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Cutaneous Sarcoidosis with Huge Muscle Hypertrophy

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Abstract

Observation: Sarcoidosis is a multisystem granulomatous disease of unknown etiology with variable manifestations, which may affect virtually any organs including skin and muscle. Cutaneous sarcoidosis without involving any other system is quite common. Muscular sarcoidosis is a rare entity, and the combination of huge muscular swelling with cutaneous involvement without any other visceral affection is rare still. We report herein a case of sarcoidosis that presented with a chronic swelling of deltoid muscle with papules in a generalized distribution.

Introduction

Sarcoidosis is a multisystem granulomatous disease in which multiple organs are involved. Between 20% and 35% of patients with systemic sarcoidosis have skin lesions, but cutaneous sarcoidosis can also occur without systemic disease and the extent of any cutaneous lesions does not correlate with the extent of systemic disease. Various forms of cutaneous sarcoidosis have been described which includes erythema nodosum, ery th ematopapular, scar sarcoidosis, papular ('small nodular'), erythrodermic, nodular, annular, angiolupoid, subcutaneous, plaque and lupus pernio [**1**]. Muscle involvement occurs rarely in comparison to cutaneous involvement, and usually it is concurrent with other systemic involvement; though isolated myopathy have also been reported. There a re several forms of muscular involvement in sarcoidosis; namely polymyositis, muscle weakness and tenderness, tumour-type muscle sarcoidosis, steroid-induced myopathy, small fibre neuropathy, reduced physical activity induced myopathy, palpable myopathic nodules and a chronic progressive myopathy; all are seen uncommonly in clinical practice.[**1,2**] Random muscle biopsies in patients with sarcoidosis showing granuloma are found in about 50–80% cases [**3, 4, 5, 6**]. We present here a case of sarcoidosis because of an un usual presentation of huge deltoid muscl e swelling which was present for long time along with subsequent appearance of cutaneous papular sarcoidosis.

Case Report

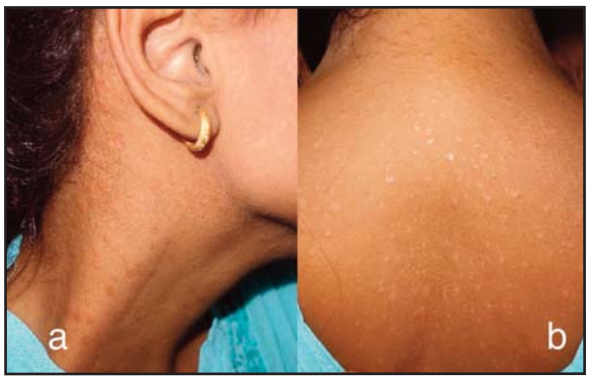
A 21-year-old female presented with multiple small elevated skin lesions all over the body and huge swelling of the right arm. Her ailments started about four years ago when she first noticed tightening of her clothes, on the right arm. The swelling gradually increased in size with feelings of warmth over the area. She sought consultation with a number of physicians, undergone thorough investigation and treated with various drugs without any beneficial effect. There was no history of redness, burning, itching, or dryness of eyes, neither there was any history of shortness of breath, abdominal pain, pruritus or yellowish discolora-

tion of urine. Features suggestive of peripheral neuropathy as well as bleeding episodes like epistaxis or hematuria was also absent. Past history and family history was unremarkable. History of long term treatment with high dose systemic corticosteroid was given by the patient.

Bilateral pedal edema and cushingoid features were evident on general examination. Cutaneous examination showed numerous shiny dome-shaped erythematous to slightly violaceous scaly papules in a generalised distribution. (**Figures 1a and b**) Right sided deltoid muscle was grossly hypertrophied and firm (**Figure 2**), having increased local temperature. Hairs, nails and mucosae were apparently normal.

Routine hematological and biochemistry panel including muscle enzymes and serum angiotensin converting enzyme were within normal limits. Chest X-ray and electrocardiogram were also non contributory. Mantoux test was negative. Color Doppler study of upper extremity shows normal triphasic flow with good peak systolic velocity in brachial artery while sluggish flow in radial and ulnar artery with reduced peak systolic velocity. MRI suggested marked signal changes involving deltoid, supraspinatus and infraspinatus with loss of muscle planes. Histopathology from a representative skin lesion revealed numerous epithelioid cell granulomas with a good number of Langhan's giant cell and scanty lymphocytes (**Figure 3**). Muscle biopsy also had similar findings.

On the basis of history, evolution of the disease and investigatory findings a final diagnosis of cutaneous sarcoidosis with tumor like myopathy was made. Treatment was started with oral hydroxychloroquine and methotrexate. Cutaneous lesions subsided satisfactorily but muscular swelling did not show encouraging response and regular follow up is still going on.



Figures 1a and b. Numerous shiny dome-shaped erythematous to slightly violaceous scaly papules

Discussion

Sarcoidosis is a systemic disease that can affect any organ in the body. One of the most common extrapulmonary organs affected is the skin [**7**]. Cutaneous lesions may occur before, coincident with, or after systemic involvement. The most common presentation is the papular form and these lesions occur most commonly on the face and neck [**8**]. Our patient presented with papular lesions in generalized distribution.

Asymptomatic muscle disease in sarcoidosis occurs with a much greater frequency than symptomatic disease. Studies regarding isolated sarcoid myopathy without prior or concurrent organ involvement are lacking [**9**]. Our patient, though first noticed few papules over her right arm, ignored it and became concerned only after the right arm get swelled and papules spread subsequently all over the body.

While the granulomas of systemic sarcoidosis are frequently observed histologically in skeletal muscles, they are not often associated with muscle symptoms or signs such as weakness, wasting, tenderness, or myalgia. Our patient presented with myopathy in the form of huge, tumor like hypertrophy of the deltoid muscle. Both muscle as well as skin biopsy specimen showed numerous epithelioid cell granulomas in histopathology. Asymptomatic muscle involvement in sarcoidosis has been found almost exclusively in the early stages of the disease [**10,11**].

At the beginning, our patient also had asymptomatic swelling of the deltoid muscle before she noticed extensive cutaneous involvement which suggested that initially the disease activity was at an early stage and gradually a ppeared in full- blown form. Symptomatic involvement of muscles and myopathic electromyographic abnormalities are rare and only found in chronic sarcoidosis. Symptoms other than raised local temperature of arm were absent in this young female and features suggestive of chronic sarcoidosis were also not found. Occasional presentation in any stage with acute myalgia and muscle tenderness had also been reported [**5**]. but such complaints were also not encountered in our patient till date.

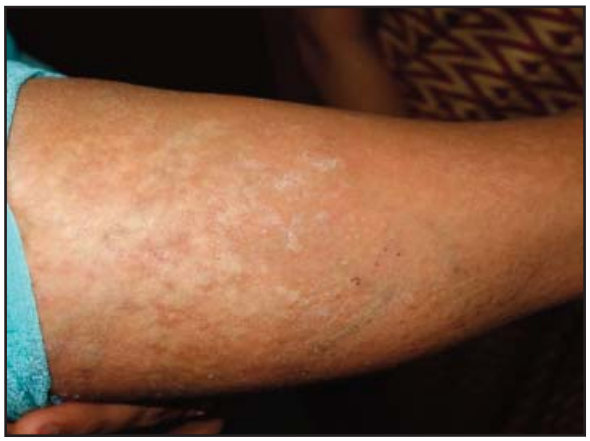


Figure 2. Deltoid muscle was grossly hypertrophied and firm

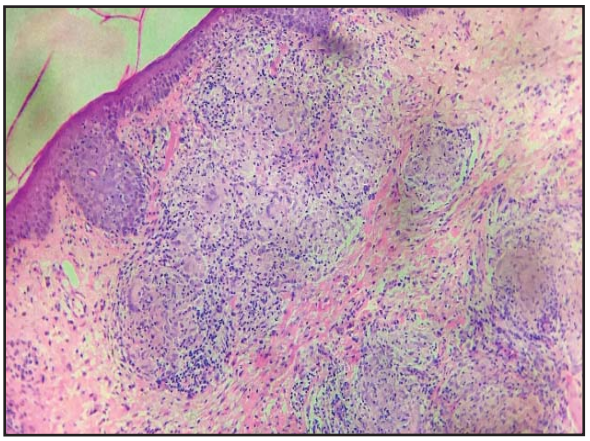


Figure 3. Numerous epithelioid cell granulomas with a good number of Langhan's giant cell and scanty lymphocytes

The myopathic type occurs predominantly in elderly females with a painful bilateral involvement and often results in muscle weakness, atrophy and even muscle contracture [**6**]. The rarest type reported is the acute sarcoid myositis [**4,6**]. This is usually found in younger p atients with proximal muscle weakness mimicking acute polymyositis. These features were lacking, too, in our patient.

Gallium scintigraphy is considered the main method6 that can demonstrate muscular involvement. Magnetic resonance imaging (MRI) can reveal a broad range of musculoskeletal abnormalities including focal and diffuse muscle lesions and soft-tissue masses which is helpful for diagnosis, but small lesions may be overlooked. In our case, MRI suggested marked signal changes involving deltoid, supraspinatus and infraspinatus with loss of muscle planes which is corroborative with the reported radiological changes of sarcoidal myopathy.

Skin lesions of sarcoidosis are usually asymptomatic. Cosmetic disfigurement is the most common complaint. Treatment of sarcoidosis skin lesions is not required if they are stable and not of cosmetic importance. Localized lesions may be treated with topical corticosteroids. If the skin lesions are diffuse or not responsive to topical agents, systemic therapy is required. Corticosteroids are the drug of choice for the treatment of skin sarcoidosis. Effective alternative agents for skin sarcoidosis include methotrexate, hydroxychloroquine, chloroquine, thalidomide, and tetracycline derivatives [**7**].

There are currently no studies available on the treatment of sarcoid muscle involvement. For patients with muscle weakness, controlled physical training programmes are advocated [**12**]. Sarcoid muscle involvement is usually asymptomatic and resolves spontaneously [**3**]. In severe cases, glucocorticoids and immunosuppressive drugs are given [**13**]. Before coming to us, this patient was treated with high dose systemic corticosteroid for several months without any appreciable benefits. Moreover, she had developed steroid induced posterior subcapsular cataract and cushingoid features. Hence, we started treatment with hydroxychloroquine and methotrexate. Significant improvement was noted for cutaneous lesions and regular follow up is still going on.

Conclusion: We presented here a case of cutaneous sarcoidosis with huge muscle involvement without any other systemic features. Though papular cutaneous sarcoidosis is fairly common, tumor-like muscle swelling a ssociated with extensive cutaneous involvement is extremely rare in the English literature, and after extensive PubMed search we could not locate any case of sarcoidosis having similar combination of cutaneous and muscular features.

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