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Research Article

Adult-onset Still's Disease in a Health Center: A Report of 10 Cases

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Abstract

Introduction: Adult-onset Still's disease is a rare systemic inflammatory disease in Africa. Observations have been published in Senegal. Materials and Methods: We conducted a retrospective descriptive study to establish the epidemiological profile of patients presenting with adult-onset Still's disease (AOSD) in a health center. The patients included met the diagnostic criteria of Fautrel and/or Yamaguchi. Results: During our study (2020-2024), we included 10 patients with a hospital incidence of 2 patients per year. The average age of the patients was 29.5 years ±11.9, ranging from 15 to 49 years. Females predominated with a sex ratio of 0.67. Clinical manifestations were polymorphic, dominated by fever (100%), inflammatory polyarthralgia (60%), tachycardia (80%), physical asthenia (70%), altered general condition (50%), and erythema (30%). The biological inflammatory syndrome was found in all patients. The glycosylated fraction of ferritin was depleted in all patients. We noted a good outcome after treatment with prednisone 1 mg/kg/day in all patients, methotrexate in six patients, and hydroxychloroquine in one patient. Conclusion: Adult-onset Still's disease is a rare condition, unknown to most practitioners. Its prognosis is severe, especially with visceral involvement.

Keywords

Still’s Disease, Glycosylated Ferritin Fractions, Corticosteroid Therapy

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1. Introduction

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disease characterized by a symptomatic triad of prolonged fever, polyarthritis, and skin rash [1]. It is a rare condition with a prevalence estimated between 0.73 and 6.77 per 100,000 people. It is rarely described in sub-Saharan Africa, although some hospital case series have been reported. The prognosis can be severe, particularly in cases of visceral involvement, which requires appropriate management [2]. We conducted a cross-sectional descriptive study to establish the epidemiological profile of patients with adult-onset Still's disease (AOSD) in a health center.

2. Materials and Methods

We conducted a cross-sectional study in the Department of Internal Medicine at the Mamadou Diop Health Center in Dakar, over 5 years from January 1, 2020, to March 6, 2024.

We included all patients aged 16 years or older, either outpatients or hospitalized in the Department of Internal Medicine, presenting with signs suggestive of adult-onset Still's disease and meeting the diagnostic criteria of Yamaguchi and/or Fautrel. We did not include patients under the age of 16 or those with incomplete medical records. The parameters studied were as follows:

1) Sociodemographic data: including age, sex, and geographic origin.
2) Medical history and background: We studied atopy, consanguinity, diabetes, hypertension (HTN), infectious and systemic diseases (RA, lupus, etc.).
3) Clinical examination:
   a) Functional signs: We looked for fever, asthenia, arthralgia, odynophagia, myalgia, cough, and abdominal pain.
   b) General signs: We assessed temperature, heart rate, blood pressure, respiratory rate, overall condition, skin fold, and mucosal pallor.
   c) Physical signs: We examined all signs across different organ systems.
4) Paraclinical data:
   a) Biological parameters studied included: complete blood count, C-reactive protein, sedimentation rate, fibrinogen, ferritin, glycosylated ferritin fraction, liver function tests, immunological and infectious workup, and tumor markers.
   b) Electrocardiogram (ECG):
   c) Imaging: including chest X-ray, abdominal ultrasound, and thoraco-abdomino-pelvic CT scan.

Data collection was done using a survey form created in Word, and data analysis was performed using Excel 2023 and SPSS software.

3. Results

1) Sociodemographic data:

During our study (2020-2024), we included 10 patients with a hospital incidence of 2 patients per year.

The average age of the patients was 29.5 years ±11.9, ranging from 16 to 49 years. The median age was 29.5 years. The age groups 16-24 years (40%) and 25-34 years (30%) were the most represented. Figure 1 illustrates these results.

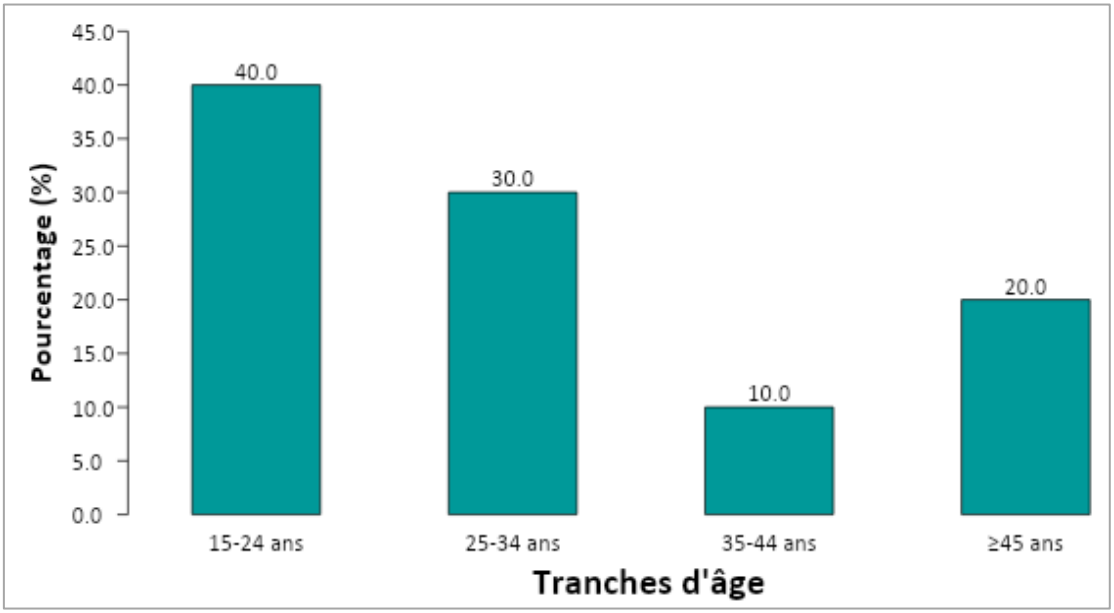

Figure 1. *Distribution of patients by age group (N=10).*

Females predominated (60%) with a sex ratio (M/F) of 0.67. All our patients were from the Dakar region. One patient had a history of pulmonary embolism.

1) Clinical data:

Clinical manifestations were polymorphic and dominated by fever (100%), inflammatory polyarthralgia (60%), and asthenia (70%).

A dermatological examination found erythema in 30% of the patients. It was pruritic in one patient and maculopapular in another. This erythema was located on the face in two patients, the thorax and four limbs in one patient, and the trunk, upper limbs, and cervical region in one patient. Melanoderma was found in one patient. The different clinical manifestations are listed in Table 1.

Table 1. *Distribution of patients according to different clinical signs.*

Clinical Signs	Types of signs	Number (n)	Pourcentage (%)
Functional signs:	Inflammatory polyarthralgia	6	60
	Vomiting	1	10
	Headaches	2	20
	Odynophagia	3	30
	Acute chest pain	2	20
	Dry cough	1	10
General signs:	Physical asthenia	7	70
	Fever	10	100
	Altered general condition	5	50
	Clinical anemia	3	30
	Tachycardia	8	80
	Hypertension	1	10
	Tachypnea	1	10
Physical signs:	Erythematous lesions	3	30
	Melanoderma	1	10
	Systolic murmur	1	10
	Bilateral pulmonary consolidation syndrome	2	20

2) Biological data

The inflammatory syndrome was consistent in our study, observed in most patients. The main biological signs were an increase in C-Reactive Protein (CRP) (100%), hyperleukocytosis (90%), and anemia (50%). The glycosylated fraction of ferritin was depleted in all patients, with levels below 20%. The autoantibody assays and the search for infectious foci were not contributory. The various biological manifestations are listed in Table 2.

Table 2. *Distribution of patients according to biological manifestations.*

Biological signs	Number (n)	Pourcentage (%)
Microcytic anemia	3	30
Normocytic anemia	4	40
Hyperleukocytosis	9	90
Leukopenia	1	10
Thrombocytosis	3	30
Increased CRP	10	100
Accelerated sedimentation rate	2	20
Hepatic cytolysis	6	60
Hyperfibrinogenemia	4	40
Hyperferritinemia	9	90
Depletion of glycosylated fraction	10	100
Increased LDH	3	30

3) Radiological and electrical investigations

The frontal chest X-ray revealed:

1) One case of bilateral basal infiltrative pneumonia with a non-specific appearance, and a right pleural effusion
2) One case of interstitial pneumonia predominantly on the right side
3) One case of bilateral bronchial syndrome

Abdominal ultrasound revealed in three patients:

1) One case of multiple retroperitoneal coelio-mesenteric and inter-aorto-caval lymphadenopathies
2) One case of splenomegaly and non-necrotic adenopathies
3) One case of terminal ileitis

The electrocardiogram was performed on three patients and revealed:

1) One case of ST-segment elevation with a concave shape in the anteroseptal region and ST-segment depression in the inferior region
2) One case of incomplete right bundle branch block
3) One case of anterior hemiblock left atrial and left ventricular hypertrophy, with a negative T wave in the lateral leads

All patients met the Yamaguchi and Fautrel criteria. The

manifestations according to the diagnostic criteria are listed in Table 3.

Table 3. *Distribution of patients according to classification criteria.*

Major criteria	Number of cases (n)	Pourcentage (%)
Fever higher than 39°C for more than 1 week	10	*100,0*
Leukocytosis with 80% neutrophils	10	*100,0*
Arthralgia or arthritis lasting 2 weeks or more	5	*50,0*
Typical skin rash	4	*40,0*
Minor criteria	Number of cases (n)	Pourcentage (%)
Absence of rheumatoid factor and antinuclear antibodies	10	*100,0*
Hepatic dysfunction	7	*70,0*
Odynophagia	3	*30,0*
Lymphadenopathy or splenomegaly	2	*20,0*

4. Treatment

Corticosteroid therapy was prescribed for all patients; it consisted of prednisolone at 1 mg/kg/day for all patients and one case of methylprednisolone bolus at 5 mg/kg/day for 3 days.

Analgesics were prescribed in 90% of cases, consisting of paracetamol (1g) and injectable tramadol (10mg).

Immunosuppressants were prescribed in 70% of cases, including 6 cases on methotrexate (10 mg/week) and 1 case on hydroxychloroquine (400 mg/day in two doses).

In terms of progression, we observed nine cases of systemic forms, including two polycyclic forms and one case of chronic articular form. The systemic forms were marked by four complications: two cases of macrophage activation syndrome, one case of acute cholestatic hepatitis, and one case of acute myocarditis. The activity was evaluated with the SDAS. Good progress was noted in most patients.

5. Discussion

During the study period (2020 to 2024), 10 patients were recorded, representing an incidence of 2 patients per year. This is consistent with the annual incidence of adult-onset Still's disease (AOSD), which is estimated to be between 1 and 10 new cases per million inhabitants, depending on the country, according to the study by Fautrel B [2]. Lower rates, 1.04 patients/year and 1.2 patients/year, have been reported in Senegalese [3] and Cameroonian [4] series.

The incidence in our series, higher compared to the Senegalese series reported 2 years earlier [3], could be explained by improved diagnostic tools, with the availability of glycosylated fraction testing in our facility, which has also reduced its relatively high cost.

Juvenile age is described in most studies. In our series, the mean age was 29.5 years ±11.9. Similar results were found in the Cameroonian series of 12 cases by Singwé et al., with a mean age of 28 years [4], as well as in the Gabonese series by Josaphat Iba Ba et al., with a mean age of 24.7 years [5].

Certainly, the mean age observed in our study was lower than those found in the series by S. Diallo, which reported a mean age of 43 years [3], and in the study by Frikha O, which reported a mean age of 36.49 years [6].

Our study revealed a predominance of females, with a sex ratio of 0.67. This is consistent with the literature on adult-onset Still's disease (AOSD). In a study by Diallo et al., a sex ratio of 0.26 was reported in a cohort of 24 patients followed in Dakar [3]. Similar rates were observed in a Tunisian series of 65 cases, with a sex ratio of 0.25 [6].

Our study revealed a predominance of the following signs: inflammatory polyarthralgia (60%), fever (100%), and physical asthenia (70%). These findings are consistent with descriptions in the literature. Fever is a constant feature of AOSD, usually hectic, with temperatures often exceeding 39°C. It typically occurs in the late afternoon, with apyrexia between febrile peaks. Fever can be associated with weight loss and altered general condition. Weight loss is found in 47.8% of cases, and polyarthralgia in 99.4% of cases. Articular manifestations may be delayed in certain circumstances [7].

Skin manifestations are the third cardinal sign of AOSD. Among the 10 patients in our study, 40% (n=4) had cutaneous lesions, sometimes pruritic, consisting of erythematous rashes (50%) and maculopapular rashes (25%), with one case (25%) of diffuse melanoderma. These lesions were primarily located on the trunk, four limbs, and face. This skin manifestation is frequently reported in studies and can sometimes lead to the discovery of the disease [8, 9].

Other common manifestations of AOSD include lymphadenopathy (50%), splenomegaly (40%), hepatomegaly (30%), myalgia (45%), pleurisy (20%), pericarditis (15%), weight loss (30%), and abdominal pain (20%). Other, more anecdotal conditions have also been reported, such as organized pneumonia and meningoencephalitis [10].

The inflammatory syndrome was present in all our patients. In addition to the previously mentioned hyperleukocytosis, our series noted: increased CRP levels in all patients (100%), thrombocytosis in 3 cases (30%); the sedimentation rate was available for 2 patients and was elevated at 70 and 85 mm/h (20%), and fibrinogen levels were available for 5 patients, with elevated levels in 4 patients (80%). These results are similar to those reported in several other series, including the study by Frikha O, which

found a consistent biological inflammatory syndrome with hyperleukocytosis in 87.3% of cases [6].

Biological markers of inflammation are consistently elevated in AOSD. The increase in ferritin levels, along with the depletion of its glycosylated fraction, is a key factor in diagnosing AOSD. These findings are well recognized and used by most authors for diagnosing the disease [11-13].

Regarding treatment, corticosteroid therapy was used as the first-line treatment in all of our patients (100%). Immunosuppressants, such as steroid-sparing agents, were used in 7 of our patients (70%), including 6 cases with methotrexate and 1 case with hydroxychloroquine.

However, the frequency of methotrexate use in our series was justified according to current data and allows for remission in certain cases [1, 6, 10].

Similarly, previous studies are in agreement with this management approach, particularly those of S. Diallo et al. and Liu et al. [3].

In the series reported by O. Frikha et al., treatment was based on corticosteroid therapy as first-line treatment in all patients, with methotrexate prescribed as second-line therapy for 28 patients (43.1%) [6]. New therapies such as biotherapies have been shown to be highly effective in treating adult Still's disease. They often involve inhibitors of IL-1, IL-6 (tociluzumab), IL-18 and IL-17, as well as inhibitors of TNFα or Janus kinases (JAKs), administered to patients who do not respond to CS and DMARDs or who do not obtain an adequate response [14-16]. Unfortunately, these innovative molecules are inaccessible in Africa, despite all the efforts made by practitioners to disseminate knowledge about this disease [17-19].

6. Conclusion

Adult-onset Still's disease (AOSD) is a rare and challenging condition to diagnose due to the non-specificity of its clinical presentation. In our study, the incidence of AOSD was consistent with global estimates, though slightly higher than previously reported in Senegal. Improved diagnostic tools, such as the measurement of glycosylated ferritin fractions, have likely contributed to this increase in diagnosis. The disease predominantly affects younger patients, with a female predominance. Treatment typically involves corticosteroids, with immunosuppressants used in steroid-sparing strategies. The outcomes in our series were favorable, with no recorded fatalities, highlighting the importance of early diagnosis and appropriate management to prevent complications. Further research and awareness are needed to improve recognition and outcomes in AOSD.

Abbreviations

AOSD	Adult-onset Still's Disease (AOSD)
CT Scan	Computed Tomography Scan
HTN	Hypertension
RA	Rheumatoid Arthritis
ECG	Electrocardiogramme
mg	Milligram
LDH	Lacticodeshydrogenase
AH	Arterial Hypertension
LDH	Lactate Dehydrogenase
ESR	Erythrocyte Sedimentation Rate
SDAS	Still's Disease Activity Score

Conflicts of Interest

The authors declare no conflicts of interest.

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