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 vasospasm 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/l) 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.

Figure 1. Coronary angiogram showing coronary vasospasm in the left circumflex artery (panel A, yellow arrow), reversible after nitroglycerin infusion (panel B, yellow arrow)

Results: The eosinophilia and the history of asthma rose the suspect of EGPA vasculitis. The patient was treated with intravenous methylprednisolone 250mg once daily for 3 days, followed by oral prednisone 1 mg/kg/day, with rapid and complete resolution of the recurrent angina episodes. Intravenous cyclophosphamide 10mg/kg was administered every 2 weeks for 2 times, then 12mg/kg every 4 weeks. Oral corticosteroid was tapered, with the persistence of a complete remission of the symptoms, after 2 months of immunosuppressive therapy.

Conclusion: Coronary involvement in EGPA can mimic atherosclerotic artery disease and can be life threatening, if not promptly recognized. An accurate medical history and complete serological and immunological tests are crucial to detect an atypical onset of EGPA, prompting early immunosuppressive therapy which is pivotal for the patient survival.

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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 afebrile 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 pelvic 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 multilocular 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.

NO other localization of hydatidosis were detected, body ct scan was normal

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 BRIEF LITERATURE REVIEW.

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Background: IgG4-related disease (IgG4-RD) is a polymorphic autoimmune disease leading to tumorous swelling and/or fibrosis of affected organs. Riedel's thyroiditis is – besides chronic periaortitis, 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 (5,3mg/l) 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, obliterative 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 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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