Is Subcutaneous Sarcoidosis a Marker for Systemic Disease?

History
A 40-year-old diabetic woman presented with slightly painful multiple subcutaneous nodules on extensor aspect of both forearms [Figure 1a] with dactylitis of right index and ring fingers [Figure 1b] of 2-month duration. There was no regional lymphadenopathy. Systemic examination was normal. All relevant hematological, serological, and biochemical investigations including serum calcium and phosphorus were within normal limits. Mantoux was nonreactive. X-ray chest revealed suspicious bilateral hilar lymphadenopathy. Biopsy from subcutaneous nodules showed multiple epithelioid granulomata deep in the dermis and within the adipose tissue [Figure 2a and b].

What is your diagnosis?

Discussion

Diagnosis: Subcutaneous sarcoid

Our patient presented with asymptomatic subcutaneous nodules with dactylitis and asymptomatic bilateral hilar lymphadenopathy. The histology of subcutaneous nodules showed multiple epithelioid granulomata deep in the dermis and within the adipose tissue. These granulomata were naked and confluent. Special stains like Fite–Faraco, Ziehl–Neelsen, and periodic acid schiff (PAS) were negative. Tissue cultures for acid fast bacilli and fungal infections were also negative. The histopathological diagnosis was sarcoidal granuloma. These findings led us to investigate the case further to look for systemic disease. Serum angiotensin-converting enzyme levels were elevated with 82 U/l (normal: 8–52 U/l). CT chest showed multiple enlarged pre-tracheal, right paratracheal, pre-carinal, left para-aortic, and bilateral hilar lymph nodes. Based on clinical, histopathological, and imaging studies, we made a diagnosis of subcutaneous sarcoidosis (SCS) with dactylitis and asymptomatic systemic disease.

SCS is a distinct subtype of cutaneous sarcoidosis. It was initially considered as Darier–Roussy sarcoid. This usage caused confusion. In 1984, Vainsencher and Winkelmann[1] cleared this confusion and preferred the term SCS. They proposed the following to diagnose SCS: (1) typical painless subcutaneous nodules principally on the extremities; (2) typical histopathological features of naked epithelioid granulomas with scanty lymphocytes, localized to the subcutaneous tissue; and (3) association with early benign hilar adenopathy. SCS occurs mostly in women, most often in the third and fourth decades. Unlike other specific cutaneous lesions of sarcoidosis like plaques, papules, annular, lupus pernio, lichenoid, psoriasiform, scar sarcoidosis, and erythroderrmic form, subcutaneous nodules have been more frequently (1.4–6%) associated with systemic disease.[2–4] Several studies reported the association between SCS and systemic disease.[5–7] The systemic component consisted of hilar lymphadenopathy, lung infiltrates, uveitis, dactylitis,[8] abnormal pulmonary function tests, lacrimal gland involvement, and hepatosplenomegaly.
Histopathologically, SCS shows an inflammatory infiltrate composed of non-caseating granulomas involving fat lobules that are usually sharply demarcated in the dermo-hypodermal junction. The granulomas are small, uniform in size, and mainly composed of epithelioid cells with a discrete amount of multinucleate giant cells and scanty lymphocytic components. SCS can be easily differentiated from other subcutaneous nodular lesions like erythema nodosum, epidermal cysts, multiple lipomas, calcinosis, rheumatoid nodules, morphea, cutaneous metastasis, tuberculosis, and deep mycotic infections due to the presence of naked granulomas in the adipose tissue.[2]

In contrast to the skin lesions in erythema nodosum, the lesions in SCS are non-tender and flesh colored which may persist for much longer periods of time and may be free of notable lymphocytic infiltration.

The overall prognosis of SCS lesions is good. The majority of SCS lesions have spontaneous resolution including the systemic disease in 2–3 years. For patients with severe systemic involvement or disfiguring skin lesions, systemic steroids are the first-line therapy. Alternate drugs like methotrexate and hydroxychloroquine have been reported helpful. Our patient was treated with oral prednisolone 40 mg daily in tapering doses over a period of 3 months. Complete resolution of nodules was observed after 8 weeks. During the follow-up period of 6 months, there was no recurrences of lesions.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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