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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