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Clinical case

Lymphoma followed by Still's disease in adults: an unusual case in Chad

Lymphome suivi d'une maladie de Still de l'adulte : un cas inhabituel au Tchad

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Résumé

Introduction : L'association entre maladie de Still de l'adulte (MSA) et lymphome est rare et source de difficultés diagnostiques.

Cas clinique : Patiente de 31 ans, suivie pour lymphome diffus à grandes cellules B en rémission, qui a développé 16 mois plus tard une MSA confirmée (critères de Yamaguchi, ferritine glycosylée < 20 %). L'évolution a été favorable sous corticothérapie et méthotrexate avec rémission durable.

Conclusion : Ce cas illustre une séquence inhabituelle (lymphome → MSA) et souligne l'importance d'exclure une rechute hématologique devant un tableau fébrile avec arthrites et rash.

Mots-clés : Maladie de Still de l'adulte ; Lymphome ; Ferritine glycosylée ; Critères de Yamaguchi ; Méthotrexate ; Tchad.

Abstract

Introduction: The association between adult Still's disease (MSA) and lymphoma is rare and a source of diagnostic difficulties.

Clinical case: A 31-year-old patient with diffuse large B-cell lymphoma in remission, who developed a confirmed ASM 16 months later (Yamaguchi criteria, glycosylated ferritin < 20%). The course was favourable on corticosteroid therapy and methotrexate with durable remission.

Conclusion: This case illustrates an unusual sequence (lymphoma → MSA) and underlines the importance of excluding a haematological relapse in the presence of a febrile picture with arthritis and rash.

Keywords: Still's disease in adults; Lymphoma; Glycosylated ferritin; Yamaguchi criteria; Methotrexate; Chad.

Introduction

Adult Still's disease (MSA) is a rare systemic disorder of unknown etiology, first described by Bywaters in 1971, which occurs in both young adults and children

[1]. It is clinically characterized by the association of high fever occurring by evening peaks, polyarthritis or arthralgias and an evanescent maculopapular skin rash contemporaneous with febrile attacks [2].

The diagnosis of MSA remains difficult, because it is based on non-specific clinical and biological criteria, such as those proposed by Yamaguchi [6] or Fautrel [7]. Among these criteria is the mandatory exclusion of an infection, an autoimmune disease and especially a hematologic malignancy, in particular lymphomas [3,5]. However, MSA is not one of the rheumatological syndromes classically associated with a high risk of neoplasia, which makes its association with lymphoma exceptional.

The relationship between MSA and hematologic malignancies, particularly non-Hodgkin's lymphomas and, more rarely, leukemias, has been reported in the literature [4,8,9,14]. This relationship can manifest itself in several scenarios: MSA precedes hemopathy, it occurs simultaneously, or more rarely it appears after lymphoma remission [3]. The distinction between these two entities is delicate, especially since certain clinical manifestations (fever, polyadenopathy, alteration of general condition, biological abnormalities) are common.

We report here the case of a patient who presented with MSA 16 months after diffuse large B-cell lymphoma in complete remission, an even rarer situation, and propose an update of the literature on this association.

Clinical case

We report the case of a 31-year-old patient, with no notable pathological history, hospitalized in 2021 for a febrile polyarthritis picture associated with a localized evanescent maculopapular rash on the elbows, arms and neck, as well as cervical polyadenopathy evolving in a context of deterioration in general condition.

Initial assessment: The assessment revealed an inflammatory syndrome (ESR: 51 mm, CRP: 26 mg/L, fibrinogen: 8.21 g/L), hypoalbuminemia (23.1 g/L) with polyclonal hypergammaglobulinemia. The blood count was normal (Hb: 13.2 g/dL; leukocytes: 9600/mm³ including 6100 PNNs; normal platelets). The ferritin was 385 ng/mL. The infection (HIV, EBV, CMV, hepatitis B and C) and autoimmune (antinuclear antibodies, ANCA, anti-CCP) were negative.

A lymph node biopsy revealed diffuse large B-cell lymphoma. The cervico-thoraco-abdominal CT scan showed supra- and subdiaphragmatic lymphadenopathy. The diagnosis of diffuse large B-cell lymphoma was retained and the patient was treated in haematology with chemotherapy, achieving complete remission.

Secondary course: Sixteen months after remission, the patient again presented with febrile polyarthritis, an evanescent maculopapular rash in the arms and neck, and bilateral laterocervical lymphadenopathy.

The assessment showed a major inflammatory syndrome (ESR: 100 mm, CRP: 68 mg/L, hypoalbuminemia), leukocytosis at 14,100/mm³ (PNN: 10,400/mm³), normocytic anaemia at 10 g/dL, and ferritinemia > 1500 ng/mL with glycosylated fraction < 20%. The infectious and immunological assessments remained negative. Imaging showed only bilateral cervical lymphadenopathy, with no evidence of lymphoma relapse.

Diagnosis and management: In view of the association of evening fever, polyarthritis, suggestive rash, lymphadenopathy and hyperferritinemia with collapsed glycosylated fraction, and in the absence of an argument for a relapse of lymphoma or an infectious/autoimmune cause, the diagnosis of Still's disease in adults was retained (Yamaguchi criteria met).

Bolus treatment with corticosteroids, followed by oral corticosteroid therapy (1 mg/kg/day), led to partial improvement. The addition of methotrexate 15 mg/week allowed a favourable course with disappearance of fever, arthritis and rash, as well as a normalization of biological parameters in 8 weeks.

After 3 years of follow-up, the patient remained asymptomatic, with a strictly normal clinical examination and laboratory tests. The assessment of quality of life by the SF-36 and NHP questionnaires showed a significant initial alteration in the physical and psychosocial dimensions (pain, vitality, mobility), a significant improvement from the 8th week, and then a complete recovery. The results of the quality of life questionnaires are presented in Tables III and IV.

Table I: Reported cases of association of adult Still's disease and lymphoma

Author(s) / Reference	Country	Age / Gender	Sequence (MSA vs Neoplasia)	Type of Neoplasia	Treatment (MSA/ Neoplasia)	Evolution
Trotta et al.	Italy	52/F	MSA → NHL	Large B-cell NHL	CC (IR) / Chemotherapy	RC
Kawasaki et al.	Japan	58/F	MSA → NHL	NHL T	Batch CC / Chemotherapy	RC
Bimbalal et al.	India	19/M	Simultaneous	NHL (Angiotropic)	CC / Chemotherapy	Death
Sono et al.	Japan	50/M	MSA → NHL	NHL B	CC, CPM, MTX / Chemotherapy	Refractory
Otrocol et al.	Romania	42/F	MSA → NHL	NHL B	CC / Chemotherapy	Relapse
Kato et al.	Japan	25/M	Simultaneous	NHL NK	CC / Chemotherapy	No rating yet
Gratton and Imboden	USA	23/F	MSA → Hodgkin	Hodgkin's disease	NSAIDs / Chemotherapy	No rating yet
Voinchet et al.	France	38/M	MSA → NHL	NHL marginal area	CC / Chemotherapy	RC
Smaali et al. (PAMJ 2020)	Morocco	28/M	NHL→ MSA	Large B-cell NHL	CC + MTX / Chemotherapy	RC
Cabane et al. (1988)	France	2 cases	MSA ↔ Neoplasia	Carcinoma of the larynx	—	Variable evolution
Ahn et al. (2010)	Korea	52/M	Simultaneous	Papillary thyroid cancer	CC / Surgery	RC
Shibuya (1997)	Japan	77/M	MSA → Cancer	Esophageal cancer	CC / Surgery	Death
Jardin et al. (2005)	France	Series	MSA ↔ Lymphoma	Miscellaneous NHL	—	Diverse
Josaphat Iba et al. (2023)	Gabon	10 cases (22 years ±)	MSA alone	—	CC ± NSAIDs	Favourable development
Bayala et al. (2024)	Burkina Faso	23/F	Simultaneous	Non-Hodgkin's NHL	CC / Chemotherapy	RC
In this case, Garba H.A. et al.)	Chad	31/F	NHL→ MSA	Large B-cell NHL	CC + MTX / Chemotherapy	RC (3 years)

Table II: Diagnostic criteria for Still's disease in adults according to Yamaguchi and Fautrel

Criteria	Yamaguchi (1992)	Fautrel (2002)
Major criteria	- Fever ≥ 39 °C ≥ 1 week- Arthralgias ≥ 2 weeks- Typical skin rash- Hyperleukocytosis> 10,000/mm³ with PNN ≥ 80%	- Fever ≥ 39 °C- Arthralgias- Typical skin rash- Pharyngitis- Polynucleosis > 80 %- Glycosylated ferritin ≤ 20 %
Minor criteria	- Pharyngitis- Adenopathy and/or splenomegaly- Hepatic damage- Biological abnormalities (transaminases, LDH, etc.)- Rheumatoid factors and negative ANA	- Elevated polynucleosis- ALT/AST- Rheumatoid factors and negative ANA
Mandatory exclusion	Infections, neoplasias, autoimmune diseases	Infections, neoplasias, autoimmune diseases
Diagnostic threshold	≥ 5 criteria, including ≥ 2 major	≥ 4 majors, or 3 majors + 2 minors

Table III: Results of the SF-36 questionnaire according to clinical course

SF-36 Domains	At relapse (MSA diagnosis)	At 8 weeks	At 3 years of follow-up
Physical function	45/100	70/100	90/100
Limitation due to physical condition	40/100	65/100	90/100
Body pain	30/100	60/100	85/100
General Health	35/100	60/100	80/100
Vitality (energy/fatigue)	40/100	65/100	85/100
Social function	50/100	70/100	90/100
Limitation due to emotional state	55/100	75/100	90/100
Mental health	60/100	75/100	90/100

The SF-36 scores show a rapid improvement in quality of life from 8 weeks on corticosteroid therapy and methotrexate, consolidated at 3 years with almost complete recovery. Scores are expressed from 0 (highly altered state) to 100 (optimal state).

Table IV: Nottingham Health Profile (NHP) results according to the evolution

NHP Domains	At relapse	At 8 weeks	At 3 years old
Energy	55 %	35 %	10 %
Pain	60 %	30 %	5 %
Emotional reactions	45 %	30 %	10 %
Sleep	50 %	35 %	10 %
Social isolation	40 %	25 %	5 %
Physical mobility	50 %	30 %	5 %

The NHP scores confirm a significant initial alteration (pain, energy, sleep), followed by a clear improvement from 8 weeks and almost complete at 3 years. Values are expressed as a percentage of perceived limitation; A high score indicates significant impairment.

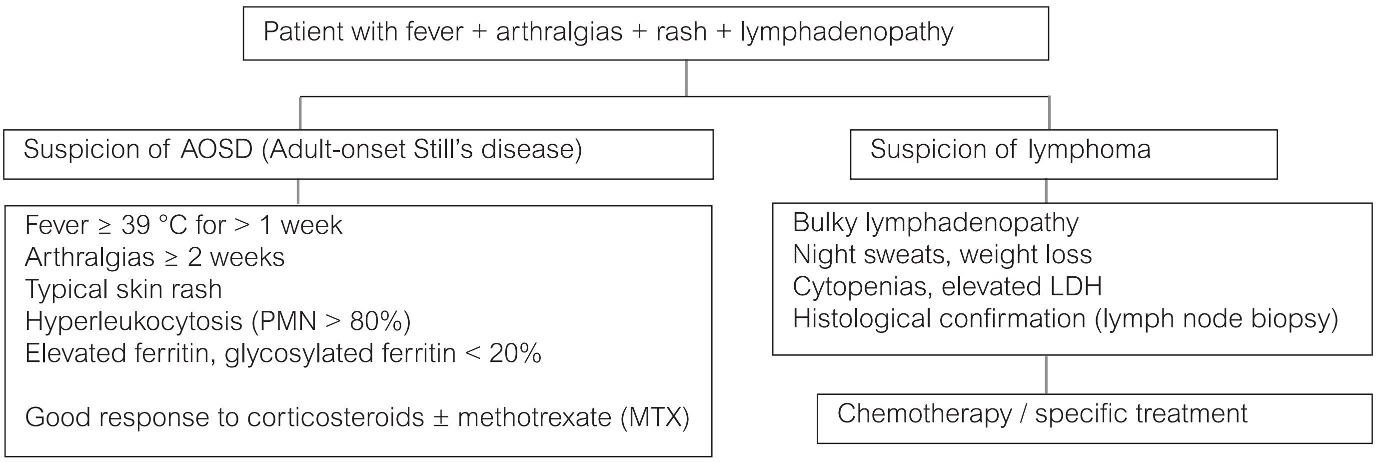

Figure 1. Comparative flowchart between adult Still's disease (MSA) and lymphoma. This diagram illustrates the diagnostic approach in the case of a patient with fever, arthralgia, rash and lymphadenopathy, highlighting the discriminating criteria and the importance of glycosylated ferritin.

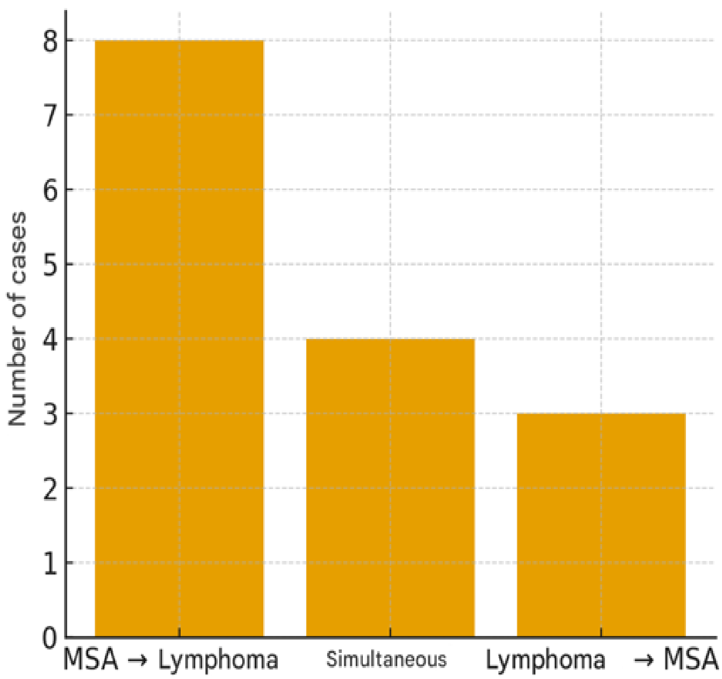

Figure 2. Distribution of reported cases of association of adult Still's disease (MSA) and lymphoma according to time sequence (MSA preceding lymphoma, occurring simultaneously, or lymphoma preceding MSA).
It highlights the relative rarity of cases where MSA appears after lymphoma remission.

Discussion

Adult Still's disease (MSA) is a rare and complex clinical entity, the diagnosis of which is based on a range of clinical, biological and exclusion arguments [1,2]. One of the main challenges is to differentiate this condition from malignant pathologies, in particular non-Hodgkin's lymphomas, which share several common characteristics: fever, polyadenopathy, alteration of general condition, biological abnormalities [3,5]. In this context, the elimination of a hematologic malignancy is an essential prerequisite before the diagnosis of MSA is retained.

The association between MSA and neoplasia remains rare, but it is described in the literature in different forms: MSA precedes lymphoma or solid neoplasia, both conditions occur simultaneously, and more rarely, MSA appears after lymphoma remission [6–9]. Our observation illustrates this third, more unusual situation. The patient, initially followed for diffuse large B-cell lymphoma, developed a typical MSA 16 months later, confirmed by the Yamaguchi criteria and the demonstration of hyperferritinemia with glycosylated fraction < 20%, a biomarker recognized as particularly suggestive [6,7]. This reverse sequence (MSA → lymphoma) has been reported only in rare cases, notably in the series analyzed by Smaali et al. in Morocco in 2020, highlighting the added value of our observation in the African context. [11:12].

Glycosylated ferritin is a major discriminating element. While total ferritin can be increased in many inflammatory or neoplastic settings, a collapsed glycosylated fraction (< 20%) is strongly suggestive of an active MSA [6]. In our case, this biological abnormality helped to guide the diagnosis and avoid confusion with lymphoma relapse. [7-13].

From a therapeutic point of view, corticosteroid therapy remains the cornerstone of the treatment of MSA. However, in the case of a partial response or corticosteroid-dependent forms, the addition of a methotrexate is effective, as observed in our patient, with complete and sustained clinical and biological remission achieved over three years of follow-up. [11-14].

This observation reminds us of the need for close follow-up and systematic re-evaluation of patients with a history of lymphoma and a febrile picture with joint involvement. In such contexts, it is essential not to hastily conclude that a haematological relapse has occurred, but to consider the possibility of MSA, the management of which differs radically. [12-14]. Quality of life analysis is an essential element in assessing the overall impact of MSA and the effectiveness of treatment. In our observation, the SF-36 [15] and NHP [17] scores showed rapid and then sustained improvement after initiation of treatment, reflecting the restoration of the patient's functional and psychosocial capacities, as also suggested by recent multicenter data in people with MSA [16]. Few African studies systematically document these dimensions of quality of life; Their integration into follow-up usefully complements clinical and biological evaluation and could refine the measurement of therapeutic response and long-term prognosis [18].

Conclusion

The association between adult Still's disease and lymphoma remains rare but represents a major diagnostic challenge. Our observation illustrates an unusual sequence, where MSA occurs after remission of diffuse large B-cell lymphoma. It underlines the need for rigorous clinical and paraclinical monitoring, as well as the decisive contribution of glycosylated ferritin to differentiate this entity from a haematological relapse. Management combining corticosteroid therapy and methotrexate has led to a lasting remission, reinforcing the importance of accurate diagnosis and appropriate treatment.

Statements

Authors' contributions

All authors participated equally in the design, writing, critical review, and final approval of the manuscript. The lead author provided coordination and submission.

Informed consent

The patient's informed consent was obtained for the publication of this observation, in respect of anonymity and confidentiality.

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