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Systemic Form of Juvenile Idiopathic Arthritis: Epidemiological, Clinical, Paraclinical and Therapeutic Aspects of 13 Cases in Abidjan

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Abstract

Objective: To describe the epidemiological, clinical, paraclinical and therapeutic aspects of systemic juvenile idiopathic arthritis observed in Abidjan. **Materials and Method:** This retrospective and descriptive study covered 13 children suffering from systemic juvenile idiopathic arthritis selected in the Rheumatology Department of University Hospital Center of Cocody in Abidjan (Cote d'Ivoire) from January 2005 to December 2015. We were interested to the sociodemographical, clinical, paraclinical and therapeutic aspects. **Results:** The systemic form of the juvenile idiopathic arthritis represented 0.2% of the 4608 rheumatologic diseases and 70.58% of the JIA. We selected 6 boys and 7 girls, with an average age of 10.8 years and mostly going to school (84.61%). The diagnostic delay was 18 months. The main clinical signs were fever and joint damage observed each in 100% of cases, impaired general condition (92.30%) and tumor syndrome (83.33%). Biological signs were characterized by hyperleukocytosis (69.20%) and the presence of a biologic inflammatory syndrome (on average, erythrocyte sedimentation rate 59.6 mm and C Reactive Protein 56.4 mg/l). The cervical damage was the essential functional complication (38.46%). The major treatment has been a therapeutic combination based on corticotherapy and methotrexate (100%) with 1 death case by macrophage activation syndrome. **Conclusion:** Systemic juvenile idiopathic arthritis is rarely diagnosed in the rheumatologic practice in Abidjan. It concerns children relatively big, and is characterized by a febrile polyarthritis with impaired general condition and tumor syndrome. This systemic form is

treated by corticotherapy and methotrexate.

Keywords

Systemic Idiopathic Juvenile Arthritis, Juvenile Idiopathic Arthritis, Profile, Children, Abidjan

1. Introduction

Juvenile idiopathic arthritis (JIA) is a heterogeneous set of chronic inflammatory rheumatism beginning before the age of 16 and grouping together 6 groups of arthritis (systemic arthritis, oligoarthritis, polyarthritis with positive rheumatoid factors, polyarthritis with negative rheumatoid factors, arthritis with enthesitis, psoriatic arthritis) to which is added a last group of unclassable arthritis [1]. They involve the functional prognosis of the child as well as the vital prognosis especially on the most severe form which is the systemic form (SF-JIA) or Still's disease of child. Still's disease has been described for the first time on children (SF-JIA) but it appears to be less frequent in this age group, judging by the numerous publications about this disease in adults (Still's disease of adult) [2]-[9]. It has evolved conceptually and is now classified as an autoimmune disease [10]. It represents 4% to 17% of all JIA [11]. In the Maghreb region of Africa, we notice several publications on JIA [12] [13] [14] [15]. In sub-Saharan Black Africa, few studies have been published on JIA in their whole, but they remain poorly known as SF-JIA, too [16] [17] [18] [19]. To our knowledge, only one study has been devoted specifically to this SF-JIA in Sub-Saharan Black Africa [20]. The quest for a better knowledge, in our context, guided the realization of this study whose objective was to describe the epidemiological, clinical, paraclinical and therapeutic aspects of the SF-JIA observed in Abidjan.

2. Materials and Method

A retrospective descriptive study was conducted in the department of rheumatology of University Hospital Center of Cocody during a period of 10 years from January 2005 to December 2015. We enrolled 13 children with SF-JIA. The records of patients with infectious arthritis, metabolic, autoimmune or tumor origin and those without articular imaging were not included. The diagnosis of SF-JIA was based on signs according to the 1997 Durban criterias revisited in Edmonton in 2001 [11] [21]. For each observation, we analyzed the following aspects:

- Sociodemographical aspects: age, sex, school level;
- Clinical aspects: morbid past medical history, duration of disease, reason for hospitalization, axial and/or peripheral joint damages and general or visceral extra-articular injuries;
- The paraclinical aspects: biological (Blood count, erythrocyte sedimentation rate (ESR), C reactive protein (CRP), transaminase, serum ferritin), radio-

logical (joint and thoracic standard x ray, cardiac echography);

- The therapeutic aspects: non-steroidal anti-inflammatory drugs (NSAIDs), corticotherapy, methotrexate;
- The prognostic aspects and evolution: functional prognosis, prognosis of life, evolution of the disease under treatment.

3. Results

Our sample was constituted of 6 boys and 7 girls with an average age of 10.8 years (Extremes: 3 and 15 years). This sample size represented 0.2% of the 4608 rheumatologic diseases identified during the study period. Patients attending school were from primary school (5 cases) and secondary school (college in 6 cases) and 2 patients were not schooling. The delay of diagnosis of the disease was on average 1 year and 6 months. An impaired general condition was present in 12 of 13 cases. Microcytic anemia was observed in all cases with a mean hemoglobin level of 10 g/dl. We noticed the presence of an inflammatory biological syndrome with a mean ESR and CRP respectively of 59.6mm at first hour and 56.4 mg/l (respective extremes: ESR 40 et 101 mm et CRP 26 et 142 mg/l). The assessment of autoimmunity (rheumatoid factors, anti-CCP antibodies and antinuclear factors) was performed in 3 cases and was negative. The main clinical, paraclinical and therapeutic data are listed in Table 1 and Table 2.

4. Discussion

4.1. At the Socio-Demographic Level

The weakness of our sample size is a limit to make it a generalization. However, it provided information on the rarity of JIA in general, then on SF-JIA in our African context. Other studies on this subject had made the same observation

Table 1. observations summary.

Parameters patients	Age sex	Elevated fever ≥ 39˚C	Polyarthritis	Skin rash	Angina or odynophagia	Tumor	Complications
1	4 M	Yes	Yes	Yes	No	HPM SPM	
2	10 F	Yes	Yes	Yes	No	ADP	Cervical damage
3	10 M	Yes	Yes	Yes	No	No	
4	8 F	Yes	Yes	No	Yes	ADP	Cervical damage
5	3 M	Yes	Yes	No	No	ADP	SPD Cervical damage
6	11 F	Yes	Yes	No	No	SPM	
7	15 F	Yes	Yes	Yes	No	No	
8	15 F	Yes	Yes	No	No	ADP	
9	15 M	Yes	Yes	Yes	No	No	
10	12 M	Yes	Yes	No	No	ADP HPM	
11	15 F	Yes	Yes	No	No	HPM	Cervical damage
12	14 F	Yes	Yes	Yes	Yes	ADP	Cervical damage
13	09 M	Yes	Yes	No	Yes	ADP	

M (male); F (female); HPM (hepatomegaly); SPM (splenomegaly); ADP (adenopathy); SPD (staturo-ponderal delay).

Table 2. Continued from the summary of observations.

Parameters patients	White blood cells	Transaminases	Ferritinemia	Pleurisy	Pericarditis	Osteo-articular lesions	Treatment
1	Hyperleukocytosis	Normal	Increased	No	No	Absent	C + M
2	Hyperleukocytosis	Normal	Increased	No	No	Absent	C + M
3	Hyperleukocytosis	Normal	Increased	Yes	No	Bones erosions	C + M
4	Hyperleukocytosis	Normal	Normal	No	No	Absent	C + M
5	Normal	Normal	Normal	No	No	Absent	C + M
6	Normal	Normal	Increased	No	No	Absent	C + M
7	Normal	Normal	Normal	No	No	Absent	C + M
8	hyperleukocytosis	Increased	Increased	No	No	Absent	NSAID then C + M
9	Hyperleukocytosis	Normal	Normal	No	No	Absent	NSAID then C + M
10	Hyperleukocytosis	Increased	Increased	No	No	Absent	NSAID then C + M
11	hyperleukocytosis	Normal	Increased	No	No	Bones erosions	NSAID then C + M
12	Normal	Normal	Normal	Non	Non	Absent	NSAID then C + M
13	Hyperleukocytosis	Normal	Increased	No	No	Absent	C + M

NSAID (nonsteroidal anti inflammatory drugs); C + M (corticoid and methotrexate).

[16] [22] [23] [24]. The SF-JIA remains unknown in sub-Saharan Africa because it gives diagnostic problems due to the absence or deficit of pediatric rheumatologist. A study on JIA in general, carried out in Côte d'Ivoire showed a high frequency of SF-JIA, as in Togo and Saudi Arabia, respectively, representing 70.58%, 83.33% and 36.5% of cases [16] [25] [26]. However, in other sub-Saharan black African countries, the frequency was much lower: 2% in Senegal, 13.04% in Nigeria and 14.10% in Zambia [18] [19] [24]. This frequency variability would be explained by the intervention of environmental, genetic and ethnic factors [26]. In western countries, it is the oligoarticular form which predominate and SF-JIA represented 5 up to 15% of JIA [27] [28] [29]. The SF-JIA affected as well male as female as in most cases [28]. The mean age of the disease arise appeared rather high in our study (10.8 years) compared to other countries: 6.3 years in Senegal, 8 years in South Africa, 11.3 +/− 3 years in Morocco, 6.6 years in France [13] [20] [30] [31]. The lack of knowledge of children's rheumatologic disorders and their almost systematic orientation in the pediatric services in our context could justify this diagnostic delay compared to Europe where the disease is better known. Nonetheless, the study of Diallo *et al.* carried out in Senegal was an exception with an age identical to that found in France [20].

4.2. At the Clinical, Paraclinical and Diagnostic Level

The SF-JIA is characterized by a clinico-biological polymorphism associating rather evocative signs such as the fever and the joint involvement, observed in 100% of our cases. These associated with the evanescent rash, constitute the classical presentation of this disease [32]. When an impaired general condition was added as was the case in 12 of our patients, these signs reflected the systemic nature of the disease [20] [32]. On the biological level, apart from hyperferritine-

mia and the decrease of its glycosylated fraction, which could be considered as an evocative sign [33] [34], the biological inflammatory syndrome and neutrophilic hyperleukocytosis remain constant. The other clinico-biological signs were observed at various degrees in our study as described by several authors [35] [36] [37].

4.3. Radiologically

This form and the polyarticular form are the two major forms of bone destruction [38] [39]. This structural damage is characterized by radiographic bones erosions observed in 2 of our patients.

These structural destructions represent a functional evolutionary challenge of the disease because the functional prognosis is engaged. The illustration in our study was a cervical damage marked by ankylosis of the posterior articular masses in 5 cases and a destructive coxitis in 1 case. These last localizations constitute with the temporomandibular injury, not observed in our study, the 3 major functional and prognostic impairments of the disease. The staturo-ponderal delay observed in 1 of our patients and osteoporosis are other potential complications that threaten these patients due to the long-term use of corticosteroids as a basic symptomatic treatment. The vital prognosis is singularly engaged in the case of macrophage activation syndrome (MAS), which represent 10% of the systemic forms. It is the most important complication of SF-JIA because it can be fatal [40] [41]. In our study, the only case of MAS occurred in a male child and was fatal. The rare African JIA studies did not identify MAS apart from the Diallo's study which showed 3 cases out of 16 cases of Still's disease (study involved children and adults) [20]. Elsewhere in the West, the study of Minoa *et al.* revealed 22.2% of MAS cases [41].

4.4. Therapeutically

All our patients had as treatment the combination corticoid and methotrexate with a rather favorable evolution outside the case of death. It is the basic conventional treatment of any systemic form of JIA [14] [42] [43]. Refractory forms are eligible for other therapies such as biotherapy based on anti-TNF alpha and especially anti-interleukin 6 or anti-interleukin 1 [14] [42]. Biotherapy is common practice under skies. Its excessive cost and our context of tuberculosis endemic area are the main limiting factors.

5. Conclusion

SF-JIA is very rare in rheumatologic practice in Abidjan but is the most seen JIA form. It affects schooling children at a relatively high age. It is manifested in our context, by a clinico-biological polymorphism characterized by fever, polyarthritis, impaired general condition with more or less a tumor syndrome and a hyperleukocytosis with polynucleosis. This disease is marked by a functional complication mainly cervical involvement and a vital complication which is a macrophage activation syndrome. Its management involves corticosteroid ther-

apy and methotrexate.

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