Rare disease

Scar sarcoidosis with bone marrow involvement and associated musculoskeletal symptoms

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Summary

Sarcoidosis affects the lungs most commonly and can present with cutaneous lesions. It can also involve the bone marrow in rare instances, often presenting with non-specific symptoms such as fever and malaise, with occasional haematological abnormalities. The authors present the case of a 41-year-old caucasian female who was diagnosed with scar sarcoidosis, but who also reported fatigue, night sweats and polyarthralgia. No haematological abnormalities or lung involvement were detected. A magnetic resonance scan of the spine, performed due to disc prolapses, demonstrated areas of bone oedema and stress response that were reported as a possible sarcoid infiltration of the bone marrow. An improvement occurred after 8 weeks, despite conservative treatment. Bone marrow involvement in sarcoidosis is rarely reported, and was an accidental finding here. Due to the non-specific presentation, cases such as this are potentially underdiagnosed and should be considered in patients with systemic symptoms, regardless of their haematological status.

BACKGROUND

Sarcoidosis is a multisystem disorder characterised histologically by non-caseating granuloma formation.^{1,2} Its aetiology remains unknown and it can affect all races, ages and both sexes.^{1,2} The disorder most commonly affects the lungs (>90%), skin, lymph nodes and eyes.^{1,2} A specific but relatively rare presentation of the disease is the reactivation of preexisting scars, which demonstrate histological

features typical of sarcoidosis.¹⁻³ Bones are infrequently affected (1–13% of cases), with involvement of the vertebrae being uncommon.^{4,5} Similarly, sarcoidosis affects the bone marrow rarely, with occasional haematological abnormalities.^{6,7} Patients can also present with non-specific systemic symptoms such as malaise, lethargy, night sweats and fever of unknown origin, which may be indicative of bone marrow involvement.^{1,6,7} We present the case of a

Figure 1 Skin biopsy of inflamed scar demonstrating non-caseating granulomas. H&E, original magnification x100.

Figure 2 (a) MRI spine T1 sequence demonstrating hypointense lesions in multiple vertebral bodies. (b) The vertebral abnormalities reflected as high signal intensities on the MRI spine STIR sequence.

patient with scar sarcoidosis, musculoskeletal symptoms and bone marrow involvement.

CASE PRESENTATION

A 41-year-old caucasian female presented with inflammation of two previously atrophic scars. She also reported a 2-month history of night sweats, fever, fatigue and polyarthralgia. Her medical history included chronic back pain due to lumbar disc prolapses. Other than the contraceptive pill, she was not taking any regular medications and did not report any respiratory or visual symptoms.

On examination both scars were erythematous, elevated, hot and painful to touch. One was located on the right knee resulting from a fall when she was 8-years-old, and the other was on the right lower back, resulting from the excision of a lipoma at the age of 20. There was no peripheral lymphadenopathy or organomegaly. A skin biopsy was performed which demonstrated granulomatous

inflammation consistent with sarcoidosis (figure 1). Stains for fungi and mycobacteria were negative. A chest radiograph revealed a slightly prominent left hilum but a subsequent high resolution CT scan indicated no abnormality. Pulmonary function tests were within normal ranges. Laboratory tests showed raised levels of C-reactive protein at 18 (<8 mg/l) and rheumatoid factor at 62 (<20 IU/ml), with the level of ACE at the upper limit of normal. Full blood count, renal, liver and thyroid function tests and calcium were all within the normal ranges. An ophthalmology review revealed sarcoid granulomas in the lower tarsal conjunctivae bilaterally.

Due to severe worsening of the lower back pain, MRI of the spine was performed. Although this revealed no change in the L4/5 and L5/S1 disc prolapses compared with previous scans, abnormalities in the bone marrow were indicated. There were hypointense lesions near the end plates and posterior vertebral bodies on the sagittal

Figure 3 (a) Sarcoidosis with infiltration of a scar. Image reprinted with permission from Medscape.com, 2011. Available at: <http://emedicine.medscape.com/article/1123970-overview>. (b) Sarcoidosis with infiltration of 50-year-old scars in a 75-year-old woman. Image reprinted with permission from dermatology online journal. Available at: http://dermatology.cdlib.org/1411/letters/scar_sarcoid/larangeira.html

T1 sequence, which appeared as high signals on the short tau inversion recovery (STIR) sequence (figure 2a,b). A bone marrow biopsy was offered but was declined by the patient. Protein electrophoresis and urine Bence–Jones protein were negative.

The diagnosis of scar sarcoidosis with systemic features including bone marrow involvement was made. Treatment was conservative with non-steroidal anti-inflammatory medications.

OUTCOME AND FOLLOW-UP

The patient improved over the course of 6 months, with complete resolution of the cutaneous and ophthalmological signs, and improvement in the systemic symptoms. A repeat MRI of the spine, 8 weeks after the original scan, revealed a reduction in the bone marrow abnormalities with a decrease in oedema on the sagittal STIR sequence.

DISCUSSION

Cutaneous manifestations of sarcoidosis affect between 9–37% of patients with scar sarcoidosis reported in 2.9–29% of these cases.^{1–3 8} As scar sarcoidosis may be the only skin manifestation of systemic disease, as in the case described here, it is of great diagnostic importance (figure 3).^{3 8} The discovery of bone marrow involvement was an accidental finding when investigating the known lumbar discs prolapses in the patient. Bone marrow involvement is estimated to affect only 10–17% of patients with sarcoidosis,⁹ but is reported to be associated with skin lesions in 50% of these cases.⁶ Although haematological abnormalities such as anaemia and leucopaenia are expected and have been reported,^{2 6 7 9} bone marrow involvement can present with normal haematological parameters,^{6 7} as was the case here. In terms of imaging, MR scanning is the most sensitive modality for detecting skeletal sarcoidosis, especially in cases of axial skeleton involvement including bone marrow where the lesions are demonstrated by a decreased signal intensity on T1 sequences and increased signal intensity on STIR, T2 and fat-saturated

proton-density sequences.^{4 5} This is consistent with the MR results in our patient. However, these changes are non-specific and the differential diagnosis could include osseous metastases, lymphoma, myeloma and disseminated infections such as tuberculosis.⁵ Although a bone marrow biopsy would provide a definitive diagnosis,⁴ our patient declined the procedure. The subsequent reduction in bone marrow abnormalities on repeat MR imaging, concomitant with the clinical improvement, further corroborated the diagnosis of bone marrow sarcoidosis. No specific data on prognosis of sarcoidosis associated with bone marrow involvement is available. A series of five cases⁷ revealed improvement without any specific therapy in two, and with oral steroids in three patients. Single case reports showed positive results of treatment with immunosuppressive medications.⁹

In conclusion, this case demonstrates a rare presentation of cutaneous sarcoidosis, in the form of scar sarcoidosis, with the accidental finding of bone marrow involvement. Given the non-specific symptoms of bone marrow sarcoidosis, it may be underdiagnosed. Therefore, it is suggested that bone marrow sarcoidosis should form a part of the differential diagnosis in patients presenting with systemic and non-specific symptoms, regardless of their haematological status. This could be further assessed by MRI, although a definitive diagnosis may require a bone marrow biopsy.

Learning points

- ▶ Scar sarcoidosis is a relatively rare but specific cutaneous presentation of sarcoidosis.
- ▶ Bone marrow sarcoidosis may present with or without haematological abnormalities but systemic symptoms may indicate the need for further investigations.
- ▶ MR imaging can be helpful in diagnosing bone marrow sarcoidosis, although other possibilities include bone metastases, lymphoma, myeloma and infections.

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Competing interests None.

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