Background: Systemic AA amyloidosis is a serious and life-threatening complication of chronic inflammatory diseases such as rheumatoid arthritis, spondyloarthritis (SpA), and periodic fever syndromes. While most common cause of AA amyloidosis is Familial Mediterranean Fever; Ankylosing Spondylitis (AS) is another frequent cause of AA amyloidosis in Turkey.

Objectives: We aimed to evaluate the response of secukinumab (SEC) treatment in three patients with AS and AA amyloidosis (AS-AA) in our tertiary referral centre.

Methods: We retrospectively evaluated three AA amyloidosis patients who fulfilled Modified New York Criteria for diagnosis of AS in our AA amyloidosis cohort with 163 patients. Diagnosis of AA amyloidosis was confirmed by Congo red stain and by monoclonal AA-specific antibodies.

Results: *Patient 1:* 61-year-old male patient with inflammatory back pain (IBP) and peripheral arthritis for 14 years was evaluated in our clinic. After methotrexate (MTX) failure, he used adalimumab (ADA), etanercept (ETA) and certolizumab (CZP). Nephrotic range proteinuria was detected when he was on CZP, and rectum biopsy documented AA amyloidosis 3 years ago. After the diagnosis, CZP treatment was switched to infliximab (IFX). IFX was ineffective in controlling inflammatory findings. SEC was started 15 months ago and he responded partially. The dose of SEC was increased to 300mg monthly, which resulted in a sustained improvement in clinical and laboratory findings.

Patient 2: 69-year-old woman was admitted to our clinic with peripheral arthritis in addition to the history of IBP for 19 years in 2005. MTX, NSAID and prednisolone were started. Because of inefficacy to conventional treatments and development of nephrotic range proteinuria, ETA was added to treatment. The patient responded to ETA and was followed-up for 13 years without symptoms of AS and proteinuria. ETA was switched to IFX due to secondary inefficacy two years ago. On the third month of IFX treatment, she developed demyelinating polyneuropathy. IFX treatment was switched to SEC and she is still being followed-up on SEC without any findings of AS and proteinuria.

Patient 3: 49-year-old woman who was on sulphasalazine for 24 years for treatment of ulcerative colitis (UC) was evaluated for recent onset IBP and peripheral arthritis in 2007. After failure of MTX, she started to receive IFX. She did not respond to first IFX and then ADA and CZP, and she developed nephrotic range proteinuria when she was on anti-TNF. Her serum creatinine increased progressively, and haemodialysis (H/D) was started six months later. Due to ongoing IBP and elevated acute phase response with CZP treatment, SEC was started. Significant improvement was observed in both clinical and laboratory findings with no worsening of UC.

Table 1. Clinical characteristics, laboratory findings and treatment responses of patients

	Patient 1	Patient 2	Patient 3
Age (year)	61	69	49
Sex	Male	Female	Female
Age of AS onset (year)	47	35	25
Age of amyloidosis diagnosis (year)	57	50	46
Amyloidosis duration (months)	42	168	30
Family history	No	AS and amyloidosis in siblings	No
HLA-B 27 status	Positive	Positive	Not available
MEFV status	Negative	Negative	M694V heterozygous
Organ involvement of amyloidosis	GIS and kidney	Kidney, liver, heart, bone marrow	GIS, kidney
Secukinumab duration (months)	15	18	24
CRP (mg/L)			
Before	27	15	23
After	2.4	4.5	2.8
Creatinine (mg/dl)			
Before	0.5	1.6	H/D
After	0.5	1.3	H/D
Proteinuria (g/day)			
Before	4.2	5.5	H/D
After	2.4	0.9	H/D
Albumin (g/dL)			
Before	2.7	2.5	3.1
After	3.1	4.3	4.2
ASDAS			
Before	4.1	1.6	4.6
After	1.8	1.1	1.7

GIS: Gastrointestinal System, ASDAS: Ankylosing Spondylitis Disease Activity Score

Conclusion: AA amyloidosis is a rare complication of SpA, and SEC treatment was found to be safe and effective in our three patients with AS-AA. Although anti-TNF agents have previously used successfully in treatment of AS-AA, SEC may be a new option especially in patients who are resistant or intolerant to anti-TNFs.

Disclosure of Interests: None declared
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AB0767 SEVERE BREAST ULCERATION IN BEHÇET'S DISEASE TREATED WITH CYCLOPHOSPHAMIDE: A CASE REPORT

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Background: Behcet's disease is a systemic autoimmune vasculitis that typically involves mucocutaneous surfaces, the eyes and the skin with varying manifestations. Skin ulceration often secondary to pathergy is a common feature.

Objectives: To raise awareness of breast ulceration as a clinical manifestation in Behcet's disease.

Methods: We report a case of complex Behcet's disease associated with severe refractory breast ulceration successfully treated with cyclophosphamide.

Results: A 27 year old female patient presented to the rheumatology department with a non-healing lesion on her face secondary to a cat scratch. She reported previous episodes of ulceration usually as a result of minor trauma such as IV cannulation, as well as a two year history of severe oral and genital ulceration. She had also been diagnosed with 'colitis' requiring ileocaecal resection two years prior to presentation. Previous treatments for her bowel included azathioprine, infliximab, adalimumab and vedolizumab. Over the next three years she developed recurrent eye involvement in the form of scleritis with ongoing oral and genital ulceration. Further treatment under our care included ustekinumab, etanercept, benepali, certolizumab and tacrolimus with background prednisolone. She then developed a small area of broken skin on her left breast which continued to extend and ulcerate despite treatment with IV steroids. Tissue biopsy showed deep focal vasculitis with intravascular thrombi. Anticoagulation was commenced and she was treated with tocilizumab IV for 3 months with no clinical improvement. Involvement extended to include 80 percent of the breast surface and the nipple self-amputated. Due to the severity and extent of the ulceration which was extremely painful and distressing, she was given 3 months of IV cyclophosphamide at a dose of 15mg/kg every 2-3 weeks. The ulceration showed rapid clinical improvement (see Figure 1).

Figure 1. Breast ulceration before and after 3 months of cyclophosphamide treatment

Conclusion: Involvement of the breast in systemic vasculitides (such as GPA or PAN) has been reported, although usually manifesting as a palpable mass with diagnosis on biopsy¹. Similar presentations in Behcet's disease have been recognised², although much less commonly. Our case demonstrates that severe breast ulceration can be a clinical manifestation of this condition and can be refractory to usual therapies.

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AB0768 NEUROLOGICAL MANIFESTATIONS IN GRANULOMATOSIS WITH POLYANGIITIS: ABOUT 11 CASES

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Background: Granulomatosis with polyangiitis (GPA) is a rare systemic vasculitis, affecting mainly the superior airways, lungs and kidneys. Neurological impairment is frequent, described in 22 to 54% of cases, and rarely inaugurates the disease.

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 x 10⁹/L), eosinophilia (4 x 10⁹/L), thrombocythaemia (500 x 10⁹/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°. 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.