Niasse M, Ndao AC, Bouchrane R, Charifah S, Bah A, Diallo S. Ankylosing Spondylitis associated with Adult Still's Disease: The First Senegalese Case Report. J Rare Dis Res Treat. (2021) 6(2): 1-4

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Journal of Rare Diseases Research & Treatment

Case Report Open Access

Ankylosing Spondylitis associated with Adult Still's Disease: The First Senegalese Case Report

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Article Info

Article Notes
Received: October 12, 2021
Accepted: Decemeber 10, 2021

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Introduction

Ankylosing spondylitis is the most common type of spondyloarthritis[1], a group of diseases characterized by axial skeleton and sacroiliac involvement, peripheral joints and/or entheseal involvement and extra-articular manifestations[1,2]. Other types of spondyloarthritis are psoriatic arthritis, reactive arthritis, arthritis associated with cryptogenetic inflammatory bowel disease, S.A.P.H.O. (Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis), juvenile spondyloarthritis and undifferentiated forms. All other forms of spondyloarthritis can develop into ankylosing spondylitis[1,2]. This form, which can also occur at the outset, is characterized by the presence of sacroiliitis, often associated with stiffness and spinal deformities[1,2,3].

General signs such as fever are rarely found in ankylosing spondylitis[4]. Their existence is often linked to the association of other diseases, notably infectious diseases or other types of auto-inflammatory diseases, including adult Still's disease. It is characterized by a triad of constant hectic fever, skin rash and arthritis and other miscellaneous manifestations[6]. The association between ankylosing spondylitis and adult Still's disease is rarely reported in the literature[5]. We report the first case in Senegal.

Case Report

A 43-year-old man followed up since April 2019 for ankylosing spondylitis diagnosed in accordance with the ASAS and modified New York criteria[7,8]. He presented a radiographic axial form with bilateral stage 3 sacroiliitis (Figure 1), associated with enthesitic peripheral involvement (bilateral plantar talalgia). HLA-B27 antigen was positive. Ankylosing spondylitis was active with a BASDAI of 45/100 and an ASDAS-CRP of 3.26. The patient was treated with non-steroidal anti-inflammatory drugs and methotrexate (15 mg per week); which is indicated in front of the peripheral attacks of spondyloarthritis. Evolution was marked by remission after three months of treatment.

In January 2020, he suddenly presented with a hectic fever oscillating between 38 and 39°5 c associated with peripheral, bilateral asymmetric polyarthritis involving the left knee, the right wrist and both ankles. The patient also complained of odynophagia, prompting ENT examination which revealed a non-exudative pharyngitis. The examination did not reveal any skin rash. Biological workup showed an inflammatory syndrome with an erythrocyte sedimentation rate of 82 mm at the first hour and a C-reactive protein of 212 mg/l.

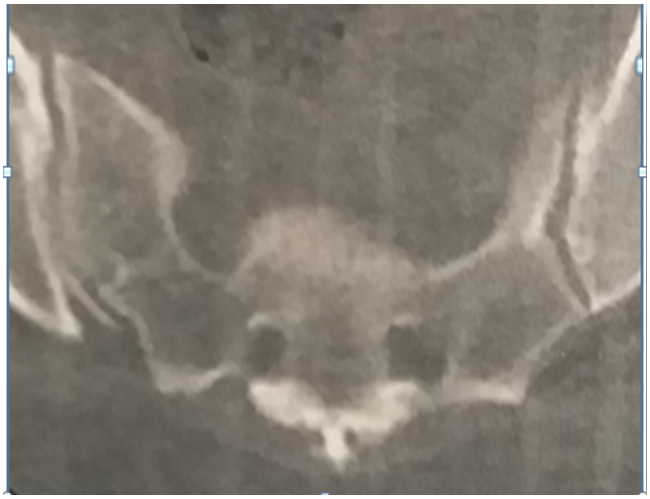

Figure 1: CT image showing bilateral sacroiliitis in our patient. We note diffuse sclerosis and irregular erosions of the sacroiliac joints giving « a postage stamp » appearance.

Complete blood count showed hyperleukocytosis (14,000 leukocytes/mm3) with neutrophil predominance (85%), normochromic normocytic anaemia with a haemoglobin level of 9 g/dl and a normal platelet count (420,000/mm3). Aspartate amino transferase values were elevated at 51 IU/l and alanine amino transferase was normal at 37 IU/l. There was hyperferritinemia at 10 times normal levels (1587 ng/ml).

The glycosylated fraction was reduced (10%). Infectious tests were negative. These included thick blood drop, blood cultures, throat swabs, urine cultures, viral serology tests (hepatitis B, hepatitis C and HIV) and syphilis. Immunological tests including rheumatoid factor, anti-cyclic citrullinated peptide antibodies and antinuclear antibodies were also negative. The diagnosis of adult-onset Still's disease (AOSD) was thus made in accordance with the criteria of Fautrel[9], and with those of Yamaguchi[10]. Non-steroidal anti-inflammatory drug was replaced by prednisone 1 mg/kg/day. Methotrexate, which was still being used before the onset of Still's disease, was continued. The course of treatment was favourable, with gradual reduction of the corticosteroids after 4 weeks.

Commentary

Ankylosing spondylitis is a chronic inflammatory rheumatic disease in which fever is rarely present[4]. Thus, the presence of fever suggests its association with other febrile conditions[5]. In our case, ankylosing spondylitis was associated with adult-onset Still's disease, which caused fever at nine months of follow-up.

The association of these two conditions suggests a coincidence or a pathophysiological link[5]. Indeed, both diseases are classified in the group of polygenic auto-inflammatory pathologies[11,12] which are mediated by innate immunity with a high production of pro-inflammatory cytokines (IL-1 β, IL-18, IL-6 and TNF-α). IL-1β, and IL-18 are activated by caspase-1, which is the main effector protease of the inflammasome[13,14].

Nonetheless, despite the pathophysiological links, the association between ankylosing spondylitis and adult Still's disease is rarely reported[5,15]. However, this rarity may reflect an underestimation, especially as Cush found sacroiliitis in 9.1% of adult Still's disease in a large review of articles[16]. To the best of our knowledge, between 2000 and 2020, eight (8) cases of this association in four (4) studies were published (see table, below).

These cases, as ours, all concerned men. Spondyloarthritis, as in our case, had preceded adult Still's disease. The types of spondyloarthritis were: ankylosing spondylitis (5 cases), psoriatic arthritis (2 cases) and undifferentiated spondyloarthritis (1 case). Psoriatic arthritis was found in a 21-year-old man and in his father. Our case is about a 43 years old who presented with ankylosing spondylitis. The average age of the 8 patients was 40 years (extremes: 21 and 62 years), similar to that of our patient (43 years). In adult Still's disease, fever and arthralgia were constant in all patients (Table 1).

Skin rash was present in 6 cases. On the biological level, the C-reactive protein was highly elevated except in 1 case, in which it was normal[17]. Hyperleukocytosis was noted in 6 cases. Ferritin levels in 4 patients, including our patient, were between 4 and 31 times normal. The glycosylated fraction was measured in our patient and in Aradoini[5]. It was collapsed (<20%) in both cases. The values were 10% and 15% respectively. The immunological tests, as well as infection workups, were negative in all cases.

Treatment of our patient is in accordance with the literature. Indeed, in spondyloarthritis, non-steroidal anti-inflammatory drugs are used as first line treatment and in adult Still's disease, high dose corticosteroids are indicated as first line treatment[12]. These anti-inflammatory treatments are often combined with conventional disease-modifying antirheumatic drugs (DMARDs)[18].

The use of DMARDs has anti-inflammatory drug sparing effect. Thus, in our patient, the maintenance of methotrexate allowed an early reduction of the corticosteroid dose after 4 weeks of treatment.

Conclusion

Our observation indicates the existence of the association between ankylosing spondylitis and adult Still's disease in our context. Thus, the association is certainly rare but possible. This couple must be evoked each time a known patient with spondyloarthritis presents a fever with a significant biological inflammatory syndrome and hyperleukocytosis with neutrophil predominance, after infectious and neoplastic diseases have been ruled out.

Table 1 : Comparing the characteristics of 8 reported cases in literature of the association Spondyloarthritis and adult Still's disease to those of our patient

Authors	Maeda H, et al. [17]	Akkoc N, et al. [15]	Aradoini N, et al. [5]	Duran TI, et al. [18]	Our case
Country	China	Turkey	Morocco	Turkey	Senegal
Year	2000	2008	2017	2020	2021
Number of cases	2	4	1	1	1
Age (in years)	Case 1 : 21 Case 2 : 46	N.F	31	62	43
Types of spondyloarthritis	Psoriatic arthritis : 2	A.S: 3 Undifferentiated Spondyloarthritis : 1	A.S : 1	A.S : 1	A.S : 1
Presence of HLA-B27	N.F	N.F	N.F	N.F	Oui
Delay between spondyloarthritis and adult Still's disease onset	Case 1 : 4 years Case 2 : 4 years	N.F	10 years	15 years	9 months
Fever	40°C	High (N.Q)	40°C	Oui (N.Q)	38-39,5°C
Skin rash	Absent	Present	Present	Absent	Absent
Arthritis	Present	Present	Present	Present	Present
Odynophagia	Case 1: Present Case 2: N.F	3 of the 4 cases	N.F	Present	Present
Organomegaly	Case 1: Present Case 2: N.F	N.F	Present	Present	Absent
Leukocytes/mm3	Case 1: 13800 Case 2: 10300	N.F	15000	11650	14000
C-reactive protein (mg/l)	Case 1: 12,5 Case 2: 3,4	High (N.Q)	300	192	212
Ferritinemia (ng/ml)	Case 1: 4 N Case 2: N.F	N.F	31 N	9 N	10 N
Glycosylated fraction	N.F	N.F	15%	N.F	10%
Rheumatoid factor	Negative	Negative	Negative	Negative	Negative
Antinuclear antibodies	Negative	Negative	Negative	Negative	Negative
Infectious tests	Negative	Negative	Negative	Negative	Negative

N.F: not filled in ; N.Q: not quantified; A.S : Ankylosing spondylitis ; N : normal

Conflict of Interest

The authors have no conflicts of interest.

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