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CASE REPORT

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Sydenham's chorea in a Nigerian girl: A case report

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Abstract: Sydenham chorea is a major manifestation of Rheumatic fever (RF). Chorea occurs primarily in children and is rare after the age of 20 years. Females are more often affected and it almost never occurs in post-pubertal males. Sydenham's chorea is characterized by emotional lability, uncoordinated and erratic movements, and muscular weakness. The choreiform movements can be a source of irritation or a severe functional disability to the individual requiring appropriate management. The rarity of documentation of SC in sub-Saharan Africa raises the possibility of missed or misdiagnosis. We present the case of a 13 year old girl whose SC was initially misdiagnosed and conclude that though uncommon in Sub-Saharan Africa, SC requires proper diagnosis and prompt medical attention.

Introduction

Chorea has been defined as a state of excessive, spontaneous movements, irregularly timed, non-repetitive, randomly distributed and abrupt in character. These movements may vary in severity from restlessness with mild intermittent exaggeration of gesture and expression, fidgeting movements of the hands, unstable dance-like gait to a continuous flow of disabling, violent movements[1]. Sydenham's chorea was first described by Thomas Sydenham in 1684 as a clinical syndrome[2] and its link to rheumatic fever was proposed by Stoll in 1780[2]. Since then, Chorea has been recognized as a major manifestation of acute Rheumatic Fever (ARF) and may be the only evidence in approximately 20% of cases[2]. Chorea occurs primarily in female children. The prevalence of chorea in patients with RF varies from 5–36% in different reports[3]. The incidence in Africa is not precisely known but one pooled study has put it at 8.8%[4].
Incidence of ARF is decreasing in almost all the WHO Regions of the world[4], and as such Sydenham's chorea is also decreasing[4]. Currently, younger doctors may have never seen a patient with rheumatic chorea and have become unfamiliar with the disease. We report the case of a 13-year-old girl, who presented in our clinic with acute onset of abnormal involuntary movements and whose rheumatic chorea was initially misdiagnosed.

Case report

OA is a 13 year old girl who presented at the Paediatrics clinic of Reddington Hospital, Lagos, Nigeria with a three day history of rhythmic and involuntary jerking of the hands and five months history of joint pains.
She was well until five months previously when she developed severe pains on her right knee which was managed with topical analgesics and bandaging. Three weeks later she experienced similar pains in her left knee and then the left elbow. There was no history of joint swellings.
Three days before presentation, she developed twitching and involuntary jerking of her upper limbs and face. The jerky movements worsened whenever she became agitated and tried to stop them but disappears while asleep. The abnormal movements made her very anxious and self-conscious.
She was taken to a hospital where a diagnosis of upper limb dystonia was made and was commenced on Artane (Trihexyphenidyl). She was brought to our facility the next day because the jerking persisted.

General examination was normal except that she looked anxious with intermittent, involuntary jerks of both upper limbs and facial twitches. The twitches were subtle and the involuntary jerks consisted of fidgeting movements of the upper limbs which occurred every few minutes and were of very low amplitude, though she reported occasional involuntary flinging of her arms. She was lucid and had age-appropriate mentality. Both hands were not able to sustain a strong grip (milkmaid hand). Her knees revealed no signs of inflammation. Both knees demonstrated crepitus. Her respiratory and cardiovascular systems were normal on physical examination.
Full blood count done at presentation showed slight neutrophilia(78%) and elevated erythrocyte sedimentation rate of 43mm/hr (Reference range 0-10mm/hr). The throat swab culture yielded no growth but anti-streptolysin O-titre (ASOT) was elevated at 420IU (Reference range <200 I.U). Electrocardiogram, echocardiogram and electroencephalogram showed normal findings. Antineuronal Antibody (ANA) titre was positive but DNAse B titre was negative.
A diagnosis of Sydenham's chorea (SC) was made. She was commenced on Sodium Valproate, Ibuprofen and Penicillin V. These medications were given respectively to control the involuntary movements, reduce pain and

inflammation of the joints and eliminate any streptococcal organism remaining in her system. The patient's condition improved within 48 hours. By the second week, the involuntary movements had disappeared. The patient has resumed normal activities and has been followed up in our clinics for about 8 months. No recurrence was reported.

Discussion

Reliable data on the incidence of SC and RF are scarce. The incidence of SC in Africa is not known but one pooled study has put it at 8.8%[4]. No known data exists for SC in Nigeria but Okoroma et al[5] in Enugu reported it as being rare in their study. The diagnosis of SC depends on a high index of suspicion. Depending on the experience of the health provider the diagnosis may be entirely missed due to lack of awareness of the clinical entity. Our patient presented early to a peripheral hospital where the diagnosis was missed delaying management of her rheumatic chorea.

Considering the high contribution of RHD to acquired heart disease in Sub-Saharan Africa[6,7], the low incidence of SC appears paradoxical and the explanation is not clear. Studies of ARF outbreak in the USA during the 1990's attributed SC to highly virulent mucoid strains of group A Streptococci[8]. Though, it may be attractive to speculate that the strains of group A Streptoccoci in our environment are less virulent and therefore have lower propensity for causing SC, further studies need to be done on the virulence and immunogenicity of the strains of group A Streptoccoci in our environment that cause ARF to establish that.

According to Jones criteria[4] chorea alone is sufficient for diagnosis of RF[4] provided other causes of chorea had been excluded. Other possible causes of childhood chorea include cerebrovascular accidents, collagen vascular diseases, drug intoxication, hyperthyroidism, Wilson's disease, Huntington's disease, abetalipoproteinemia, Lesch-Nyhan syndrome and hormonal imbalances[2]. Unlike SC, Huntington chorea is generally a disease of adult onset with an unremitting autosomal dominant movement disorder and dementia; unlike in Huntington disease, neuroimaging in Sydenham's chorea is normal and other family members are usually unaffected. Our patient manifested the main features of SC as listed by WHO[2] viz: involuntary movements, hypotonia, and muscular weakness. Chorea can be generalized or unilateral[2]. Patients with SC may also have psychiatric symptoms such as depression, anxiety, personality changes, etc[7] Whether the psychological manifestations are secondary to the movement disorder or an integral part of the disease is not clear. It is also common for some patients with SC to present with other manifestations of RF[2]. Our patient had chronic arthralgia but had no clinical evidence of carditis. This may not be unusual as carditis has been documented to occur in only about 40-50% of patients with ARF at the time of diagnosis[9]. Averagely, rheumatic chorea resolves spontaneously within 3-6 months but may last longer if untreated. With treatment our patient's symptoms resolved within two weeks. Fast resolution with treatment has also been reported by other researchers[4,6,7]. About 20% of patients may experience recurrences and relapses, usually within 2 years after the initial attack[7] and this underscores the need for adequate follow up.

Treatment of SC is typically limited to supportive care, palliative medications, to eliminate streptococci and treat movement disorders, and long-term follow up.

Conclusion

Sydenham's chorea is a disabling manifestation of rheumatic fever, and its rarity in sub-Saharan Africa may have contributed to cases being misdiagnosed or even missed. It requires proper diagnosis as prompt and appropriate medications can bring quick resolution to symptoms. There is need for continual awareness creation on manifestations of ARF and its proper managements.

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