

Asymptomatic swelling in a 60-year-old scar

A 65-year-old lady presented with two hard swellings over the forehead for two years before seeking dermatology consultation. She had a penetrative injury in a linear fashion on the forehead when she was 5-year-old. The wound healed with secondary intention leaving behind a single linear scar. Two years back, she noticed a swelling underneath the scar. The swelling was gradually progressive in size, hard to feel, and was associated with slight stretching and purplish discoloration of the overlying scar. Later, she developed another round swelling similar to the previous one in the vicinity underneath an unrelated scar. She had exertional dyspnea of insidious onset for six months not associated with any other cardiac or pulmonary symptoms. There was no history of fever, loss of weight or appetite, arthralgia, or other systemic complaints. She did not have pain, itching, ulceration, or finger-like extensions on the scar. She was a known diabetic which was controlled with metformin.

On cutaneous examination, she had two distinct lesions over the left side of the forehead [Figure 1]. The larger lesion was of dimension 8 × 2 cm, disposed



Figure 1: Subcutaneous swelling extending beyond the margin of overlying stretched scar

linearly. The lesion was subcutaneous, hard, nontender, minimally mobile, and extending beyond the margin of overlying stretched scar. A circumscribed similar swelling measuring 2 × 2 cm was noted in the vicinity. Histopathological features of skin biopsy specimen are shown in Figures 2 and 3.

WHAT IS YOUR DIAGNOSIS?

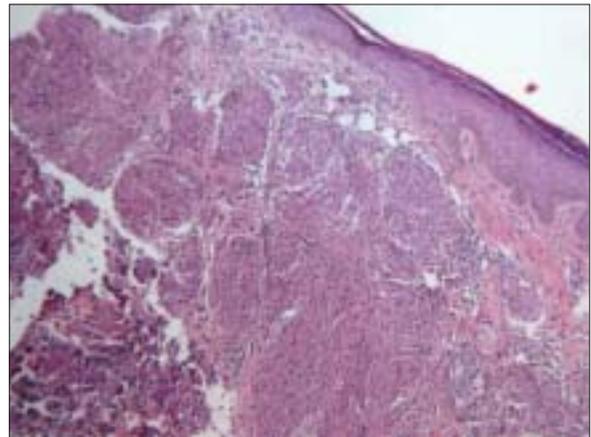


Figure 2: Multiple nonnecrotizing granulomas comprising of epithelioid cells and Langhans giant cells without lymphocytic cuffing, (H and E, x280)

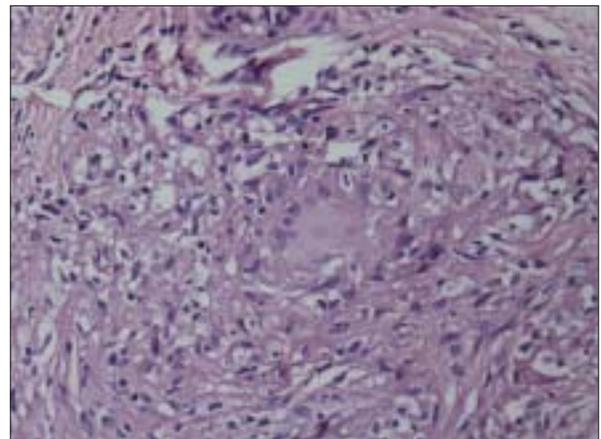


Figure 3: Close-up view, (H and E, x550)

How to cite this article: De D, Narang T, Dogra S, Radotra BD, Kanwar AJ. Asymptomatic swelling in a 60-year-old scar. *Indian J Dermatol Venereol Leprol* 2009;75:97-9.

Received: October, 2007. **Accepted:** January, 2008. **Source of Support:** Nil. **Conflict of Interest:** None declared.

Diagnosis: Scar sarcoidosis

Hematoxylin and eosin staining of a deep punch biopsy specimen taken from the linear lesion showed multiple non-necrotizing granulomas comprising of epithelioid cells and Langhans giant cells without lymphocytic cuffing, consistent with sarcoidosis. There was no nerve twig invasion. No organism was found in special stains including Ziehl Neelsen, Fite, and periodic acid-Schiff stain (PAS).

Her complete blood cell counts, liver and renal functions tests, and serum calcium and phosphate values were normal. Routine urine examination was normal. Chest X-ray showed perihilar shadows. Mantoux test was negative. Pulmonary function test revealed restrictive pattern. Ophthalmologic examination was unremarkable. Serum angiotensin converting enzyme level was twice the normal value.

DISCUSSION

Sarcoidosis is a multisystem disorder of unknown origin characterized by the accumulation of lymphocytes and mononuclear phagocytes that induce the formation of noncaseating epithelioid granulomas with secondary derangement of normal tissue anatomy and function. Cutaneous involvement occurs in about 20–35% patients of sarcoidosis, with only 2% having exclusive involvement of the skin.^[1] Most often, the cutaneous involvement occurs at outset of the disease.

Cutaneous sarcoidosis can manifest with specific and nonspecific lesions; erythema nodosum being the only nonspecific manifestation of sarcoidosis. The specific lesions, lupus pernio and plaques are associated with more severe systemic involvement and more chronic course, while nonspecific erythema nodosum is the hallmark of acute and benign disease.^[2]

Scar sarcoidosis is a specific manifestation of sarcoidosis, occurring in 5.4–13.8% patients of cutaneous sarcoidosis in adults.^[2] It has been found to occur at the scar sites of pseudofolliculitis barbae, Kveim test biopsy site, tuberculin test site, tattoos, venipuncture, mechanical trauma, radiation, herpes zoster, and following hyaluronic acid injection. How the old scars, as reported up to 60 years after their onset, develop into a nidus of sarcoidal granuloma is not known. They may appear early before pulmonary involvement or parallel chronic systemic findings.^[2] Previous contamination of scar with foreign material

possibly at the time of trauma has been suggested to be an underlying cause. Persistent granulomatous reaction to vaccines and allergen-extract preparation for desensitization are usually attributable to the adsorbing agent aluminium hydroxide.

Scar sarcoidosis is characterized by reoccurrence of activity at the site of previous scar and clinically represented by swelling, erythema, and purplish red hue that subsequently turn brown with the conspicuous absence of itching.^[3] This factor is important in differentiating between scar sarcoidosis from hypertrophic scar and keloid, which are the clinical mimickers. It occurs during the acute eruptive phase of sarcoidosis paralleling changes in the lung or can occur at the late phase as a sign of exacerbation in a previously quiescent sarcoidosis.

Chronic active systemic sarcoidosis along with iritis has been estimated to coexist in 84.6% of scar sarcoidosis patients in contrast to around 30% patients with other form of cutaneous sarcoidosis having systemic involvement.^[4] Even though there is no systemic involvement at the onset, 30% of patients may develop systemic manifestations in long-term follow up.^[2]

In the absence of systemic involvement, scar sarcoidosis can be managed with topical or intralesional corticosteroids. For those patients who have systemic involvement, hydroxychloroquine is the first-line drug of choice. Prednisolone and methotrexate are the second-line choices. Other drugs which can be used are tetracyclines, isotretinoin, allopurinol, thalidomide, leflunomide, pentoxifylline, and infliximab, etc.

Our patient was treated with intralesional triamcinolone acetonide (20 mg/ml) injections every three weeks, and hydroxychloroquine 400 mg/ day. With this regimen, she had improvement in her skin lesions, though systemic problems were persisting. Oral prednisolone was then started at a dose of 30 mg/day, with which she had improvement in pulmonary symptoms after four weeks of treatment.

In conclusion, scar sarcoidosis has two important clinical implications associated with it. First, it can be very easily confused with hypertrophic scar or keloid. Second, scar sarcoidosis is an important clue to the significant risk of systemic involvement due to disease.

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