Rare disease

Bilateral ballism following streptococcal infection, associated with psychiatric disorder and purpura

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A woman in her 30s was brought to the hospital with abnormal movements. Three months prior, the patient had exacerbation of the movements after an episode of recurrent pharyngitis. Neurological examination revealed, violent involuntary movement that affected both upper and lower limbs, hypotonia and ataxia. Other findings including emotional instability and involuntary movements were considered ballistic. Throat culture showed β haemolytic streptococci, tonsillectomy and specific antibiotic improved bilateral ballism and psychiatric disorder. This is the first report of bilateral ballism poststreptococcal infection.

BACKGROUND

Bilateral ballism (biballism) is very rare, 23 cases were reported, and only two of these were in children.¹ It has been attributed to a variety of aetiologies, or it has occurred upon other underlying diseases such as: multiple sclerosis² or Guillain-Barré syndrome.³ Our case is important because it provides a new aetiology to the involuntary movements of the extrapyramidal syndromes. The study of this case can be viewed as reference for the clinical awareness of the physician. We wrote this article with the intention that it may be useful for clinical research in the etio-pathogenesis of other diseases of the central nervous system (CNS).

CASE PRESENTATION

A white female in her 30s with a history of recurrent episodes of febrile pharyngitis since childhood was treated with penicillin. Three years prior to admission, following an episode of pharyngitis, the patient was observed walking with large steps, throwing off her shoes, for a period of over 1 week. This movement progressed in the following weeks to involve the trunk and neck, with protrusion of the tongue, and grimacing affecting speech. She was seen by a psychiatrist who diagnosed organic depression, prescribing antidepressant without positive results. At admission, the patient had several symptoms of epistaxis, pain and swelling of the knees, and complained of hypermenorrhea. Neurological examination showed: vigorous, continuous and high-amplitude movements, affecting the proximal muscles of the four limbs, particularly the kicking movements of the legs. Dorsal flexion of the neck, torsion of the trunk and rhythmic rotation of the pelvis in standing position. The movements intensified by voluntary action or excitement and subsided at rest. Consciousness was clear. Dysarthria, explosive words, cerebellar ataxia, hypotonia and areflexia were observed, with occasional pendular reflex. Plantar reflexes were flexor, and ballistic movements were associated with choreoathetoid movements. In addition, spontaneous ecchymoses was

observed. No sociopsychological factors were found that might be related to her disease.

INVESTIGATIONS

Two throat cultures during episodes of pharyngitis showed β haemolytic streptococci with elevated antistreptolysin O titres. Two studies of coagulation were normal. Serum copper and ceruloplasmin were normal. Electroencephalogram, cerebrospinal fluid and neuroimaging were normal.

DIFFERENTIAL DIAGNOSIS

Bilateral ballism was mainly differentiated from chorea, in which involuntary movements are repetitive, resembling fragments of voluntary action, affecting the distal muscles and are less visible in the lower limbs. Hepatolenticular degeneration (Wilson's disease) was excluded by negative family history, absence of liver damage and Kayser-Fleischer rings. Furthermore, serum copper and ceruloplasmin were normal in our patient.

TREATMENT

Tonsillectomy was followed by penicillin 2 million units intravenously every 6 h for 1 week.

OUTCOME AND FOLLOW-UP

The outcome was initially favourable. The causal therapy led to decrease of involuntary movements, improvement in depression, previously ineffective to antidepressant drugs and abolition of the purpuric symptoms, including the hypermenorrhea without hormonal treatment. Thereafter, the patient left medical care. Two years later, the patient had a relapse of depression. At that time, the patient suffered a fall resulting in a fracture of the humerus which was complicated by nosocomial pneumonia, and died.

DISCUSSION

In our case, the clinical diagnosis of bilateral ballism was made obvious by the typical involuntary movements, the

mode of onset by the lower limbs as kicking movements, and the presence of torsion of the trunk. Cerebellar ataxia and pendular reflex are indicative of cerebellar dysfunction. The aetiology of bilateral ballism most frequently reported is vascular⁴ such as bilateral haemorrhagic infarction in caudate nucleus and putamen.⁵ In our case, the streptococcal infection enlarged the aetiological varieties of this rare extrapyramidal syndrome. The above mentioned improvement after treatment, especially antibiotic, reaffirmed the aetiology. Prompt antibiotic treatment of pharyngeal infection lead to an improvement in paediatric autoimmune neuropsychiatric disorders associated with streptococcal infections neuropsychiatric symptoms,⁶ as it occurred in other dyskinesias.⁷ In our case, the psychiatric disorder accompanied biballism. We believed our patient's depression relapsed as a result of ceasing medical care, specifically the use of antibiotics. In bilateral ballism previously reported, the most common anatomical lesion was extensive damage to both basal ganglia, including subthalamic nucleus.⁴ Based on the clinical course and the neurological signs in our case, a diffuse lesion in the subthalamic nucleus involving the globus pallidus was identified, with which it is reciprocally linked. Neuroimaging was normal. The MRI is commonly normal in poststreptococcal disorders of the CNS.⁷ Likewise, it was normal in bilateral ballism induced by hypoglycemia in a diabetic patient⁸ by thyrotoxicosis⁹ fever induced¹⁰ or by drugs.^{11 12} All of the above mentioned, were reverted by causal treatment. The natural history and clinical features found in our patient were similar to rheumatic fever (RF), except for the absence of carditis in our case. It is possible that our patient was protected from RF by the treatment of recurrent pharyngitis with penicillin. This observation suggests a possible role of immunopathogenesis in our case. However, the disease's tendency may be multifactorial including immune and other factors.¹³ If our patient had been diagnosed earlier, she would have had a better outcome. Some patients can develop a permanent lesion of the basal nuclei probably due to chronicity of the process for which a cause is still unclear.¹⁴

In many diseases considered idiopathic, congenital or degenerative, as it occurred in our case, the true aetiology was found. The interrelation of multiple syndromes led us to the unique cause of the disease. It is our hope that this study will be useful to physicians in the clinical practice.

Learning points

- ▶ Those patients who suffer from an acute infection disease, such as febrile pharyngitis in children should be properly identified and adequately treated with prophylaxis against later disease.
- ▶ An early and accurate diagnosis often leads to a more favourable outcome.
- ▶ Consistent clinical observation may be the beginning of a promising research.

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Competing interests None.

Patient consent Not obtained.

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