

A rare manifestation of systemic sarcoidosis with livedo reticularis–like eruption in a pediatric patient: A case report



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INTRODUCTION

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. It can affect all age groups, but it is relatively rare in children.¹ Clinical manifestations can vary substantially depending on multiple factors, such as the patient's age and the different organs involved. Infants and children younger than 4 years of age usually present with the triad of cutaneous eruption, uveitis, and arthritis, without classic pulmonary disease.^{1,2} Cutaneous sarcoidosis is rare and can manifest with different clinical presentations but rarely with livedo reticularis–like eruption.^{3–5} However, older children have a classic presentation resembling that of adults, which includes involvement of the lungs and lymph nodes predominantly.⁶ It can also be accompanied by generalized constitutional symptoms, such as fever, fatigue, malaise, and weight loss. The diagnosis of sarcoidosis is mainly established by confirming a typical noncaseating granuloma on a tissue biopsy specimen and excluding other granulomatous diseases clinically, microbiologically, and histologically. Other causes of granulomatous disease include infections, such as tuberculosis, autoimmune diseases (eg, Sjögren syndrome and Behçet disease), neoplasms (eg, lymphoma), and other diseases.^{7–9} There is no significant difference between childhood sarcoidosis and the adult type, although most children have a better prognosis.¹ We describe a unique clinical variant of cutaneous sarcoidosis manifesting

as livedo reticularis–like eruption in a pediatric patient with a history of intermittent joint pain, cranial nerve involvement, and uveitis for 1 year.

CASE REPORT

A 13-year-old girl was admitted to King Abdullah Specialized Children's Hospital in Riyadh, Saudi Arabia, with a 1-year history of intermittent fevers, nonspecific skin eruptions, bilateral knee arthritis, facial and abducent nerve palsies, and uveitis. On examination, she had reticulated erythema that was blanchable in some areas, was not affected by heat or cold, and involved the trunk and upper and lower extremities. In addition, she had uveitis, bilateral knee active arthritis with effusion, and decreased range of motion (Fig 1).

The initial laboratory workup was remarkable for only a low white blood cell count ($1.27 \times 10^9/L$ [reference range, $4-12 \times 10^9/L$]). Other basic metabolic laboratory values were normal (liver function, renal function, lipid profile, and general electrolytes). Positron emission tomography–computed tomography found hepatosplenomegaly with diffuse hypermetabolic activity along with hypermetabolic lymph nodes in the internal mammary region, and left paratracheal and left supraclavicular lymph nodes, raising the suspicion for lymphoma. Results of the lymph node biopsy found necrotizing granulomatous lymphadenitis and negative acid-fast bacilli special stain. Results of the bone marrow

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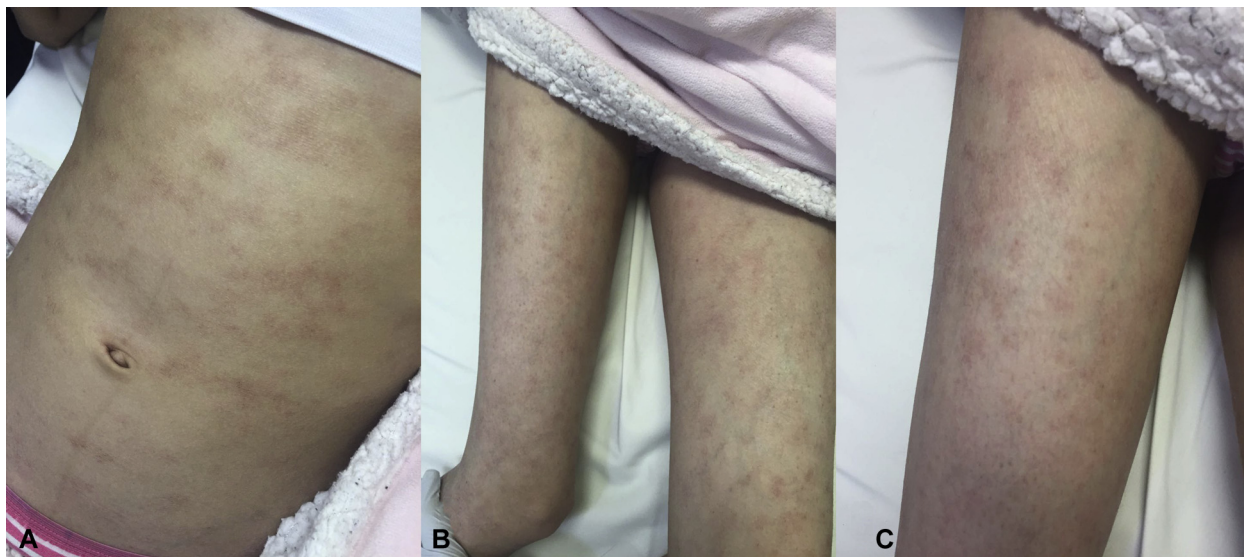


Fig 1. Clinical presentation of cutaneous sarcoidosis shows livedo reticularis–like eruption over the patient’s abdomen, lower chest, and lower limbs.

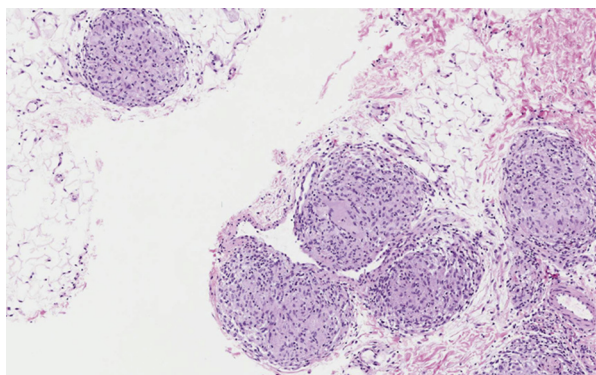


Fig 2. Microscopic examination of the livedo reticularis–like eruption shows multiple noncaseating granulomas surrounding the blood vessels.

biopsy from the right posterior iliac crest showed hypocellular bone marrow for age with no definite evidence of abnormal involvement. Multiple punch biopsies were taken from the left forearm and left thigh; the results of which showed subepidermal noncaseating granulomatous dermatitis with perivascular and periadnexal distribution with negative acid-fast bacilli special stain (Ziehl-Neelsen stain) and tuberculosis polymerase chain reaction analysis (Fig 2).

Further laboratory investigations found a high angiotensin-converting enzyme level (510 U/L

[reference range, 29-112 U/L]) and a high tumor necrosis factor- α level (163 pg/nL [reference range, up to 8.1 pg/nL]). Additional imaging studies were performed, including chest radiography and echocardiography, the findings of which were normal. Given the multisystem involvement with tissue biopsy findings of noncaseating granuloma of the skin, high angiotensin-converting enzyme level, and exclusion of other causes of granulomatous inflammation, a diagnosis of systemic sarcoidosis was made. The patient was started on methylprednisolone (2 mg/kg/d), methotrexate (15 mg/wk), and infliximab (5 mg/kg), and she showed remarkable improvement (Fig 3).

DISCUSSION

Sarcoidosis is sometimes difficult to diagnose, and the involvement of multiple medical specialties may be needed to make the correct diagnosis. A diagnosis of sarcoidosis should be considered when there is evidence of noncaseating granuloma in histopathologic samples. The most common cutaneous sarcoidosis variants include papular sarcoidosis, plaque sarcoidosis, lupus pernio, and scar-associated sarcoidosis.¹ However, cutaneous sarcoidosis manifesting with livedo reticularis–like eruption is very rare. There are a few reports in the literature linking livedo reticularis–like eruption with systemic sarcoidosis.³⁻⁵

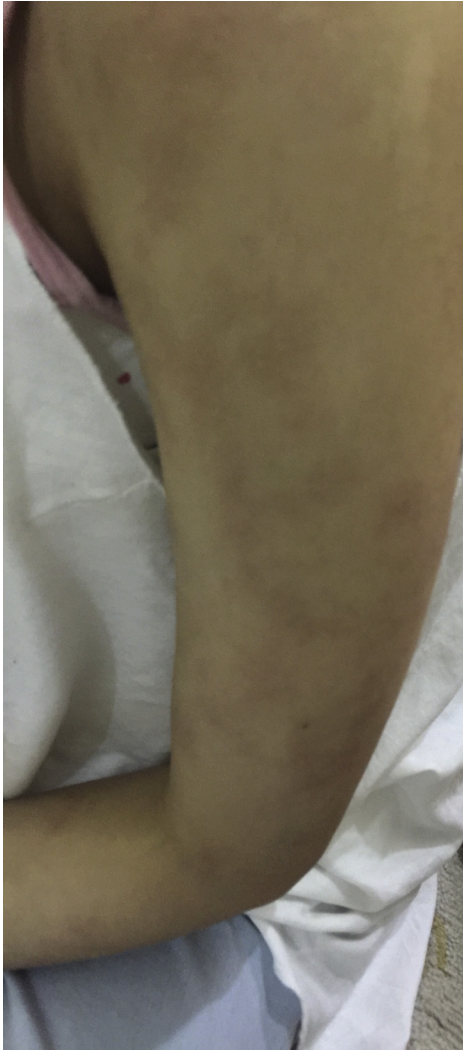


Fig 3. Remarkable improvement is shown after starting the patient on methylprednisolone, methotrexate, and infliximab.

Cutaneous sarcoidosis manifesting with livedo reticularis–like eruption is very rare and can be attributed to either granulomatous vasculitis or obliterative changes around the blood vessels by the granulomas. Given there have been a few reports, livedo reticularis–like eruption should be considered a clinical variant of cutaneous sarcoidosis.

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