Objectives: The objective of our work was to evaluate its prevalence and its different aspects.

Methods: A retrospective study including 11 files of GPA patients followed over a period of 10 years in The Internal Medicine Department at Tahar Sfar Hospital Mahdia TUNISIA.

Results: Neurological manifestations were reported in 5 patients: a pyramidal syndrome was noted in 3 patients with a normal cerebral MRI, a disabled headache was described in one patient with a normal cerebral MRI. Peripheral neuropathy in 2 cases, and the Electroneuromyogram objectified a mononeuropathy affecting the ulnar nerve, peroneal nerve, and musculocutaneous nerve in one patient and a distal sensitivomotor poly neuropathy in one patient. Cranial nerves damage was noted in two cases, one VII (1 case) and one V (1 case).

Conclusion: Our study results, show the importance of timely diagnosis of these neurological manifestations, as their evolution will depend on the early GPA treatment.

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AB0769

EROSIVE SYNOVITIS, DACTYLITIS, HYPERCALCAEMIA, LYMPHADENOPATHY, 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 swan-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 ($20 \times 10^9/L$), eosinophilia ($4 \times 10^9/L$), thrombocythaemia ($500 \times 10^9/L$), hypoalbuminaemia (25 g/L), CRP 200 mg/L and ESR 60 mm/hr. All immunoglobulins were raised with IgG 30.5 g/L. IgG4 subset was 3.0 g/L. Parathyroid hormone was low (< 0.7 pmol/L) and vitamin D was 125 nmol/L. Thyroid function, myeloma screen and blood tumour markers were unremarkable. Blood film revealed no blasts. Serum angiotensin-converting enzyme (ACE) was elevated at 171.9 U/L. A CT-chest-abdomen-pelvis revealed an enlarged thyroid with retrosternal extension and widespread lymphadenopathy above the diaphragm.

Results: Biopsies of the thyroid gland and submandibular lymph node demonstrated lymphoplasmacytic fibroinflammatory changes and $> 40\%$ of IgG4 cells. Neither biopsy showed evidence of sarcoidosis, tuberculosis or malignancy. He was treated with a weaning regimen of high-dose oral corticosteroids resulting in normalisation of haematological, inflammatory and biochemical parameters. His musculoskeletal pain, swelling and early morning stiffness significantly improved too. Subcutaneous methotrexate was commenced and titrated upwards to 20 mg. Two months later, his arthritis relapsed upon reduction of corticosteroids. Again, there was a marked inflammatory response with raised CRP and ESR. He had also developed a new hypothyroidism (TSH > 100 mU/L, T4 0.5 µg/dL, positive TPO-antibody). Repeat hand and feet radiographs

revealed severe erosive progressive disease (Figure 1). He awaits commencement of rituximab.

Figure 1. Plain hand radiograph demonstrating extensive erosive arthritis.

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.

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AB0770

A RARE CASE OF SYNOVIAL METASTASIS REVEALED BY KNEE PAIN

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Background: Intra-articular masses are not frequently encountered in clinical practice. However, the differential diagnosis can be broad. Synovial metastasis is a rare presentation that carries a poor prognosis with a poor survival rate.

Objectives: Here by a case of synovial metastasis of the knee joint in a patient diagnosed with an adenocarcinoma of the lung.

Methods: 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 for 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 $< 90^\circ$. Laboratory investigations showed high acute phase reactants, the erythrocyte sedimentation rate was 75 mm. A plain radiograph of the left knee demonstrated a lytic lesion of the upper tibia. A magnetic resonance 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.

Results: Biopsy concluded to synovial metastasis from primary adenocarcinoma. Further investigations were necessary. Chest tomography showed a tumor process of left lower lobe associated with mediastinal lymphadenopathy. The patient was treated with palliative external radiotherapy to the right knee. The evolution was marked by the appearance of multiple tracheobronchial fistulas. The patient died 3 months later due to the progression of the disease.