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CUTANEOUS SARCOIDOSIS IN A MIDDLE EASTERN MALE: A CASE REPORT

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ABSTRACT

Sarcoidosis is a multi-systemic granulomatous disorder of unknown etiology that can affect multiple organs, with the lungs and intrathoracic lymph nodes being the most involved. The skin is another organ that can be involved in systemic sarcoidosis. However, primary cutaneous sarcoidosis has been reported in about 25% of cases.

Primary cutaneous sarcoidosis is a diagnostic challenge due to its heterogenicity and resemblance of other dermatologic conditions. In such cases clinicopathological correlation plays a crucial role in identification, prognosis and mangement of the disease.

Documentation of case reports allows a better understanding of primary cutaneous sarcoidosis, its presentation diagnostic challenges and patient's manegement. In this case report we review a case of a 44-year-old male patient from middle east who presented with primary cutaneous sarcoidosis. The patient presented to the dermatology department with a non-itchy, erythematous papule without scales at the left shoulder for two months duration. Skin punch biopsy demonstrated an intradermal nodule of non-necrotizing well-defined granulomas. Chest imaging didn't show any pulmonary sarcoidosis. Cutaneous sarcoidosis can present with different manifestations; thus early identification helps in early intervention.

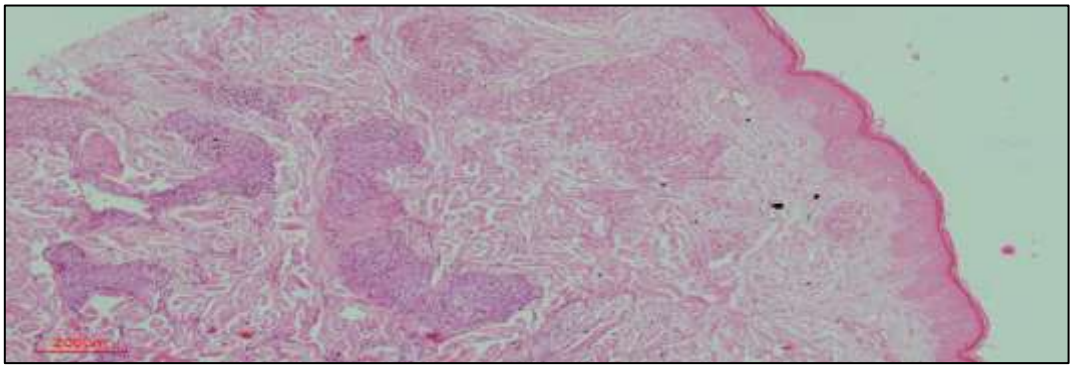
KEYWORD: Sarcoidosis, cutaneous sarcoidosis, granulomatous disorder

INTRODUCTION

Sarcoidosis is a multisystemic, chronic inflammatory disease (1), characterized by the formation of non-caseating granulomas (2). The lungs and the intrathoracic lymph nodes are regarded as the most involved organs. However, involvement of other sites including the eyes and skin might occur. Cutaneous sarcoidosis has been documented in at least 25% of cases (2) and showed equal overall male and female presentation, with females being affacted at older ages compared to males (1). Even though sarcoidosis has been reported worldwide, particularly in African American population, there is limited epidemiological data available regarding its prevalence in Middle Eastern population (3). The etiology of sarcoidosis is unknown, but several factors have been integrated in pathogenesis including genetic susceptibility, immunological and environmental factors (4, 1). A recent study has found the best response in treating these cases with oral corticosteroids, followed by a combination of hydroxychloroquine and topical steroids (5), even though the first-line treatment is initiated with topical and intralesional corticosteroids for localized lesions (1). Although, more recent studies have highlighted the importance of using anti-TNF-α and anti-JAK-STAT inhibitors, more population studies are needed before being assigned for safe use in refractory cutaneous sarcoidosis therapy (6). In this case we report a primary cutaneous sarcoidosis disease in a middle-aged male patient from the Middle East.

Case Presentation

A 44-year-old male patient presented to the dermatology clinic for the first time with a single, erythematous, non-itchy papule with indurated base and no scales over the left shoulder for two-month duration during which the nodule has increased in size. Systemic review was unremarkable for constitutional symptoms. Physical examination was normal. Upon dermatological examination the nodule showed a well-demarcated, erythematous, non-scaly features and measured about 4 mm. Chest CT image was unremarkable, demonstrating both clear lung fields and no evidence of lymphadenopathy or lung involvement. A skin punch biopsy was performed and confirmed the presence of a dermal-based nodule of multiple, well-defined, non-necrotizing granulomas. The granulomas consisted of epithelioid macrophages, surrounded by a lymphocytic cuff with a variable number of multinucleated giant cells (see Figure 1). Additional histochemical stains including Ziehl-Neelsen, PAS and GMS special stains were performed for acid fast bacilli and fungi, respectively, and were negative. The dermatologist prescribed mometasone furoate 0.1% topical cream along a total period of three months. The lesion has been resolved completely after one month of the treatment course.



DISCUSSION

Sarcoidosis is a disease of granulomatous reaction with unknown primary etiology, defined by the presence of well-fomred non-necrotizing granulomas (2). The disease predominantly affects the lungs and lymphatic system but can involve virtually any organ, including eyes, skin, liver, nervous system and others. Cutaneous involvement is present in approximately 25% of sarcoidosis cases and often serves as an early indicator of systemic disease emphasizing the need for comprehensive systemic evaluation in these patients (7,8). Skin lesions are easily accessible for biopsy, allowing for diagnosis without the need to use invasive procedures. Histopathological findings might have a vaiable features ranging from the specific features where non-caseating granulomas are identified to nonspecific lesions, such as erythema nodosum which lacks these characteristic granulomas and are not diagnostic of sarcoidosis (9).

The prevalence of cutaneous sarcoidosis varies between different populations, with African descent populations having the highest incidence (1) A nationwide registry study conducted the United States found that patients with cutaneous manifestations were more likely to be female (81%) and Black (24%) (10).

Sarcoidosis is a relatively rare diseas in the Middle East region (3). However, several studies highlighted its presence across several countries. One study in Saudi Arabia estimated a prevalence of 13 per 100,000 people, with a slight female predominance and age around the fifth decade (11). Similarly, research from Oman and Kuwait has documented cases of sarcoidosis with clinical presentations similar to those seen worldwide, including pulmonary and extrapulmonary manifestations (12). These findings necessitate differential diagnoses of sarcoidosis in the Middle Eastern, despite its rarity.

Clinically, cutaneous sarcoidosis presents with a variety of lesions, including papules, plaques, nodules, and lupus pernio (8). Lupus pernio, characterized by chronic, violaceous, indurated plaques on the face, particularly the nose and cheeks, is strongly associated with chronic disease and poor prognosis (13). The diverse morphology of skin lesions in sarcoidosis has led to its designation as "the great imitator" in dermatology.

Management of cutaneous sarcoidosis depends on several factors, most importantly the severity, extent of skin involvement and the presence of systemic disease. First-line treatment is initiated with topical and intralesional corticosteroids for localized lesions (1) and a more systemic approach, such as oral corticosteroids, hydroxychloroquine, and methotrexate, for more extensive or refractory cases. Recent studies have shown the efficacy of new biologic therapies, such as tumor necrosis factor (TNF) inhibitors like infliximab and adalimumab, and Janus kinase (JAK) inhibitors like baricitinib and tofacitinib, in treating refractory cutaneous sarcoidosis (14), making these targeted therapies promising agents in achieving clinical remission in patients unresponsive to conventional treatments.

CONCLUSION

The case reported the presence of a primary cutaneous sarcoidosis, confirmed by histopathologic confirmation of non-caseating granulomas emphassizing the importance of early detection of cutaneous sarcoidosis. Awareness of such presentations can improve diagnostic accuracy, facilitate easier and more targeted therapeutic approaches, and ultimately enhance patient outcomes.

Patient consent

Verbal informed consent was obtained from the patient for the publication of this case report.

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