EROSIVE SYNOVITIS, DACTYLITIS, HYPERCALCAEMIA, LYMHPHENADOPTHAPHY, ELEVATED SERUM ACE & HYPOTHYROIDISM – AN UNUSUAL PRESENTATION OF IGG4-RELATED DISEASE

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Background: IgG4-related disease (IgG4-RD) is a fibroinflammatory disorder which can affect almost any tissue in the body. We describe a challenging case of IgG4-RD which reflects the great heterogeneity of this disease.

Objectives: A 49-year-old Mauritanian gentleman was referred to Rheumatology with chronic progressive symmetrical synovitis affecting the upper and lower limbs. He had no features suggestive of a spondyloarthropathy or connective tissue disease. Blood tests revealed a raised C-reactive peptide (CRP) of 194 mg/L and erythrocyte sedimentation rate (ESR) of 46 mm/hr. His rheumatoid factor and anti-cyclic citrullinated peptide antibodies were negative. Anti-nuclear antibody was weakly positive at 1:80 with a speckled pattern, but extractable nuclear antigen antibodies, anti-double-strand DNA-antibodies and anti-neutrophil cytoplasm antibodies were negative. Complement proteins and creatine kinase were normal. Radiographs of the hands and feet demonstrated widespread erosive changes worst at the metacarpophalangeal, metatarsophalangeal and first tarsometatarsal joints, as well as wrist-neck deformities of the fifth phalanges in both hands. Chest radiograph and serological virology screen were both unremarkable. He was subsequently diagnosed with a seronegative inflammatory arthritis and treated with intramuscular methylprednisolone. He was awaiting subcutaneous methotrexate.

Methods: He was hospitalised shortly after with worsening polyarthritis and hypercalcaemia (3.34 mmol/L). Examination demonstrated synovitis, new lymphadenopathy and dactylitis. Other investigations revealed a normocytic anaemia (HB 80 g/dL, MCV 80 fL), neutrophilia (20 x 10^9/L), eosinophilia (4 x 10^9/L), thrombocytopenia (500 x 10^9/L), hypoalbuminaemia (25 g/L), CRP 200 mg/L and ESR 60 mm/hr. All immunoglobulins were raised with IgG 30.5 g/L. Parathyroid hormone was low (< 0.7 pmol/L) and vitamin D was 125 nmol/L. Thyroid function, countless antibodies and anti-neutrophil cytoplasm antibodies were negative. Complement proteins and creatine kinase were normal. Radiographs of the hands and feet demonstrated widespread erosive changes worst at the metacarpophalangeal, metatarsophalangeal and first tarsometatarsal joints, as well as wrist-neck deformities of the fifth phalanges in both hands. Chest radiograph and serological virology screen were both unremarkable. He was subsequently diagnosed with a seronegative inflammatory arthritis and treated with intramuscular methylprednisolone. He was awaiting subcutaneous methotrexate.

Results: A 60-year-old man with no remarkable past medical history presented to our department of rheumatology with right knee pain. He described a dull ache and swelling in his right knee with a dragging sensation, waking him up at night on a regular basis. The symptoms lasted from 6 months and were partially improved by analgesics and anti-inflammatory drugs. On examination, temperature was normal. The knee was edematous, erythematous, and warm with a range of motion of <90°. Laboratory investigations showed high acute phase reactants, the erythrocyte sedimentation rate was 75 mm. A plain radiograph of the right knee showed diffuse enlargement of the joint space due to a tissue infiltration within the synovium. These structures demonstrated heterogeneously increased T2 signal and intermediate T1 intensity characteristics. Imaging of the right knee showed diffuse enlargement of the joint space due to a tissue infiltration within the synovium. These structures demonstrated heterogeneously increased T2 signal and intermediate T1 intensity characteristics. Most of the joint space was replaced by hyper enhancing synovium. At that time, differential considerations included severe inflammatory arthritis and synovial chondromatosis rather than unusual metastasis.

Conclusion: This is a unique presentation for a rare disease. There are few case reports of IgG4-RD causing erosive synovitis with rheumatoid-like swan-neck deformities or hypercalcaemia. Furthermore, this is the first case report as far as we know, of the disease associated with dactylitis or a raised serum ACE. Prior to biopsy, our main working diagnoses were sarcoidosis or a lymphoproliferative malignancy. The peculiar presentation necessitated a second opinion via the national IgG4-RD MDT who confirmed the histological diagnosis of IgG4-RD. We hope that this interesting case highlights the importance of histology in diagnosis of complex cases. In addition, the case presentation adds to the multitude of clinical manifestations of IgG4-RD.

Disclosure of Interests: None declared

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