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

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Introduction

Ankylosing spondylitis is the most common type of spondyloarthritis1, a group of diseases characterized by axial skeleton and sacroiliac involvement, peripheral joints and/or enthesal involvement and extra-articular manifestations1,2. 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 spondylitis1,2. This form, which can also occur at the outset, is characterized by the presence of sacroiliitis, often associated with stiffness and spinal deformities1,2,3.

General signs such as fever are rarely found in ankylosing spondylitis4. 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 manifestations6. The association between ankylosing spondylitis and adult Still’s disease is rarely reported in the literature5. 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 criteria7,8. 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.
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 Fautrel⁹, and with those of Yamaguchi¹⁰. 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 present¹. Thus, the presence of fever suggests its association with other febrile conditions². 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. Indeed, both diseases are classified in the group of polygenic auto-inflammatory pathologies¹¹,¹² which are mediated by innate immunity with a high production of pro-inflammatory cytokines (IL-1β, 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 inflammasome¹³,¹⁴.

Nonetheless, despite the pathophysiological links, the association between ankylosing spondylitis and adult Still’s disease is rarely reported⁵,¹⁵. However, this rarity may reflect an underestimation, especially as Cush found sarcroilits in 9.1% of adult Still’s disease in a large review of articles¹⁶. 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 normal¹⁷. 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 Aradoini⁵. 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. 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.
### Conflict of Interest

The authors have no conflicts of interest.

### References


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### 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                  | Country | Year | Number of cases | Age (in years) | Types of spondyloarthritis | Presence of HLA-B27 | Delay between spondyloarthritis and adult Still’s disease onset | Fever | Skin rash | Arthritis | Odynophagia | Leukocytes/mm3 | C-reactive protein (mg/l) | Ferritinemia (ng/ml) | Glycosylated fraction | Rheumatoid factor | Antinuclear antibodies | Infectious tests | Our case |
|--------------------------|---------|------|----------------|---------------|---------------------------|---------------------|----------------------------------------------------------------|-------|-----------|-----------|-------------|----------------|----------------------|---------------------|---------------------|---------------------|-------------|---------------------|
| Duran TI, et al. [18]    | Turkey  | 2020 | 1              | N.F           | A.S : 1                   | 62                  | 10 years                                                        |       | Present  | Absent   | Case 1: Present | 11650          | 300                  | N.F                 | 15%                  | Negative           | Negative             | Negative | N: normal       |
|                          | Senegal | 2021 | 1              | N.F           | A.S : 1                   | 43                  | 15 years                                                        |       | Absent   | Absent   | Case 2: N.F   | 14000          | 192                  | 31 N                | N.F                  | Negative           | Negative             | Negative | N: normal       |

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