Rare disease

Sarcoid myositis: a rare presentation

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Summary

A 50-year-old lady with a background of Perthes disease and multiple bilateral hip revisions presented with symptoms of right hip pain. She was unable to raise and weight bear on the right leg. Hip examination revealed limited flexion and extension to 15–20° due to pain. Her blood tests and x-rays showed no obvious acute problems. She was treated with mobilisation and analgesia, and had a bone scan that revealed no acute issues. Further examination revealed she had generalised weakness in her upper arm. A creatine kinase marker showed a level of over 2000. Therefore, an MRI and a muscle biopsy were done. A diagnosis of sarcoid myositis was made and the patient was put on steroids. She was able to make a full recovery. This case highlights the consideration of rare diseases in more highly complex cases.

BACKGROUND

Generally, sarcoidosis is identified clinically in patients with foggy vision or mediastinal lymphadenopathy, but muscular pain may be an infrequently observed initial symptom.

CASE PRESENTATION

A 50-year-old lady was admitted to our hospital with a painful right hip after having had a mechanical fall 3 days before. She normally mobilised with a stick but was unable to weight bear on the leg. She had a total hip replacement for a fractured neck of femur 5 years previously at this hospital. Her medical history included hypertension, Perthes disease of both hips, a right hip arthodesis and pins 35 years ago, and was being treated at the Stanmore Hospital for continuing thigh pain (figure 1). She was on ramipril and had no known medical allergies.

On general examination, there was nothing remarkable to note. Hip examination showed a shortening of the right

leg length by 2 cm, internal and external rotation limited to 20°, flexion to 80°, abduction painful to 10°. These were all limited due largely to pain. Tone and reflexes were otherwise normal.

INVESTIGATIONS

The x-rays of her spine and right hip showed no obvious acute problems. Blood, including full blood count, urea and electrolytes were normal. To eliminate the possibility of a bone infection, a bone scan was requested, the result being that there were no abnormalities detected. Other explanations for the pain were explored.

Further examination of the patient identified that she had restricted movement (40° abduction) of both shoulders, which was reviewed by the rheumatologist and diagnosed as frozen shoulder. Added to the fact that she had butterfly rash on her face led to further investigations, including rheumatological antibodies and creatine kinase. The results of

Figure 1 X-ray of pelvis: right, total hip replacement and left, hip pins.

Figure 2 MRI of patient's legs showing atrophy of muscle. Highlight shows replacement of muscle tissue with connective tissue.

the test on antibodies were negative, but the creatine kinase was at the level of 1679 (2 weeks after the fall). Other blood parameters were unremarkable, including troponin, full blood count and C reactive protein (CRP). Her erythrocyte sedimentation rate (ESR), however, was raised at 53, her dipstick showed myoglobinuria, and her creatine kinase remained persistently elevated at around 2000.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included lupus, connective tissue disorder and myositis. She was referred to the rheumatologist who organised an MRI (figure 2), an EMG (results showed abnormal signals) and a muscle biopsy of the leg. The muscle biopsy showed fatty infiltration, atrophy of muscles and non-caseating granuloma. Although her CRP was normal, she had a raised ESR of 53. A diagnosis of sarcoid myositis was made.

Following this, the patient had a CT of the chest, which did not show bilateral lymphadenopathy, and the respiratory function tests were normal.

TREATMENT

The treatment of choice for sarcoid myositis was steroids.

OUTCOME AND FOLLOW-UP

The treatment allowed the patient to regain muscle strength in her upper arm and legs. The pain has been minimised, her mobility has also improved and her rehabilitation may eventually allow her to return to work. Her blood markers at this stage showed an ESR of 16 and creatine kinase within normal limits.

DISCUSSION

Sarcoid myositis is a systemic disorder of unknown cause.¹ It is characterised by non-caseating granulomas.² It can be classified into three types: chronic myopathy, palpable nodules and acute myositis. Chronic myopathy is a condition wherein the disease has a steady and slow progress. In this condition, the patient has proximal myopathy bilaterally and symmetrically, and the creatine kinase levels are normal.

The second type is palpable nodules, where the patient presents with painful nodules on the muscles of the extremities, and muscle biopsy specimens reveal non-caseating granulomas.³ This type of sarcoid myositis is not associated with muscle weakness.

The third type is the rarest manifestation that this patient presented with – acute sarcoid myositis.⁴ There have only been 20 acute cases reported in the literature. It usually presents in females aged 40–55 years. Most patients report acute or insidious respiratory problems, as well as proximal muscle weakness. It mimics polymyositis. It usually involves chronic self-limited episodes of relapse and remission of symptoms. Investigations reveal a creatine kinase of around 2000, and the condition itself is diagnosed via MRI and muscle biopsy. The treatment of choice is steroids.⁵

Our case is different from most other published cases in the literature in that the patient had not been diagnosed with sarcoidosis. The patient had no respiratory symptoms, and the chest x-ray did not show hilar lymphadenopathy. It was also unusual that it was initially thought for many years to do with the underlying hip prosthesis.

The patient is doing better with regard to her condition. However, certain complications as a result of the diagnosis must be monitored. Complications with regard to the condition include respiratory complications.⁶ She now has regular lung function tests and chest follow-up. Complications that are important as a result of the steroid treatment include osteoporosis. This is particularly important given her orthopaedic issues. She is currently being regularly followed up by the orthopaedic and rheumatology team, and is on regular bisphosphonates.

Learning points

- ▶ Clinical history and examination are the most important tools in making a diagnosis.
- ▶ There should be early involvement of the multidisciplinary team in complex cases.
- ▶ Sarcoidosis may present with the symptoms of muscular pain.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Ahmad Z, Skinner J, Field R. Sarcoid myositis: a rare presentation. *BMJ Case Reports* 2010;10.1136/bcr.02.2010.2759, date of publication

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