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MONOARTHRITIS: PROBABLE OUTCOMES

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Background: Numerous joint disorders initially produce swelling in a single joint and new onset monoarthritis will probably further lead to the involvement of other joint groups and development of extraarticular manifestations. It is essential to take a proper diagnostic approach for organizing appropriate treatment and lowering possibility of disease progression.

Objectives: The aim of this study was to investigate joint distribution, determine rheumatological diseases of patients with acute monoarthritis and reveal the development of further systemic manifestations.

Methods: 100 patients (age 18-75 years) with clinically apparent monoarthritis of less than 6 weeks duration were included in the study. Criteria of exclusion were infection, trauma and crystal induced arthritis. Joint distribution, presence of systemic manifestations and development of chronic inflammatory rheumatic disease were evaluated. Presence of arthritis was proved with help of ultrasound examination. Complete blood count, ESR, CRP, RF, anti-CCP; HLAB27; MEFV mutations and X-ray of swollen joint were performed for all patients. Temperature was also measured.

Results: Mean age of patients with acute monoarthritis was 46±13 years. Female predominance was noted (61%). 71% of patients had elevated ESR, 69%- CRP. In 24% of cases homozygous or heterozygous mutations of MEFV gene were revealed. 21% of patients had positive RF and 18% - anti-CCP. 11% patients carried HLA-B27 antigen. 28% of examined patients had subfebrile fever. Hepatosplenomegaly was determined in 16%, uveitis in 5%, psoriatic plaque in 4%, interstitial pneumonia in 2% of cases

At the baseline 82 patients were diagnosed with rheumatological disease. Baseline data is shown in the Table 1 below.

Table 1. Baseline data

Diagnosis	Number of patients
FMF	23
Osteoarthritis (reactive synovitis)	16
Rheumatoid arthritis	15
Reactive arthritis	10
Ankylosing spondylitis	6
Psoriatic arthritis	4
SLE	3
Schönleyn-Henoch purpura	2
Sarcoidosis	2
Behcet diseases	1

Conclusion: In this study monoarthritis in majority of cases underlies FMF. Though FMF is not considered as a frequent cause of acute monoarthritis, more attention should be paid on this pathology in focus of monoarthritis, especially in specific for FMF region. Further follow up of acute monoarthritis progression is needed.

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“RICE AND SARDINES”: AN UNUSUAL CAUSE OF RASH AND JOINT SWELLING

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Background: Systemic vasculitis presents with numerous widespread manifestations. Various diseases can masquerade as systemic vasculitis (1). It is crucial to carefully evaluate patients with unusual presentations to look for vasculitis mimics. Scurvy, a disease caused by severe and prolonged vitamin C deficiency, still occurs in industrialized countries (2). Ascorbic acid is involved in various biologic processes including the synthesis of mature collagen. Lack of ascorbic acid especially affects blood vessel integrity leading to haemorrhagic manifestations characteristic of scurvy (3).

Objectives: Case report highlighting that scurvy is still prevalent in the west and may present with features resembling vasculitis.

Methods: Information was obtained from the patient's medical records.

Results: A 61 year old Caucasian female presented with a 3 weeks' history of bilateral lower limb pain, rash and swelling. She was referred to acute medicine to exclude deep vein thrombosis suspected by her general practitioner. She was previously fit and well apart from diagnosis of coeliac disease and various food intolerances. She believed her symptoms started following a recent course of Metronidazole for suspected dental infection which manifested with gum bleeding. She was found to have purpuric spots and ecchymosis bilaterally in her legs, from the toes to the groin. The left knee was swollen with evidence of hematoma around the joint. She denied fever, weight loss, night sweats, arthralgia, myalgia, rash, Raynaud's, alopecia, mouth or genital ulcers, ear-nose-throat manifestations. She denied chest pain, difficulty in breathing or symptoms of peripheral neuropathy. There was no history of asthma, inflammatory bowel disease or uveitis. She reported occasional alcohol intake, she did not smoke cigarettes nor use recreational drugs. Examination otherwise revealed no other major abnormalities with normal vital signs. Initial differential diagnosis included platelet or coagulation disorders or vasculitis. Initial blood tests revealed normal full blood count, coagulation screen, ESR, CRP, liver and renal biochemistry. Urine dip was positive for blood but not for protein. Knee aspiration was attempted but no fluid could be aspirated. Knee x-ray was unremarkable. ANA, ANCA and anti-GBM antibody tests were negative. On further review and enquiry into her symptoms and lifestyle she admitted having a diet exclusively of "rice and sardines" for nearly 2 years. This information along with clinical features led to the suspicion of Vitamin C deficiency that was subsequently confirmed. Ascorbic acid levels were <2.8 micromol/l (normal range 26.1-84.6 micromol/l). Replenishment of vitamin C was followed by a rapid and sustained recovery. Detailed investigation for additional macro and micronutrient deficiencies were undertaken with referral to a dietician and to gastroenterology.

Conclusion: This case illustrates scurvy can present in previously "well" individuals and should be considered as a possible differential diagnosis of vasculitic lesions in the appropriate clinical context. Detailed dietary history is invaluable.

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SPINAL HYDATID CYST DISEASE: WHAT FEATURES IN SURGICAL DEPARTMENTS?

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Background: Cystic echinococcosis (CE) is a zoonosis caused by the larve of *Echinococcus granulosus*. Cysts can develop in any part of the body, although the liver and lungs are most frequently involved. Bone echinococcosis is one of the rarest forms of the disease, accounting for 0.5 to 4.0% of all echinococcosis. Spinal cysts are disabling causing destruction similar to malignant bone lesions, with high risk of neurological deficit.

Objectives: To increase awareness of this disease, the clinical data of eight patients with spinal CE were analyzed retrospectively.

Methods: Clinical data of eight patients with spinal CE were analyzed retrospectively, collected over ten years on the department of orthopedics in the Military hospital of Tunis.

Results: The mean age of the patients was 49 years. The median disease duration was five years. All patients presented with back pain and paresthesia without neurological deficit. Radicular pain was reported by two patients. The diagnosis of spinal CE was made after the diagnosis of visceral CE in two patients. Former X rays showed nonspecific abnormalities and patients were treated initially by symptomatic treatments based on paracetamol and non-steroidal antiinflammatory drugs without any improvement. All of the patients needed Magnetic resonance imaging (MRI) to explore chronic back pain with paresthesia, revealing spinal CE. The typical MRI appearance is a multilocular cyst. Six patients had cervical and thoracic spinal cysts, one patient had a lumbar spinal cyst and one patient had cervical, thoracic and lumbar cysts. Further examinations with Computed tomography scans (CTscans) were needed before surgery for better examination bone destruction. All patients underwent surgery.

Cysts were removed with spinal fixation. All the patients showed relapses and needed at least three surgical interventions.

Conclusion: Bone echinococcosis is rare and often misdiagnosed. Radiographic and CT images lack disease-specific characteristics whereas MRI images offers a greater chance of direct diagnosis. Treatment of spinal hydatid disease is entirely surgical with high risk of relapses.

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TREATING IDIOPATHIC RECURRENT PERICARDITIS WITH INTERLEUKIN-1 INHIBITORS - A SINGLE CENTER EXPERIENCE

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Background: Pericarditis is a common disease with significant morbidity (1). Idiopathic pericarditis, where an underlying cause cannot be identified, makes up for 80% of cases in the Western World (1). Up to 30% of these patients experience recurrence despite optimal treatment (2). Idiopathic recurrent pericarditis (IRP) is thought to represent an auto-inflammatory process rather than a reinfection (3). 2015 European Society of Cardiology (ESC) guidelines have outlined treatment of acute episodes and first recurrence with nonsteroidal anti-inflammatory drugs (NSAID), acetyl salicylic acid (ASA) and Colchicine as first line and Glucocorticoids (GC) as second line treatment (3). However GC treatment increases the risk of relapse, dependence and toxicity (2). Interleukin-1 (IL-1) inhibitors have been proposed as possible treatments in IRP (3, 4).

Objectives: The aim of this case study is to outline our first experiences treating IRP with the IL-1 inhibitor anakinra in our Rheumatologic clinic.

Methods: All patients referred to our department in 2018/2019 with pericarditis were physically seen in our outpatient clinic. All patients were screened for malignancy, infection or rheumatologic disease as possible cause by clinical measures. Following ESC guideline, patients who suffered either the third recurrence under optimal treatment or significant side effects or dependency from GC were considered for anakinra treatment. Daily injection of anakinra (100mg) were given continuously over at least three months with gradual tapering over at least three months afterwards. Physical emergency department contacts, days hospitalized, colchicine- and GC use, the year prior to Anakinra treatment was recorded retrospectively. During follow up the same data was prospectively recorded.

Results: Over the course of two years 20 patients were referred to our clinic. All fulfilled ESC diagnostic criteria for pericarditis at index episode. In none of the patients could a rheumatologic, infectious or malignant cause be identified. 16 patients could be treated according to 2015 ESC guidelines with first or second line agents. Four patients were aligned to anakinra-treatment. Prior to referral, duration of symptoms was 5 - 120 months (mean 61 months). Further relevant patient- characteristics are outlined in Table 1.

After initiation of anakinra patients were afterwards regularly followed up in scheduled visits every 3 months.

Table 1. Characteristics of the four patients aligned to anakinra prior to anakinra-initiation.

Patient	Number of recurrences	Number emergency hospital contacts related to IRP the year prior to anakinra	Days hospitalized related to IRP the year prior to anakinra	GC dose prior to anakinra	other
I	2	2	3	7.5 mg	steroidglaucoma
II	7	2	4	10 mg	
III	4	7	22	20 mg	
IV	7	3	12	20 mg	

Follow-up after start of anakinra was 6-15 months (mean 11.5 months). No patient was admitted to hospital or emergency department in that period. All four patients could taper and stop GC without recurrence. One patient experienced a mild relapse after discontinuing anakinra and was restarted on a low dose with complete remission. No patient had elevated CRP values at the end of follow-up and no patient experienced tamponade or clinical signs of constriction. No significant side effects were noted, no patient had to stop anakinra-treatment during follow up.

Conclusion: Implementation of anakinra treatment in cases of complicated IRP was both secure and successful in our rheumatologic outpatient department. In our small sample we could confirm findings from bigger trials regarding effect- and side effect rates of anakinra treating IRP.

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HUGHES-STOVIN SYNDROME: A PECULIAR AUTOIMMUNE ORIGIN OF PULMONARY ANEURYSMS

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Background: Hughes-Stovin Syndrome (HSS) is a rare potentially fatal vasculitis supposedly belonging to the spectrum of Behçet disease without ocular involvement. HSS tends to play by a temporal pattern, starting with thrombosis and followed by formation of pulmonary aneurysms. Since its mortality can reach 25% of cases, the early recognition and the appropriate therapy represent the major challenges.

Objectives: In this report, we aimed at describing a rare case of HSS successfully treated in accordance with a prompt combined management with endovascular approach and immunosuppressive treatments.

Methods: We described a Caucasian 33-year-old man presented to the Emergency Room of our Hospital with hemoptysis associated with dyspnea and chest pain, without fever and hypotension, with a 2-year history of painful genital and oral ulcers. Blood test revealed anemia, elevated white cell count with increased C-reactive protein (77 mg/L) and D-dimer (2740 ng/mL). Doppler ultrasonography of legs showed thrombosis of the left femoral vein, while CT-angiography reported multiple aneurysms of the pulmonary arteries. An early and extensive work-up for infections and autoimmunity did not document relevant abnormalities. An appropriate genetic assessment revealed the HLA-B51 positivity. The patient immediately underwent endovascular embolization with coils and plugs of the largest pulmonary aneurysm, and inferior vena cava filter placement. Then, he started methylprednisolone (1 mg/kg/day) IV for three days in association with LMWH (6000 IU/day). Prednisone (1 mg/kg/day) in combination with cyclophosphamide (100 mg daily) were started orally, with tapering of steroids within a month. At the tight follow up (1 month and 3 months later), no genital and oral ulcers nor vascular thrombosis occurred, and acute phase reactants were in normal range. The 3-month-CT angiography showed a complete resolution of the aneurysms and no new changes.

Results: The peculiar diagnosis of HLAB51 positive HSS with multiple pulmonary aneurysms was made. The appropriate imaging followed by a prompt endovascular embolization of aneurysms with LMWH treatment were successfully performed. Interestingly, the oral combination therapy with corticosteroid and cyclophosphamide showed a rapid efficacy with a relevant safety profile. At the tight follow up, the young man improved significantly with clinical signs and pulmonary changes (Figure 1 next page).

Conclusion: This case is of interest because of the early recognition of such a rare disease that allowed an adequate combined radiological, vascular, and rheumatologic approach. In our case, LMWH resulted useful in preventing the pulmonary embolism, despite the use of anticoagulants is still debated in HSS. Moreover, the endovascular embolization let a less invasive approach to surgery without the need for multiple surgical procedures. For the first time, we documented that oral cyclophosphamide showed an early efficacy as a first line therapy of a HLA-B51 HSS.

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