crystals. They are not pathognomic of sarcoidosis as they are found in other conditions like tuberculosis, berylliosis and lymphogranuloma venerum.[2]

Cutaneous tuberculosis is now rarely encountered in Western countries, but is seen in developing countries with a prevalence of 0.1% where the incidence of tuberculosis is high. Lupus vulgaris is the most common type of cutaneous tuberculosis with most varied manifestations seen in persons with moderate to high degree of immunity.[1] It is usually acquired from an exogenous source although it may arise from hematogenous dissemination of bacilli.

The common sites of involvement are hands and neck followed by arms and legs. In India, the buttocks, thighs and legs are more common sites of involvement.

The lesion is usually single and starts as a tiny soft erythematous nodule slightly elevated above the surface. Clinically lupus vulgaris presents as five general patterns: plaque form, ulcerative and mutilating form, vegetating form, tumor like form and papular and nodular form.[3]

In plaque form, there is well defined border with little infiltration and scaling and minimal scarring in the center. In ulcerative form, crusts form over the areas of necrosis and scarring and ulceration predominates. Vegetating form is characterized by marked ulceration and extensive infiltration. The tumor like form presents as a deep infiltration in the form of soft and smooth grouped nodules or a reddish-yellow plaques. In papular and nodular type, multiple lesions occur in disseminated pattern. Besides these clinical types of lupus vulgaris, atypical forms are becoming more common.[4]

Histopathological examination of sarcoidosis

The cutaneous lesions of chronic, persistent sarcoidosis are characterized by the presence of

circumscribed collections of epithelioid histiocytes so called epithelioid cell tubercles which show little or no necrosis. The well demarcated islands of epithelioid cells contain few, if any, giant cells. Those that are present are usually of the Langhans type. Giant cells contain Asteroid bodies and Schaumann bodies.[5]

Asteroid bodies which are common are star shaped eosinophilic structures that, when stained with phosphotungstic acid hematoxylin, produce a center that is brown red with radiating blue spikes.

Schaumann bodies are round or oval, laminated and calcified, especially at their periphery. They stain dark blue because of presence of calcium.

Neither of these two bodies are specific for sarcoidosis. They have been observed in a variety of granulomas.[6]

Histopathological examination of Lupus vulgaris

Chronic granulomatous inflammation is characterized by formation of epithelioid cell granulomas. Tuberculoid granulomas are composed of epithelioid cells and giant cells.[7] Epithelioid cells are activated macrophages that appear on microscopic examination as large cells with abundant pale, foamy cytoplasm. Granulomas are usually surrounded by lymphocytes, plasma cells, fibroblasts and collagen. Caseation necrosis within the tubercles is slight/ absent. Although the giant cells usually are of Langhans type with peripheral arrangement of nuclei. There is associated lymphocyte infiltrate.[8] Sometimes this may be so prominent that the granulomatous component is obscured. The inflammation is most pronounced in the upper dermis, but in some areas, it may extend into subcutaneous layer. Secondary changes in epidermis are common.[8] The epidermis may undergo atrophy and subsequent destruction causing ulceration, or it may become hyperplastic, showing acanthosis, hyperkeratosis and papillomatosis.[9]

Tuberculous bacilli are present in such small number that they can very rarely be demonstrated by staining methods. Polymerase chain reaction (PCR) detection of mycobacterial DNA is more often positive. In some instances of lupus vulgaris when an old focus of primary infection cannot be detected, a positive tuberculin test and the response to antitubercular therapy must suffice as a proof of a tuberculous etiology.[3]

In our case, lesion was a single, irregular erythematous plaque which gradually progressed to present size of 8x6 cm in 3 years which was associated with scaling.

Histopathological difference between lupus vulgaris and sarcoidosis

Lupus vulgaris and sarcoidosis are both chronic granulomatous conditions. Histologically, both contain epithelioid cells and giant cells of Langhans type. Sarcoidosis has sparse lymphocytes when compared to lupus vulgaris which has prominent lymphocytes.[6]

Non caseating necrosis is seen in sarcoidosis and caseating necrosis is seen in lupus vulgaris. Schaumann bodies and asteroid bodies are commonly seen in sarcoidosis but it is very rare to see schaumann bodies in tuberculosis.[10] This is the first case of lupus vulgaris with Schaumann bodies.

Conclusion

Lupus vulgaris, arguably the next great mimic, after syphilis can present in protean forms. Based on clinical features and with the help of histopathological examination, we confirmed the

case as lupus vulgaris. Thus, in our case we highlight the role of histopathological examination in confirming the diagnosis of Lupus vulgaris and to rule out other differential diagnosis. Though Schaumann bodies are commonly seen in sarcoidal granulomas but can be rarely seen in granulomas due to other causes especially tuberculosis.[1]

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