CASE REPORT

An unusual case of steroid responsive idiopathic ectopic calcification

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Abstract

This case explores an unusual calcified lesion of the hand and its dramatic response to steroids. A 30-year-old lady presented to rheumatology with a 1-year history of swelling on the radial side of her right middle metacarpophalangeal joint. Over a 2-week period, she had developed swelling throughout her right hand. She was treated with intramuscular methylprednisolone injection and a weaning course of prednisolone. A series of photos and X-rays demonstrates the resolution of swelling and calcification after steroid treatment. This case reports a chronic calcified mass associated with an acute inflammatory episode in the hand. This is likely to represent rupture of a calcific deposit with the release of crystals into the soft tissue. While there is prior literature on treatment with bisphosphonates or surgery, a fast and complete response to modest dose steroids suggests that this would be the first treatment to try in such a case.

INTRODUCTION

Calcific tendonitis often affects the tendons of the rotator cuff. This case explores an unusual calcified lesion on the dorsal aspect of the hand and its dramatic response to steroid treatment.

CASE REPORT

A 30-year-old civil servant was referred to the rheumatology department in March 2018 with a 1-year history of a swelling on the radial side of her right middle metacarpophalangeal joint (previously shown to be calcified, Fig. 1). She first noticed swelling and pain after long periods of typing. This initially settled down with rest and was asymptomatic for 5 months. However, over a 2-week period, she had developed rapid swelling throughout her right hand (Fig. 2) with all the characteristics of crystal arthropathy (acute calcium pyrophosphate deposition disease).

Blood tests revealed a C-reactive protein of 50 and an ESR of 34. Phosphate was low at 0.63 mmol/l, but calcium and parathyroid hormone were normal. Vitamin D was reduced at 44. Rheumatoid factor, anti-cyclic citrullinated peptide, anti-nuclear antibody and extractable nuclear antigen were all negative.

She was treated with 160 mg intramuscular methylprednisolone injection and 10 mg prednisolone daily for 2 weeks, followed by 5 mg daily for a further 2 weeks. Although the pain subsided within 5 days of the intramuscular steroids, the stiffness and swelling persisted for a further 7 days. After a month of treatment, the patient was pain free with near normal dexterity. The series of X-rays below demonstrate the radiographic appearance before and 2 months after treatment.

Figure 1 taken on 06 June 2017 shows a calcific deposit 1.1 × 0.7 cm. Figure 3 taken on 14 March 2018 showed progression from a discrete mass to diffuse calcification to almost complete resolution of the calcific tendonitis (Fig. 4 taken on 15 May 2018). The patient’s swelling resolved and function returned to near normal (Fig. 5).
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Further imaging was performed in the form of ultrasound on the same day as the first X-ray of the patient’s right hand on 06 June 2017. This confirmed a calcific density within the subcutaneous tissue adjacent to the third metacarpal head. It has a smooth outline demonstrating benign sonographic features. The ultrasound also confirmed normal appearance to the second and third metacarpophalangeal joints and extensor tendons.

DISCUSSION

Idiopathic ectopic calcification is a rare clinical and radiological finding, especially without biochemical abnormalities such as hyperparathyroidism or hyperphosphataemia. While there is prior literature on treatment with bisphosphonates or surgery, systemic steroid therapy remains the most effective treatment [1].

Chondrocalcinosis is common in the elderly population with 25% prevalence in those above 85 years of age. Calcium pyrophosphate may be deposited in synovium and cartilage but much less often in periarticular soft tissues [2]. This case is unusual due to the patient’s young age at presentation, the atypical location of the disease within the periarticular tissues and brisk response to steroids.

This case can be compared and contrasted to the condition of hyperphosphataemic familial tumoral calcinosis, which is an autosomal recessive condition involving metastatic mineralization [3]. The pathogenesis is due to mutations in genes that regulate phosphate metabolism. However, this patient had a slightly low level of phosphate rather than hyperphosphataemia. Other forms of ectopic calcification are secondary to vitamin D intoxication, hyperparathyroidism, scleroderma, uremia from chronic kidney disease, ochronosis, milk alkali syndrome and bony destruction due to malignancy [6]. The management of secondary calcification is best managed by effectively treating the underlying cause.

Idiopathic calcific tendonitis within the hand is caused by calcium hydroxyapatite deposition in and around tendons. Acute pain, oedema and erythema, such as that reported in this case, are thought be secondary to the rupture of the calcific deposit into the surrounding soft tissues [4]. The most common site of deposition is in the supraspinatus tendon. It is theorized that local hypoxia and subsequent metaplasia lead to
fibrocartilage formation before dystrophic calcification occurs. Histological examination reveals collections of hydroxyapatite crystals surrounded by inflammatory cells [5].

Biopsy of the lesion was considered in this case, but deemed not to be the best course of action, given that the patient’s history was more suggestive of an acute inflammatory process rather than malignancy. This was also supported by the benign sonographic features found on ultrasound. In this case, the risks of complication from an invasive biopsy procedure were felt to outweigh the benefits and was therefore not requested.

In some cases, successful treatment has followed surgical excision [6]. However, steroids offer a much less invasive, cheaper and less risky alternative to surgery.

Ectopic calcification and inflammation that is steroid-responsive can occur in joints, tendons and periarticular tissues. This case reports a chronic calcified mass, which was associated with an acute inflammatory episode in the hand. This is likely to represent rupture of a calcific deposit with the release of crystals of either calcium pyrophosphate or hydroxyapatite into the soft tissue. A fast and complete response to modest dose steroids suggests that this would be the first treatment to try in such a case.

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CONFLICT OF INTEREST STATEMENT
None declared.

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ETHICAL APPROVAL
Not applicable.

CONSENT
Written consent obtained from the patient. Please see attached file for reference.

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