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Sydenham's Chorea: Apropos of Three Observed Cases at the N'Djamena Mother and Child University Teaching Hospital (CHU-ME)-Chad

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Abstract

Introduction: Post-streptococcal chorea is the most common form of acquired chorea in children. **Objective**: The objective of this study was to contribute to better management of this condition by giving the characteristics of the epidemiological, clinical, biological, etiological, therapeutic and evolutionary aspects. **Methodology**: This was a retrospective and descriptive study from January 2021 to December 2021 which took place in the pediatrics department of the Mother and Child University Teaching Hospital of N'Djamena (CHU-ME). All the children in whom we observed abnormal involuntary movements whose age was less than or equal to 15 years were concerned. **Result**: there were three female children. The average age was 7.5, of which the two are 7 years old and the third is 8 years old. Two had a history of angina. A notion of polyarthralgia and dental caries was found in the third. The diagnosis of post-streptococcal chorea was retained on the basis of clinical arguments: observation of abnormal movements of the limbs and the face, which are involuntary, sudden with an insidious and progressive onset, muscular hypotonia, and psychic disorder. Medical imaging, in particular echocardiography, which objectified two cases of associated rheumatic valvular disease. Remission was observed in all three children after two weeks of treatment with haloperidol, penicillin. Those with associated rheumatic valve disease also received corticosteroid therapy. Complete remission of chorea was observed in

all three girls. Prophylaxis in two patients based on Penicillin V was instituted. **Conclusion**: Although less frequent, post-streptococcal chorea is still seen in our country and is still an important health problem that needs more real medical efforts.

Keywords

Sydenham Chorea, AAR, Streptococcus, CHU-ME, Chad

1. Introduction

Post-streptococcal chorea, also called Sydenham's chorea, or rheumatic chorea, is the most common form of acquired chorea in children. For the first time it was described by Thomas Sydenham in 1686 [1]. Chorea derives from the Latin word "choreus" which means "dance" [2]. Its characteristic clinical picture is marked by involuntary movements, linked to the contraction of several muscles. They are ample, irregular, can sometimes be limited to half of the body and can even involve the face [3]. The movements appear gradually or suddenly, predominating in the upper limbs. Exacerbated by fatigue, the movements disappear during sleep. The evolution is unpredictable, interspersed with relapses which can last for about ten years with the risk of persistence of residual character disorders [4]. However, the seriousness of the condition lies in the occurrence of valvular damage which constitutes the major progressive risk [2] [5]. This disease is predominant in girls [2].

In developed countries, its incidence has decreased, in parallel with that of AAR following the judicious use of antibiotics in the treatment of angina [6] [7]. In developing countries such as Chad, the abnormal movements observed in children often pose diagnostic and therapeutic difficulties. Abnormal movements can be of variable etiology and Sydenham's chorea represents the leading cause of acquired chorea in children in the Third World [1]. The objective of this study is therefore to contribute to better management of this condition by giving the characteristics of the epidemiological, clinical, biological, etiological, therapeutic and evolutionary aspects of children followed for Sydenham's chorea in the pediatric department of the N'Djamena Mother and Child University Teaching Hospital (CHU-ME).

2. Patients and Methods

2.1. Patients and Study Period

This was a one-year retrospective and descriptive study, from January 2021 to December 2021. It took place in the pediatrics department of the N'Djamena Mother and Child University Teaching Hospital (CHU-ME). Included in this study were all children in whom abnormal involuntary movements were observed, associated with psychic disorders and/or contractions of the muscles of

the trunk and extremities and whose age was less than or equal to 15 years. Incomplete records are not included in the survey.

2.2. Method

It is a retrospective study of all the records of the children over a period of one year, 2021. The diagnosis of the Korean was retained based on the criteria of Jones, modified in 1965 and revised in 1992 [8] [9] [10] (**Table 1**). The following variables were identified: socio-demographic characteristics, clinical and paraclinical data, therapeutic and evolutionary aspects.

3. Results and Comments

3.1. Results

During the study period, 2367 files were analyzed, of which 5 patients were diagnosed with Sydenham's chorea. However, among the 5 files, 2 were excluded from the study for non-compliance because they were incomplete and not usable. The hospital prevalence of this study is 0.12%.

3.2. Comments

3.2.1. Observation No. 1

The child AM aged 07, female, 3rd of a sibling of 05 children. AM was the result of a sustained pregnancy, delivery at term without any complications. AM was correctly vaccinated according to the extended national vaccination program (EPI) recommended by the WHO [11]. His psychomotor development was normal. She had an undocumented history of angina, and was admitted for physical asthenia, character disorders such as abnormal movements and progressive language disorders. The clinical examination objectified a poor general condition, with a stable hemodynamic state (blood pressure, pulse, heart rate), normal consciousness. The neurological examination objectified a drunken gait, ample abnormal movements at the level of the roots of the limbs, dysarthria, and coordination disorders such as dysmetria, asynergy, and adiadochokinesia. Tendon reflexes were present and symmetrical, and muscle hypotonia. The inflammatory biological assessment (NFS, ESR and CRP) was normal. ECG and Doppler

Table 1. Revised Jones criteria [8] [9] [10].

Major events	Minor manifestations	Evidence of strep infection
Cardites	Fièvre Fever	High and increasing level of anti-treptococcal antibodies (Aslo, Dnase B)
Polyarthritis	Arthralgia	
Chorea	Elongation of space PR on electrocardiogram	Positive pharyngeal cultures or identification by rapid strep test or recent scarlet fever
Erythema marginatum		
Subcutaneous nodules	Non-specific inflammatory signs (raised ESR, CRP, gammaglobilins)	

echocardiography were normal. The diagnosis of post-streptococcal chorea was retained. The child was put on a neuroleptic (Haloperidol) starting at 0.5 mg to gradually reach 1.5 mg/day for 6 months. The evolution was marked by a clear improvement under treatment, made of the regression of abnormal movements and gait disorders, with a follow-up of 6 months.

3.2.2. Observation No. 2

It was YA, 8 years old, female from a consanguineous marriage. YA was the result of a sustained pregnancy, delivery at term without any complications. She was properly vaccinated [11]. His psychomotor development was normal. She had in her history migrating poly-arthralgia and treated dental caries. YA was admitted for abnormal movements of the type of hyperactivity and behavioral disorders, progressive insomnia for three months. On clinical examination, she had an altered general condition, consciousness was normal, with an unstable hemodynamic state (tachycardia, galloping noises and systolic murmurs). The neurological examination objectified sudden, brief, and rapid involuntary choreic movements, predominant in the upper limbs. The inflammatory biological assessment was normal (NFS, VS and CRP). Doppler echocardiography revealed rheumatic valvulopathies. The diagnosis of post-streptococcal chorea was retained. The child was kept under neuroleptic (halopéridol) at increasing dose (2 mg/day) and corticosteroid therapy (2 mg/kg/day) for 10 days, as well as antibiotic prophylaxis (Pénidelay/month). The evolution was marked by the disappearance of choreic movements and persistence of object manipulation disorder with a follow-up of 6 months.

3.2.3. Observation No. 3

She was FM, a small child of 07 years old, attending primary school 2, 3rd of a sibling of five children. She is the result of consanguineous marriage (first degree consanguinity) and a sustained pregnancy, delivery at term without any complications, her psychomotor development was normal; she had had no vaccination contact. She had a history of recurrent angina. She was seen in an outpatient setting, then hospitalized for walking and language disorders that had been evolving for two months in an apyretic context.

The clinical examination revealed an altered general state, consciousness preserved, with an unstable hemodynamic state (tachycardia, galloping noises and systolic murmurs), physical asthenia. The neurological examination objectified sudden involuntary movements, predominant in the upper and lower limbs with slight hypotonia. The osteotendinous reflexes were present and symmetrical. The inflammatory biological assessment was normal (NFS, VS and CRP). Doppler echocardiography revealed rheumatic valvulopathies. The diagnosis of post streptococcal chorea was retained. The child was placed under neuroleptic (haloperidol) at a gradually increasing dose (2 mg/day) and corticosteroid therapy (2 mg/kg/day) for 10 days, as well as antibiotic prophylaxis (peni-delay/month). The evolution was marked by the initial improvement under treatment with a follow-up of 5 months.

4. Discussion

Sydenham's chorea (SC), also called "Saint Guy's Dance", is one of the neurological manifestations of rheumatoid arthritis (RAA) and is one of JONES' major criteria [8] [9] [10]. This rheumatism, which is being eradicated in industrialized countries due to the use of recommendations and the improvement in the quality of life, is still frequent in Third World Countries. The pathophysiology of this chorea remains poorly elucidated but the most accepted hypothesis is that of a post-infectious cross-autoimmune reaction by molecular mimicry between the epitopes of the membrane of the streptococcus and the isoganglioside GM1 of the neurons of the gray nuclei of the base [11] [12] [13]. In this study, the frequency of post-streptococcal chorea was 0.12%. This annual frequency is lower than those of several studies, including Kiliç [4] in Turkey, Panamonta [13] in Thailand and Tumas [14] in Brazil, *i.e.* 4.4 respectively; 4.1 and 8.12. According to Cardoso, the incidence of Sydenham's chorea in the United States and in southern countries in general is 0.2 to 0.8 per 100.000 inhabitants per year [15]. This study, which is the first of its kind in our country, also the low incidence can probably be due to the delay in the consultation of children in pediatric services, because the clinical signs are misinterpreted by parents who consider them as clumsiness and therefore do not entrust them to the doctor. But also to the difficulties of diagnosis based on a bundle of epidemiological, clinical and biological arguments coupled with the normality of cerebral imaging which requires a certain financial means. As a background, our study reveals repetitive angina in the first two little girls. Carapetis [16] reports in his study a high incidence of AAR and chorea in families of children with Sydenham's chorea. Peña found a family history of AAR in only one patient [3]. Other authors among others [17]-[22] have found that some patients have an insidious and progressive onset of the disease. On the neuromuscular level, the upper limbs and the face were solicited by involuntary movements, hypotonia. For several authors [2] [17] [21] [23] [24] involuntary movements are at the forefront of the disease. The predominance of hypotonia has been demonstrated by several studies [25] [26] [27]. In our case, the neuropsychic disorders were highlighted in a single child, essentially in the type of hyperactivity, incessant cries (Observation 1).

On the paraclinical level, the blood count does not give characteristic signs as in the AAR: the anemia is inconstant and very discreet. None of our patients had an elevated level of antistreptococcal antibodies (ASLO). As the time between the causative infection and the onset of symptoms is often prolonged, the level of antistreptococcal antibodies (ASLO) may be normal [17]. This explains why the relationship with a streptococcal infection is sometimes difficult to establish [25]. Two of our patients had rheumatic valve disease on Doppler echocardiography. Report described by several authors including [17] [21]. The three patients benefited from injectable penicillin G for 10 days, then relayed by intramuscular injection of Benzathine penicillin. It is recommended in all cases, even if formal proof of a rheumatic etiology is not provided [1] [3]. The majority of

authors are formal on the use of the prophylaxy [1] [2] because streptococcal reinfestation remains the major provider in triggering relapses of Sydenham's chorea. Nevertheless, a few reports mention that prophylactic treatment has no effect on recurrences or clinical course [3].

All our patients received haloperidol, at a gradually increasing dose to reach a daily dose of 1.5 mg per day. It gives good results, as has been observed in our patients and in various series in the literature [3] [11]. Two of three children with associated rheumatic carditis received corticosteroid therapy for 8 weeks. Corticosteroid therapy, whose rapid effect on chorea was highlighted by Mehta *et al.* [6], seems to be reserved for severe and febrile forms [26].

Improvement in choreic movements is observed during the first 2 weeks, which is consistent with the results of several studies [6] [27].

The evolution was favorable with a regression of choreic movements after 6 months of treatment with haloperidol. Vialet *et al.* [23] and Gilbert [26], noted a favorable evolution respectively, over a period of 1 to 8 years. We can conclude that the immediate evolution of Sydenham's chorea is favourable, but one cannot pronounce on a definitive cure, given the uncertain evolution which can be enamelled by the possibility of a cardiac attack or one or more relapses.

5. Conclusion

Sydenham's chorea is an affection which represents the main cause of acute chorea in children in Chad. The common clinical picture of this condition is characterized by the finding of abnormal involuntary movements. The diagnosis is mainly clinical. The therapeutic protocol is not codified, particularly the symptomatic treatment.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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