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

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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 bouts of sneezing made it difficult to diagnose this syndrome. Biochemical blood tests, 24-hour video electroencephalographic monitoring (EEG monitoring) and magnetic resonance imaging of the brain (MRI) made it possible to determine the condition. Prescription of antibiotic therapy and intravenous administration of immunoglobulin led to remission of the disease. It is necessary to conduct further research to determine the effectiveness of the above drugs introduced above for this syndrome.

Keywords: tics, PANDAS syndrome, EEG, disease, children.

The most common group of human bacterial infections today is streptococcal infections caused by group A β-hemolytic streptococci. Physicians of almost all specialties encounter this broad 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 superficial (erysipelas, impetigo, pharyngitis, tonsillitis), 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, myoclonias in children associated with group A β-hemolytic streptoc. Such cases, as suggested by S.E. Swedo [6], are usually

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designated as PANDAS syndrome (pediatric autoimmune neuropsychiatric disorders associated with streptococcal infection).

For 10 years, PANDAS syndrome has been considered as related to rheumatic fever and especially Sydenham's chorea, one hundred confirmed by the association with infection caused by group A β-hemolytic streptococci and the relative effectiveness of antirheumatic therapy. The pathogenesis of this disease is an autoimmune reaction in which antibodies affect nerve cells [3].

The clinical and diagnostic symptom complex of the PANDAS syndrome is similar to the symptomatology of chorea minor. The development of PANDAS syndrome is typical for prepubertal children (in contrast to 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 disorder 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 an attack is on average 12-15 weeks) and significantly reduces the patient's quality of life. Clinical symptoms are combined with such manifestations as absent-mindedness, choreiform hyperkinesis, impulsivity, motor hyperactivity, emotional lability, attention disorders, difficulty falling asleep, Tourette's syndrome, anorexia, which allows us to regard them as processes comorbid with PANDAS syndrom

The diagnostic criteria for PANDAS syndrome are [6]:

- Onset in childhood: symptoms appear between 3 years of age and puberty;
-The presence of obsessive-compulsive disorders and / or tics;
- A seizure type of the course of the disease, characterized by a sudden onset or a sharp increase in symptoms. Often the onset or worsening of symptoms can be associated with a specific day or week. Symptoms are usually significantly reduced, and sometimes completely disappear between episodes of exacerbation;
- Connection with neurological disorders. During an exacerbation, changes in the neurological status are found in patients. The most common are hyperactivity and hyperkinesis (including choreiform)
- 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 our own observation of PANDAS syndrome.

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Patient A.Kh, 11 years old. At the age of 10, she complained of frequent sneezing, which occurs paroxysm, 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; they were not observed at night.

Anamnesis vitae: baby from 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 had tonsillitis every year.

Heredity for neurological and mental illness is not burdened.

Anamnesis morbi: at the end of July 2017, at the age of 10, after being frightened (a stray dog attacked), hiccups appeared that continued throughout the day and then disappeared. On day 3, sneezing attacks (60 per minute) appeared, which also lasted 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) lasted for another 1.5 months. 10 days after recovery, paroxysms reappeared in the form of sneezing (with a frequency of 70-90 per minute). The duration of the 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 (tic twitching of the eyes was observed during sneezing), and also excluded the epileptic nature of paroxysms.

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

When conducting bacterial culture from the nasopharynx, Streptococcus mitis (106 KOE / ml) and Staphylococcus epidermidis (106 KOE / ml) were found; the number of bacteria is within normal limits.

A biochemical blood test revealed a high level of antistreptolysin-O (ASL-O) with normal values of C-reactive protein and rheumatoid factor.

- ASL-O 460 IU / ml (the norm is up to 150 IU / ml);
- C-reactive protein ≤ 1 mg / l (the norm is up to 5 mg / l);
- Rheumatoid factor (RF) ≤ 10 IU / ml (the norm is up to 14 IU / ml)

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Considering the presence of vocal and motor tics in combination with an increased level of ASL-O, the exacerbation of symptoms of the disease after suffering 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 carried out. The literature describes cases of temporal lobe epilepsy with seizures in the form of sneezing [2]; the daily EEG monitoring did not reveal either interictal or ictal (during sneezing) epileptiform change. 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 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 (twitching of the eyes) and vocal (sneezing) tics). But in this case, the diagnostic criteria for Tourette's syndrome were not met, including the duration of more than 1 year. Cases such as a connection with tonsillitis, an increase in ASL-O levels, and a culture from the pharynx make this condition more similar to PANDAS syndrome.

Sydenham's chorea was excluded due to the absence of choreiform hyperkinesis, polyarthritis and rheumatic heart disease on the basis of normal C-reactive protein and rheumatoid factor values.

A search for information in social networks revealed a description of similar cases in the Russian Federation and the United States: PANDAS syndrome was diagnosed, treatment with intravenous immunoglobulin was carried out 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 injection of immunoglobulin reduced the frequency of sneezing paroxysms by 75%. Azimak 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 had disappeared completely. The remission lasted about 7 months with the further appearance of tics and their persistence throughout the day. To date, the boy's condition has stabilized. After the appearance of tics and their subsequent disappearance,

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remission is 4 months. The ASL-O index dropped to 350 IU / ml. Drug therapy is not performed.

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.

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