SYDENHAM'S CHOREA IN PAEDIATRIC AGE, A CASE WITH ATYPICAL RESPONSE TO TREATMENT

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ABSTRACT

Sydenham's chorea is a neurological disease that appears in people with rheumatic fever, usually in childhood. It is a possible long term complication of a streptococcal infection. The disease is usually latent and shows up to 6 months after the acute infection, but it may occasionally be the symptom of rheumatic fever's presentation. Sydenham's chorea occurs more frequently in the female sex and most patients are at under 18 years of age. We report the case of a 13 year-old female patient, without significant pathological findings, who was brought to our observation for having presented involuntary movements of the head and arms associated with important difficulty in speech for about 4 days. After a complete neurologic and psychiatric evaluation, blood and instrumental examinations, we make diagnosis of Sydenham's chorea and established a specific farmacologic plan of therapy, at first with AED (anti-epileptic drugs), and then with Haloperidol with an initial benefit. During the follow up, due to the recurrence and the persistence of the symptoms, we decided for a switch of therapy, using Aripiprazole, with a very important and long-lasting amelioration, with practically complete extinction of involuntary movements.

Keywords: Sydenham's chorea; basal ganglia; movement's disorder; developmental age; treatments.

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INTRODUCTION

Sydenham's chorea (SC), also called "Saint Vitus Dance", is a vasculitis with probable autoimmune pathogenesis, hat involves the encephalic vessels and affects up to 10% of rheumatic fever cases. [1] Clinically "chorea" describes involuntary, irregular, purposeless movements which are rapid and jerky and flow from one part of the body to another. It is a disorder characterized by the sudden onset (sometimes even in a few hours) of rapid uncoordinated snap movements that occur mainly on the face, hands and feet (usually chorea involves all four limbs).[2] Often patients can show other symptoms, such as behavior change, dysarthria, gait disturbance, loss of fine and gross motor control, headache. The result of all this is deterioration of handwriting, slowed cognition, scholastic difficulties and in social relationships, with an important reduction in the quality of life [3].

Furthermore, patients affected by chorea can highlight fasciculations of the tongue or muscle twitch and the "sign of milking ", a spasm of the hand with increase and decrease of the tension, as in hand milking. This disease was first described by Sydenham in a book published in 1686, making this affection the first example of movement disorder reported in the scientific literature. The typical age of onset of SC is 5–15 years and females are more affected than males (M:F 1: 5). From an epidemiological point of view, SC is one more frequent disease in countries with a temperate climate. It affects usually in summer and autumn, after the peak of incidence of rheumatic fever in spring and at the beginning of summer. Chorea develops later than other manifestations of rheumatic fever, usually 4–8 weeks after a group A beta-hemolytic streptococcal (GABHS) pharyngitis. The most accredited pathogenetic hypothesis is based on a possible molecular mimicry between streptococcal antigens and neuronal tissue antigens (basal ganglia). Cross-reactive antibodies are specific for both streptococcal antigens, but also for neuronal antigens. Therefore, they also bind to the nervous tissue, thus triggering the inflammatory cascade and the tissue damage. An essential test, useful in all patients, is the echocardiogram, which often results free from abnormalities, but it cannot show alterations, particularly of the mitral valve. The echocardiogram represents the only instrumental examination valid for diagnosis; other tests are not helpful for diagnosis, as imaging of the brain (TC, MRI) is always normal and EEG is normal or may show minor and absolutely non-specific changes. Even the laboratory tests are not very helpful, because they can only show an increase in ASLOt, "Antistreptolysin O titer" which indicates a previous streptococcal infection, with normal or only slightly altered flogosis indexes. Therefore the diagnosis is often difficult and is based almost exclusively on clinical criteria. [4-5]The treatment of SC has as its fundamental objective the complete regression of the neurological symptomatology with the consequent rapid reintegration of the child in the social and scholastic context [6]. Currently it is based on the administration of antipsychotic drugs, such as Haloperidol (or Aripiprazole or Risperidon) and antiepileptic drugs such as Valproate, Carbamazepine and Phenobarbital. The studies in the literature show that the drugs of first choice for treatment are, today, represented by valproate and carbamazepine that have superior efficacy to antipsychotics. [7-10]

CASE REPORT

We report the case of a 13 year-old female patient, who was brought to our observation for having presented involuntary movements of the head and arms associated with important difficulty in speech for about 4 days. She described states of anxiety and sudden changes in mood. We evaluated recent, remote, physiological and

family history of the patient and performed objective examination and neurological evaluation. Our group of work analyzed blood samples and electroencephalographic and neuroradiological investigations (TC and MRI of the brain).In anamnesis was reported pharyngitis, about 3 weeks ago. In family history we found that the parents were not consanguineous and we reported a positive history of neuropsychiatric diseases in the paternal family: epilepsy (not better defined) and anxious - depressive syndrome. Our patient was born from the first pregnancy of four (2nd: miscarriage; 3rd: 8 years old male, no diseases, 4th: 16 months old female, no diseases). Normodecourse pregnancy not associated with alcohol intake or smoking habit. No reported trauma or intake of drugs or substances with potentially damaging effects on gestation. Pregnancy carried out at term with spontaneous delivery. Weight at birth: 3100 grams. Apgar score: 9/10. Good cardio respiratory adaptation to extra uterine life. She reported healthy nutrition but characterized by capriciousness, regular sleep-wake rhythm and allergy to amoxicillin. The patient appears to be vaccinated according to Italian law. Her psychomotor development was reported in the standard for timing and methods of acquisition. At the time of hospitalization, she showed an excellent academic performance but poor social integration in the group of schoolmates. In pathological anamnesis nothing relevant to report. At the physical examination we observed: skin and mucous membranes without anomalies, norm transmitted vesicular murmur, eupnoic breath. Non-painful abdomen on superficial and deep palpation, on all quadrants. Thinness. Mild bilateral exophthalmos. At the neurological examination, the patient appeared alert and reactive, oriented in spatio-temporal parameters. Isochoric, isocyclic, normoreactive to photostimulus pupils. No deficit of muscular strength. Negative Romberg. Babinski absent. Normal reflexes without apparent asymmetries. No sign of meningeal irritation. The patient showed involuntary choreoatetosic movements on the head and limbs, more expressed in upper limbs. These movements compromised the ability to maintain balance and interfere with the regular and autonomous walking and sometimes even with the maintenance of the upright bipodalic statics. Language was characterized by the presence of expressive difficulties, in particular evident dysarthria. There was an evident state of anxiety and emotional lability. The laboratory tests performed during the hospitalization gave the following results: ammonium and lactate in the standard; negative pharyngeal swab, normal blood count, electrolytes, transaminases, creatine kinase; chemical-physical examination of the urine without anomalies; inflammatory indices and thyroid function in the norm. However, an ASLOt (Antistreptolysin O titer) of 1830 IU/ml was highlighted (normal range: 0-200). Pedagogical and psychological counseling highlighted a marked deficit in attention and concentration. The echocardiogram showed mild mitral insufficiency with small clinical and hemodynamic relevance. Thyroid and abdominal ecography and electroencephalogram did not reveal substantial anomalies. Neuroimaging (TC and MRI of brain and) was absolutely normal. Based on the clinical evidence that in the absence of treatment showed a worsening of symptoms in a few hours, and after the results of laboratory and instrumental tests performed urgently, we have placed diagnosis of Sydenham's chorea and established a specific farmacologic plan of therapy. As the patient reported allergy to beta lactams, after consultation with the infectious and allergist specialists, an antibiotic therapy with macrolide (Azithromycin 200mg/die) was chosen. To act on the movement disorder, we started the treatment with an anti-epileptic drug, according to the latest evidence of the literature on their effectiveness in these conditions, using Valproate (20 mg/kg/die).

Due to the onset of side effects ascribable to Valproate (nausea, vomiting, electrolyte abnormalities), we decided to modify the therapy, replacing it with Haloperidol (0, 01 mg/kg/die).We obtained a good reduction in the symptoms that allowed us to discharge the patient from our department, organizing a monthly clinical follow-up for her. Unable to use a penicillin, antibiotic prophylaxis therapy was recommended with Azithromycin. During the first and second follow-up meetings, the patient showed the persistence of the choreic movements, which however appeared to be significantly reduced compared to the period of admission. ASLOt was down (1389 and then 886 IU/ml). Therefore the therapy was confirmed. However, the patient returned to our observation before the next scheduled check, complaining of the symptoms flare up, such as make impossible her autonomy in the main daily occupations. We observed these anomalies during our evaluation and therefore we opted for a change of therapy, administering Aripiprazole (5mg twice daily) and decommissioning Haloperidol. We have achieved considerable benefit within a few days, which has been maintained over time. Subsequent monthly clinical follow-up tests have shown complete resolution of the clinical symptomatology after about three months from the start of this therapy with a significant improvement in the quality of life and social relations.

CONCLUSIONS

Like the rheumatic disease, the incidence of Sydenham's chorea seems to be declining, even if outbreaks are reported, not only in developing countries [11-12]. Although Sydenham chorea is a rare pathology, extra pyramidal involvement may be considered in patients who show acute movement disorders, without pyramidal characteristics, more in those with pathological history of streptococcal diseases. The diagnosis is exclusively clinic. Sydenham's chorea typically does not appear at the same time as arthritis, but often it associates with the cardite. As in our case described, the early establishment of a correct pharmacotherapy allows complete regression of neurological symptomatology with the consequent rapid reintegration of the child in the social context and school. Prophylaxis with penicillin-benzatine reduces the risk of relapse. The studies in the literature show that the drugs of first choice for treatment are, today, represented by antiepileptic drugs that have superior efficacy to antipsychotics. Our case-report describes a situation in which Aripiprazole has been shown to be more effective than other drugs in the reduction of symptoms, so we suggest the possible use of this drug in the conditions in which there has been a failure of commonly used drugs.

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