A CASE OF SYSTEMIC SCLEROSIS COMPLICATED BY RENAL CRISIS: POTENTIAL ETIOPATHOGENETIC ROLE OF CYTOMEGALOVIRUS AND TREATMENT

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Background: Scleroderma renal crisis (SRC) is a rare complication of systemic sclerosis (SSc), which can be triggered by viruses, such as Cytomegalovirus (CMV). SRC presents as a new-onset accelerated-phase hypertension with/without rapidly progressive renal failure.

Objectives: Here we describe the case of a patient developing SSc complicated by the appearance of SRC after a recent episode of acute Cytomegalovirus infection.

Methods: A 66-year-old male was referred to our Scleroderma Unit in March 2019. He presented with widespread skin rash, exertional dyspnoea and peripheral oedemas. He reported a myocarditis due to CMV occurred in October 2018. Antibodies anti-CMV IgM were detected in his serum. The patient developed a progressive cutaneous involvement characterized by diffuse oedema, sclerosis and melanoderma. Subsequently, Raynaud’s phenomenon, puffy hands and pitting scars occurred. Laboratory tests showed positive ANA in a titer of 1:640 in a nucleolar staining pattern. Additionally, persistence of anti-CMV IgM was found. Skin biopsy showed scleroderma-like finding. Nailfold capillaroscopy revealed a SSc pattern. Chest high resolution computed tomography displayed basal interstitial thickening and subpleural ground-glass opacities. Therefore, the patient was diagnosed with SSc. Three weeks later he developed severe hypertension and a rapid, progressive renal impairment. Serum creatinine increased (up to 4.15 mg/dl), glomerular filtration rate impaired (25 ml/min).

Renal biopsy (picture A, B) revealed acute thombotic microangiopathy. A diagnosis of thrombotic thrombocytopenic purpura was excluded. The patient was diagnosed with SRC and we started therapy with ACE-inhibitor and loop diuretic. Even if the dosage of ACE-inhibitor was increased up to the maximum tolerate dose, his renal function did not improve and the blood pressure control was inadequate. Consequently, the patient underwent plasma exchange (PEX) sessions. Two weeks later there was an improvement of renal function and blood pressure normalized. Six months later the disease was controlled: glomerular filtration rate was 41 ml/min and blood pressure was within the normal range. The patient was treated with ACE-inhibitor and underwent fortnightly apheresis sessions. Treatment for scleroderma vasculopathy is ongoing.

Results: Viral infections may be responsible for SSc. A brief interval between an acute viral infection and the onset of SSc may suggest CMV as a possible trigger for the disease. Similarly, other infectious agents could be involved in the multistep and multifactorial mechanism of SSc. This case sheds light on the potential and intriguing role of CMV in SSc. Moreover, it is often insidious, underestimated and it has a poor prognosis. The disease usually shows a prodromal phase characterized by asthma and allergic manifestations.

Objectives: We report the case of a young patient with acute coronary syndrome (ACS) complicated by cardiac shock as the first manifestation of EGPA.

Methods: A 36 year old Indian male patient, with a previous history of asthma, rhinitis, Raynaud syndrome and allergy to ketoprofen, presented to the emergency department with a complaint of chest pain and dyspnea. Cardiac troponin was elevated and he was admitted to intensive care unit with a diagnosis of ACS. The patient conditions rapidly deteriorated due to acute cardiacogenic shock and an urgent coronary angiogram was performed. An occlusion of the left main coronary artery was treated with angioplasty and two drug-eluting stents. Echocardiography showed severe left ventricular dysfunction requiring inotropic and intra-aortic balloon pump support. A mild dermatitis after salicylic acid administration resolved with intravenous hydrocortisone 1 g. The thrombophilia

References:

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screening was negative, as well as cardiovascular risk factors. Over the next days, the clinical conditions rapidly improved with recovery of normal ventricular function on discharge. However, two weeks later he was readmitted with recurrent mandibular and chest pain. Troponin levels were elevated and fluctuated, suggesting recurrent ischemic events. Repeated ECG during angina crisis showed ischemic alterations in different coronary territories. The coronary angiogram detected coronary vasoospasm of the circumflex artery, reversible after nitroglycerin (Figure 1). Nitroglycerin and calcium channel blockers were initiated, but did not resolve the vasospastic angina crisis, occurring daily. Laboratory tests revealed eosinophilia (4390 cells/mcL), increased C reactive protein (9.4 mg/dL) and positive antinuclear antibodies (1:320). The other serological and immunological tests were negative, including MPO-ANCA and PR3-ANCA. An abdomen and chest CT scan was negative.

**Methods:**

**Objectives:**

Glucocorticoids, sometimes combined with other immunosuppressives are the standard treatment of IgG4-RD, in some situations (e.g. mechanical complications or suspected malignancy) surgery may be necessary but little is known about the management of fully resected single-organ IgG4-RD [1].

**Results:**

A woman (51 y) with pre-existing Hashimoto's thyroiditis (thyroid peroxidase antibody positive) developed a rapidly growing struma with very firm consistency (not allowing fine needle biopsy). Besides slightly increased C-reactive protein (5.3 mg/dL) there was no laboratory sign suggestive for IgG4-RD (normal serum IgG4, complement, eosinophils and IgE). Within 4 months the patient suffered from hoarseness and progredient dyspnea. Surgical thyroidectomy was performed and histopathology revealed IgG4-related Riedel's thyroiditis with extensive (storiform) fibrosis, a dense lymphoplasmacytic infiltrate, obliterator phlebitis, eosinophilia and 13 IgG4-positive plasma cells per high power field.

After referral to our department a comprehensive work-up showed no signs of other manifestations of IgG4-RD. Treatment with glucocorticoids is clearly recommended for patients with symptomatic IgG4-RD in an international consensus statement, whereas “watchful waiting” may be appropriate in some cases of asymptomatic or mild disease. While some highly fibrotic lesions may not respond well to glucocorticoids and may require surgical intervention, no clear guidance is available for the management of fully resected single organ IgG4-RD [2].

A brief review of the literature revealed that few cases of single-organ IgG4-RD remaining in remission after resection without medical treatment have been reported e.g. IgG4-related cholecytitis, autoimmune-pancreatitis, tumours of the intestinal tract, lung, thymus, meninges, paravertebral space and others [3–9]. After discussion of the options with the patient no systemic immunosuppression was given under close follow up without signs of relapse in clinical examinations, laboratory or imaging during the first 6 months.

**Conclusion:** Limited evidence from case reports suggests that a "watchful waiting" strategy without systemic immunosuppressive treatment may be reasonable in some cases of single-organ IgG4-RD after the affected organ was completely resected (e.g. due to mechanical complications or suspected malignoma). However, close follow-up monitoring should be applied due to the risk of relapse or development of new organ manifestations.

**References:**


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**THU0602**

**EXTENDED BONE HYDATIDOSIS IN THE HIP AND FEMUR WITH EXTENSION TO THE SOFT PARTS: A CASE REPORT**

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**Background:** Osseous hydatid cyst is an uncommon disease with weak response to treatment hydatid disease should be included in the differential diagnosis of cystic lesions of bone in endemic regions. Bone cysts account for only 0.5 to 2.5% of all hydatid cysts in humans.

**Objectives:** To report a case of osseous hydatid disease extended on hip and femur

**Methods:** We report a case of osseous hydatid disease

**Results:** A 49 YEAR OLD BRICKLAYER, with no past-medical history and no animal contact, was admitted to our department for a left hip pain, the patient was apyretic and in a good general health condition. He had a very painful walk, the mobility of the left hip joint was very painful and restricted. The pelvis X-rays showed osteolytic lesions in the ischiopubic branch and in the left femur and proximal extremity of the tibia. The C-Reactive protein value, the protein electrophoresis were normal. tumor markers test was negative.

An ultrasound of the hip showed a low abundance intra-articular effusion. The Pelvic MRI showed multilocal appearance extending over the bone and muscle with breach of the bone cortex of the femur very suggesting of the diagnosis of a bony and muscular echinococcosis.

**Conclusion:** Hydatid disease occurs worldwide and mainly associated with sheep farming. The liver and lungs are the most common locations. Bone cysts are uncommon but severe. Although immunofluorescent assays are useful, the final diagnosis depends on histology. The treatment is almost surgery. Recurrence is common.

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**THU0603**

**IGG4-RELATED SINGLE-ORGAN DISEASE: IMMUNOSUPPRESSION NEEDED AFTER COMPLETE RESECTION? A CASE REPORT AND A BRIEF LITERATURE REVIEW.**

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**Background:** IgG4-related disease (IgG4-RD) is a polymorphic autoimmune disease leading to tumourous swelling and/or fibrosis of affected organs. Riedel's thyroiditis is – besides chronic periarticular, Mikulicz Syndrome and many others – an organ manifestation of IgG4-RD that has been thought to be an independent disease for a long time. About 40% of patients have single organ IgG4-RD while the others suffer from multisystemic disease [1].

**Objectives:** Glucocorticoids, sometimes combined with other immunosuppressives are the standard treatment of IgG4-RD, in some situations (e.g. mechanical complications or suspected malignancy) surgery may be necessary but little is known about the management of fully resected single-organ IgG4-RD [1].

**Methods:** We report a case of single-organ IgG4-RD (Riedel's thyroiditis) after complete resection and perform a brief review of the literature to guide clinical management in this situation.

**Results:** A woman (51 y) with pre-existing Hashimoto's thyroiditis (thyroid peroxidase antibody positive) developed a rapidly growing struma with very firm consistency (not allowing fine needle biopsy). Besides slightly increased C-reactive protein (5.3 mg/dL) there was no laboratory sign suggestive for IgG4-RD (normal serum IgG4, complement, eosinophils and IgE). Within 4 months the patient suffered from hoarseness and prophredent pain. Surgical thyroidectomy was performed and histopathology revealed IgG4-related Riedel's thyroiditis with extensive (storiform) fibrosis, a dense lymphoplasmacytic infiltrate, obliterator phlebitis, eosinophilia and 13 IgG4-positive plasma cells per high power field.

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A brief review of the literature revealed that few cases of single-organ IgG4-RD remaining in remission after resection without medical treatment have been reported e.g. IgG4-related cholecystitis, autoimmune-pancreatitis, tumours of the intestinal tract, lung, thymus, meninges, paravertebral space and others [3–9]. After discussion of the options with the patient no systemic immunosuppression was given under close follow up without signs of relapse in clinical examinations, laboratory or imaging during the first 6 months.

**Conclusion:** Limited evidence from case reports suggests that a "watchful waiting" strategy without systemic immunosuppressive treatment may be reasonable in some cases of single-organ IgG4-RD after the affected organ was completely resected (e.g. due to mechanical complications or suspected malignoma). However, close follow-up monitoring should be applied due to the risk of relapse or development of new organ manifestations.

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