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# ANCA-associated vasculitis can present with episodic attacks of joint pain consistent with palindromic rheumatism

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## SUMMARY

A 64-year-old man with a 2-year history of palindromic rheumatoid arthritis, presented with recurrent flares of arthritis, weight loss, new onset Raynaud's phenomenon and one previous episode of small-volume haemoptysis. Investigations, including renal biopsy, revealed antineutrophil cytoplasmic antibodies-mediated vasculitis. This case highlights the need to consider vasculitis in patients in whom there is an atypical history of arthritis.

## BACKGROUND

Antineutrophil cytoplasmic antibody (ANCA) associated vasculitis can be difficult to diagnose. Clinical presentation is variable, but usually there is systemic involvement including the upper respiratory tract, lower airway and kidneys.<sup>1</sup> Arthralgia and arthritis occur in up to 50% of patients,<sup>1</sup> most frequently in the large joints. However, arthritis is very rarely the initial presenting problem.<sup>2</sup> Rheumatologists assessing patients in early inflammatory arthritis clinics may not be alert to the possibility of AASV. We describe a patient presenting with intermittent episodes of arthralgia diagnosed as palindromic rheumatism (PR) who subsequently evolved into AASV with crescentic glomerulonephritis. This case highlights the need to consider vasculitis in patients in whom there is an atypical history of arthritis.

## CASE PRESENTATION

A 64-year-old man presented with recurrent flares of intense joint pains and swelling in his hands and feet, each episode lasting a day, with no joint symptoms between attacks. His symptoms had started 2 years previously when he had seen a rheumatologist and a diagnosis of PR was made. The joint pains were very steroid-responsive, but both hydroxychloroquine and sulphasalazine trials for at least 3 months had not reduced the frequency or severity of his attacks. On detailed questioning, over the preceding 1 year he had lost 10 kg in weight, developed Raynaud's phenomenon while skiing and he experienced one episode of small-volume haemoptysis. He reported transient tender lower limb nodules, calf claudication symptoms and intermittent painful paraesthesia. He had no history of uveitis or inflammatory bowel disease and no other relevant medical or family history. He lived with his partner, was an ex-smoker and previous cocaine user.

On examination, his pulse was 90 beats/min, blood pressure 139/83 mm Hg, weight 85.9 kg. Urine dipstick showed protein ++, blood +++++. Clinically, he was thin, but looked well. There were no rashes or palpable nodules. Cardiovascular, respiratory and abdominal examination were normal with normal peripheral pulses. Neurologically, there was no evidence of a neuropathy or myopathy and his Achilles tendons were pain free. There was small joint tenderness in the fingers, but no synovitis. Nailfold video capillaroscopy was normal.

## INVESTIGATIONS

Investigations demonstrated a reduced estimated glomerular filtration rate of 48, normocytic anaemia with haemoglobin 113 g/L and mean corpuscular volume 86 fL. His eosinophils were  $0.2 \times 10^9/L$ , platelets  $266 \times 10^9/L$  and white cell count  $7.0 \times 10^9/L$ . Inflammatory markers were raised with erythrocyte sedimentation rate 47 mm/hour and C reactive protein (CRP) 48 mg/L. Serum urate was normal.

In view of his atypical history and above blood results, an ANCA was sent. This revealed a strongly positive proteinase 3 (PR3) antibodies 219. Antinuclear antibody, extractable nuclear antigens, dsDNA and myeloperoxidase antibodies were negative with normal complement levels. Rheumatoid factor and anticyclic citrullinated peptide antibody were negative. Two COVID-19 swabs, urine drug screen, hepatitis B and C, and HIV serology were negative.

A renal biopsy showed ANCA-mediated vasculitis (focal class) with 3 (27%) cellular/fibrocellular crescents, mild acute tubular injury with focal red cell casts and 5%–10% interstitial fibrosis and tubular atrophy.

Lung function tests were entirely normal and CT chest showed non-specific subtle areas of ground-glass changes in the lower lobes of unlikely significance.

## OUTCOME AND FOLLOW-UP

He was treated with a high dose of 60 mg oral prednisolone and intravenous rituximab 1 g 2 weeks apart with subsequent resolution of his joint symptoms, improvement in PR3 titres to 99 units/mL and normalisation of the CRP.

## DISCUSSION

PR is a clinical syndrome characterised by recurrent episodes of pain and swelling, usually of the small



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## Case report

joints of the hands. Flares of PR always resolve spontaneously and do not result in joint damage, distinguishing it from rheumatoid arthritis (RA), where joint disease is persistent, leading to radiographic bony erosions.<sup>3</sup> Although it is a distinct entity to RA, up to 50% of patients do go on to develop persistent RA.<sup>4</sup> However, little is known about the underlying pathophysiology, how to identify which patients will go on to develop RA and to what extent patients may develop other rheumatological conditions. To our knowledge, this is the first case of intermittent seronegative small joint arthritis, consistent with PR, which has resulted in a diagnosis of AASV. Although rare, there are reports of patients with rheumatoid factor positive persistent inflammatory arthritis who have developed AASV, but none of these patients presented initially with PR.<sup>5</sup>

### Learning points

- We describe a patient with antineutrophil cytoplasmic antibody-associated vasculitis presenting with episodic attacks of joint pain consistent with palindromic rheumatism and deteriorating renal function secondary to crescentic glomerulonephritis.
- Without prompt diagnosis, this patient may have developed irreversible renal damage.
- Therefore, the importance of considering vasculitis in atypical rheumatological presentations is highlighted.

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