Remitting seronegative symmetrical synovitis with pitting oedema associated with lung malignancy

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Abstract
RS3PE syndrome is an inflammatory disease which affects mainly males and responds rapidly to low-dose steroids. We describe the concurrence of RS3PE and lung malignancy in a 78-year-old woman. We recommend that an underlying malignancy should always be excluded in patients with RS3PE syndrome, even when general signs and symptoms such as weight loss, anorexia and fever are absent or uncertain.

Keywords: lung malignancy, RS3PE, paraneoplastic syndrome, elderly

Case report
A 78-year-old woman was seen at the outpatient clinic of the Department of Geriatric Medicine with a 4-week history of bilateral hand swelling and pain. For the previous 2 weeks, she had pain in the wrists, elbows, shoulders and ankles. She had a history of hypertension, peripheral arterial disease and she had been smoking 20 cigarettes a day from the age of 20 till the age of 65. She complained of the flu-like symptoms and recent weight gain.

On examination, she had pitting oedema of the dorsum of both hands, together with pain and limitation of movement at the wrists. Laboratory tests revealed raised inflammatory markers (ESR 45 mm/h, CRP 105 mg/l) with a normal white blood cell count and differential leucocyte count. Haemoglobin was 6.9 mmol/l and the rheumatoid factor was negative. Radiological examination of the hands and feet showed soft tissue swelling. The diagnosis of remitting seronegative symmetrical synovitis with pitting oedema (RS3PE syndrome) was suggested and, considering the possibility of an underlying occult malignancy, a chest radiography was performed. This showed a large tumour mass in the right lung (5 cm by 4 cm). Computed tomography scan of the chest confirmed the presence of a tumour in the right lung with massive mediastinal lymphadenopathy suggesting regional metastases. The bone scintigraphy showed no clear sign of bone metastases.

Discussion
In 1985, McCarty was the first to describe the RS3PE syndrome [1], observed in 10 patients who reported a symmetrical synovitis predominantly involving the wrists and flexor digitorum tendon sheaths associated with marked pitting oedema of the dorsum of both hands. This syndrome affects mostly the elderly (gender ratio M:F = 2:1). Diagnostic criteria for the RS3PE syndrome were proposed by Olive et al. [2] and include (i) bilateral pitting edema of both hands; (ii) sudden onset of polyarthritis; (iii) age >50 years; and (iv) seronegative rheumatoid factor. Polyarthritis of the metacarpophalangeal joints, proximal interphalangeal joints, wrists, shoulders, elbows, knees, and ankles, often is observed in decreasing frequency [3]. Elevation of acute phase reactants, persistent rheumatoid factor seronegativity, and oedema responsive to low-dose corticosteroids have also been consistently observed [4]. The aetiology of oedema in RS3PE is unknown, but recent magnetic resonance imaging studies suggest that marked extensor tenosynovitis is responsible [2]. The differential diagnosis for RS3PE includes polymyalgia rheumatica, rheumatoid arthritis, seronegative arthropathies, systemic lupus erythematosus,
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mixed connective tissue disease, systemic sclerosis, overlap syndromes, amyloidosis, pseudogout, carpal and tarsal tunnel syndromes [1, 5–7].

A variety of musculoskeletal syndromes have been associated with malignancy [8, 9] and the RS3PE syndrome has been previously reported to be associated with underlying malignancy [3, 4, 10]. The presence of systemic signs and symptoms such as weight loss, fever, anorexia, and poor response to corticosteroids are clinical characteristics of an RS3PE paraneoplastic syndrome. Our patient had minimal symptoms to suggest a malignancy. However, being aware of the possibility that RS3PE may be paraneoplastic, and considering the past smoking habits of the patient, we performed additional diagnostic procedures which led to the diagnosis of lung malignancy. It is essential to suspect the presence of a malignancy in patients with the RS3PE syndrome when systemic signs and symptoms are present, or in the absence of response to corticosteroids. We now recommend that an underlying malignancy should always be excluded in patients with RS3PE syndrome even when general signs and symptoms as weight loss, anorexia and fever are absent or uncertain.

Key points
• RS3PE syndrome is an inflammatory disease which affects mainly elderly men and responses rapidly to low-dose steroids.
• The presence of a malignancy should be suspected in patients with the RS3PE syndrome when systemic signs and symptoms are present, or in absence of response to corticosteroids.
• We recommend that an underlying malignancy should always be excluded in patients with RS3PE syndrome.

References

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