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Case Report

Idiopathic Intracranial Hypertension Associated with Adult Still's Disease

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Introduction

Adult Still's disease is a rare auto inflammatory condition characterised by fever, arthralgias and skin rash. A wide spectrum of complications have been described, such as cardiac tamponade, pulmonary hypertension, macrophage activation syndrome, thrombotic microangiopathy, diffuse alveolar haemorrhage and even acute respiratory distress syndrome. We report an 18-year old female who met enough criteria for diagnosis of Adult Still's disease, but also presented with signs and symptoms of idiopathic intracranial hypertension.

Case Report

An 18 year-old female was admitted to our hospital because of a 4-month history of fever, arthralgias and headache. She described daily fever attacks, not accompanied by chills, of up to 40°C (104°F), night sweats, a 20 kg-weight loss in that period of time and an intermittent erythematous rash which extended to the trunk and extremities. During the first few days of the presentation, she had experienced a sore throat which resolved spontaneously. Symmetrical arthralgia of the elbows, wrists, hands, knees and ankles developed over the following weeks. Her headache was intermittent, pulsatile and located in the frontal area. She did not have any past-medical records, her last period had been two months before her consultation, and did not consume any medications.

On admission, her blood pressure was 100/60 mm Hg, pulse rate 113 bpm, and her temperature was 36.3°C. Her body-mass index was 27. She had swollen elbows, wrists, hands, knees and ankles, and a 2 cm, elastic and painless lymph node was palpated in her left axilla. Other findings were hepatomegaly, and a non-palpable spleen. Funduscopic exam revealed bilateral blurring of optic discs, with no visible retinal venous pulsations. Neurologic and the remainder of the examination were unremarkable. Blood and urine cultures were negative. Laboratory tests revealed a microcytic, hypochromic anaemia, leucocytosis (12,700/μL), high levels of globular sedimentation rate and C-reactive protein (112 mm/h and 4.38 mg/dL, respectively), and lactic dehydrogenase was more than threefold above the normal upper range (1,160 IU/L). The following serologies resulted negative: antinuclear antibodies, anti-dsDNA, rheumatoid factor, anti-Sm, anti-ribonuclear proteins and anti-Scl-70. Complementemia was normal. Fourth-generation HIV test, hepatitis B and C were negative. The rest of the laboratory was normal. A CT scan revealed enlarged lymph nodes in left supraclavicular fossa and axilla, retroperitoneum, celiac and inguinal groups. There were no signs of vegetations on echocardiographic exam. Chest X-ray and electrocardiogram were unremarkable. A normal campimetry was obtained. The axillar lymph node was biopsied, and its histology and flow-cytometry were consistent with reactive polyclonal hyperplasia.

Brain computed tomography and magnetic resonance with angiography were unrevealing and a lumbar puncture was performed. Opening pressure was 28 cm H_2O. Spinal fluid analysis was normal.

Since her admission to our hospital she had daily fever. Treatment with naproxen was commenced, and fever disappeared thereafter. She reported a sudden but transient improvement of her headache after the lumbar puncture was performed, and was successfully discharged with this medication and prednisone 60 mg per day (1 mg per kg of body weight). Fundus exam performed a month later, when she returned to a scheduled visit, showed a normal retina. She reported no further fever, arthralgias nor headache.

Discussion

Adult Still's disease (ASD) is an auto inflammatory disorder characterised by daily fever, arthralgias and an evanescent rash [1]. Even though genetic factors and infectious triggers have been suggested, its aetiology remains unknown [2]. Its diagnosis is one of exclusion, and several criteria must be fulfilled. The highest sensibility for diagnosis of ASD is achieved by Yamaguchi criteria, which are presented in Table 1 [3]. Our patient met three of the mayor (although referred by her,

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Table 1: Yamaguchi criteria for diagnosis of adult still disease.

Criteria	Present
Major criteria	
Fever of at least 39°C (102.2°F) lasting at least one week	Yes
Arthralgia or arthritis lasting two weeks or later	Yes
A non-pruritic macular o maculopapular skin rash, salmon-coloured and usually found over extremities or trunk during febrile episodes	No
Leucocytosis (10,000/μL or greater) with at least 80 % granulocytes	Yes
Minor criteria	
Sore throat	Yes
Lymphadenopathy	Yes
Hepatomegaly or splenomegaly	No
Abnormal liver function (aspartate and alanine aminotransferase and lactate dehydrogenase)	Yes
Negative tests for antinuclear antibody and rheumatoid factor	Yes

we were not able to witness the rash), and four of the minor criteria, which suffices to diagnose ASD. Other causes for her symptoms were excluded by biopsy of the axillar lymph node and negative serologies.

Idiopathic Intracranial Hypertension (IIH), on the other hand, presents with signs of increased intracranial pressure, such as headache, papilledema, and even visual loss, with normal cerebrospinal fluid composition, in the absence of ventriculomegaly or mass lesions [4]. Headache is the most frequent symptom, and papilledema, typically symmetrical, is the hallmark sign of IIH [5]. It most frequently affects obese women of childbearing age [6], and the annual incidence is 20 per 100,000 persons [7]. Its diagnosis is, also, achieved by mandatory criteria [4], all of them met by our patient: 1) symptoms and signs of increased intracranial pressure (headache, nausea, vomiting, transient visual obscuration, papilledema), 2) no localising focal neurological signs except unilateral or bilateral sixth nerve paresis 3)cerebrospinal fluid pressure ≥ 25 cm of water but without cytological or chemical abnormalities, 4) normal neuroimaging adequate to exclude cerebral venous thrombosis, and 5) no other cause of intracranial hypertension apparent.

The pathogenesis of the IIH is unclear, but increased water content, excess of cerebrospinal fluid production and its reduced absorption have been proposed as causes [6]. Growth hormone, tetracycline's, hypervitaminosis A, Addison's disease, systemic erythematous lupus, hypoparathyroidism, Behcet's syndrome, uraemia, coagulation disorders and polycystic ovarian syndrome have been associated with IIH [8-15].

After diagnosis, naproxen was commenced and all symptoms were partially controlled. She was discharged with naproxen and prednisone 60 mg per day. One month later, she attended a scheduled consultation, and referred that headache, fever and arthralgias had disappeared. We repeated the optic fundus and found a complete remission of papilledema.

It is difficult to determine if the resolution of the headache and papilledema was achieved because the activity of ASD was controlled. It is certainly improbable that the use of corticosteroids constituted the whole cause of this resolution because, even though steroids have been recommended for IIH in the past, it is now known that their use may provoke worsening of IIH and, nowadays, they have no role in the treatment other than as an adjunctive therapy for aggressive cases with marked visual loss while surgery is arranged [7].

To our knowledge, there have been no other reports of ASD associated with IIH. Our attention was drawn to the fact that the beginning of the headache, fever and arthralgias, were temporarily related. On the other hand, headache and papilledema resolved with treatment for ASD. We did not find any other cause for IIH. Even though we certainly cannot imply causality, these facts suggest a direct or indirect association. It should be noted that neither of the diseases have been completely understood so far, and there might be an unknown physiopathological mechanism between them.

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