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DIAGNOSTICS, DIFFERENTIAL DIAGNOSTICS AND THE TREATMENT OF PANDAS SYNDROME. CASE DESCRIPTION

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Abstract:

This article gives a case of PANDAS syndrome in an 11-year-old boy. The course and clinic of the disease in the form of prolonged sneezing attacks made it difficult to diagnose this syndrome. Conducting biochemical blood tests, daily video electroencephalographic monitoring (EEG monitoring) and magnetic resonance imaging of the brain (MRI) allowed to determine the condition. The appointment of antibiotic therapy and intravenous administration of immunoglobulin led to remission of the disease. It is necessary to conduct further studies to determine the effectiveness of the above drugs for this syndrome.

Keywords: Tics, PANDAS Syndrome, EEG, Disease, Children

The most common group of bacterial infections in humans today is streptococcal infections caused by beta-hemolytic streptococci of group A. Doctors of almost all specialties meet with this extensive group of infectious diseases. The pathogenesis of diseases is associated with the production of toxins such as: streptokinase A and B, hemolysin, streptolysin, deoxyribonuclease, hyaluronidase. Nosoforms are represented by superficial (erysipelas, impetigo, pharyngitis, sore throats) toxin-mediated (scarlet fever, toxic shock syndrome) and invasive (necrotizing fasciitis, myositis, meningitis, endocarditis, pneumonia, postpartum sepsis) [7].

The occurrence of neurological disorders in children, manifested by tics and obsessive-compulsive disorders (OCD) is also associated with streptococcal infection. Experts observe the appearance of tics, neurosis-like obsessive states, choreiform hyperkinesis, myoclonia in children associated with beta–hemolytic streptococci of group A. Such cases, at the suggestion of S.E.Swedo [6], are commonly referred to as PANDAS syndrome (pediatric autoimmune neuropsychiatric disorders associated with streptococcal infection - a pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection).

For 10 years, PANDAS syndrome was considered as related to rheumatic fever and especially Sydenham's chorea, which was confirmed by its association with infection caused by beta-hemolytic streptococci of group A and the relative effectiveness of anti-rheumatic therapy. The pathogenesis of this disease is based on an autoimmune reaction in which antibodies attack nerve cells [3]. The clinical and diagnostic symptom complex of PANDAS syndrome is similar to the symptoms of minor chorea. The development of PANDAS syndrome is typical for prepubescent children (unlike chorea, for which the typical age group is children 5-8 years old). The disease begins and proceeds quite acutely. The clinical symptom complex of obsessive-compulsive disorders is common to PANDAS syndrome and chorea. Typical manifestations of PANDAS syndrome include obsessions of a different nature and volitional uncontrolled efforts or actions. Such conditions occur with a certain frequency (the average duration of the attack is on average 12-15 weeks) and significantly reduces the quality of life of the patient. Clinical symptoms are combined with such manifestations as absent-mindedness, choreiform hyperkinesis, impulsivity, motor hyperactivity, emotional lability, attention disorders, difficulty falling asleep, Tourette syndrome, anorexia, which allows them to be regarded as comorbid processes with PANDAS syndrome [3].

Diagnostic criteria of PANDAS syndrome are [6]: - debut in childhood: symptoms appear between the age of 3 and puberty; -the presence of obsessive-compulsive disorders and/or tics; -paroxysmal type of the course of the disease, characterized by a sudden onset or a sharp increase in symptoms. Often, the appearance of symptoms or their exacerbation can be associated with a certain day or week. Symptoms usually decrease significantly, and sometimes completely disappear between episodes of exacerbation; -connection with neurological disorders. During an exacerbation, patients find changes in their neurological status. Hyperactivity and hyperkinesis (including choreiform) are most often observed -a connection with streptococcal infection - exacerbation should be associated with the detection of streptococcus in the nasopharynx and / or with an increase in the titer of antibodies to streptococcus.

Below is a description of my own observation of PANDAS syndrome.

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Patient A.X, 11 years old. At the age of 10, she complained of frequent sneezing, which occurs paroxysmally, lasting up to 8-9 hours, observed for 1.5-2.0 months. These episodes occurred only in a state of wakefulness throughout the day, were not observed at night. Anamnesis vitae: a child from the 1st pregnancy. Pregnancy and childbirth proceeded normally. Birth weight - 3000, height - 54 cm. Discharged from the hospital on time. Up to 10 years of age, the development corresponded to the age norm, he was ill with tonsillitis every year.

Heredity for neurological and mental diseases is not burdened.

Anamnesis morbi: at the end of July 2017, at the age of 10, after a fright (a stray dog attacked), hiccups appeared, continuing throughout the day, followed by disappearance. On day 3, sneezing attacks appeared (60 per minute), which also lasted for 1 day, followed by the disappearance of symptoms. In October 2017, the boy suffered tonsillitis, after which the subfebrile temperature (37.3 – 37.6 ° C) remained for another 1.5 months. 10 days after recovery, paroxysms reappeared in the form of sneezing (with a frequency of 70-90 di per minute). The duration of episodes in general reached 8-9 hours throughout the day. By the evening, sneezing usually intensified and disappeared when falling asleep.

The examination of the child began with daily electroencephalographic (EEG) monitoring, which revealed the presence of motor tics (during sneezing, tic twitching of the eyes was observed), and also excluded the epileptic nature of paroxysms.

Magnetic resonance imaging revealed cleavage of the transparent septum, which, apparently, has nothing to do with this condition and is a variant of the norm.

Streptococcus mitis (106 KOE/ml) and Staphylococcus epidermidis (106 KOE/ml) were detected during back-sowing from the nasopharynx; the number of bacteria is within normal limits. A biochemical blood test revealed a high rate of antistreptolysin-O (ASL-O) with normal indicators of C-reactive protein and rheumatoid factor.

** ASL-About 460 IU/ml (norm - up to 150 IU/ml);*

** C-reactive protein ≤ 1 mg/l (norm - up to 5 mg/l);*

** Rheumatoid factor (RF) ≤ 10 IU/ml (norm - up to 14 IU/ml)*

Taking into account the presence of vocal and motor tics in combination with an increased level of ASL-O, the exacerbation of the symptoms of the disease after tonsillitis made it possible to suspect PANDAS syndrome as the cause of this condition.

Differential diagnosis with partial epilepsy, Tourette's syndrome and Sydenham's chorea was performed. The literature describes cases of temporal lobe epilepsy with attacks in the form of sneezing [2], the daily EEG monitoring did not reveal either interictal or ictal (during sneezing) epileptiform changes. This is not an absolute exception (in some cases, a normal EEG does not exclude epilepsy), but it serves as a fairly strong argument against epilepsy. The theory of the non-epileptic origin of this condition was supported by the presence of motor tics.

Tourette's syndrome is a condition that can give a similar clinical picture (the presence of motor (eye twitching) and vocal (sneezing) tics). But in this case, the diagnostic criteria of Tourette's syndrome were not met, including a duration of more than 1 year. Such cases as the connection with tonsillitis, an increase in the level of ASL-O, the isolation of culture from the throat, make this condition more similar to PANDAS syndrome.

Sydenham's chorea was excluded due to the absence of proper choreiform hyperkinesis, polyarthritis and rheumocarditis based on normal indicators of C-reactive protein and rheumatoid factor.

A search for information on social networks revealed a description of similar cases in the Russian Federation and the USA:PANDAS syndrome was diagnosed and treated with intravenous immunoglobulin with a good effect [1]. Research data [1,5] determined the choice of human immunoglobulin as a treatment method.

Human immunoglobulin at a dose of 0.5 g / kg / day IV was prescribed to the patient, followed by monthly administration for 6 months.

The first administration of immunoglobulin reduced the frequency of sneezing paroxysms by 75%. Azimac was also prescribed at a dose of 500 mg/day for three weeks [4]. By the end of the 2nd week of treatment, the tics disappeared completely. Remission lasted about 7 months with the further appearance of tics and their preservation throughout the whole day. To date, the boy's condition has stabilized. After the appearance of tics and their subsequent disappearance, remission is 4 months. The ASL-O index decreased to 350 IU/ml. Drug therapy is not carried out.

The analysis of this clinical case demonstrates the existence of a group of patients with a good therapeutic response to the administration of immunoglobulins and antibiotics. The mechanisms of development and course of this condition are not fully understood and require further research.

LITERATURE:

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