Ischemic Stroke: an Unusual Presentation of Acute Rheumatic Fever

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Case Report

A Caucasian 8-year old boy was referred to our emergency room for recent-onset weakness on the right side of the body. During the previous months he had two febrile episodes lasting four days each, accompanied by arthralgia of elbows, knees and wrists, with no signs of inflammation. Family history was unremarkable. Physical examination showed mucocutaneous pallor, a few erythematous round cutaneous lesions on the buttocks and the inner thighs, and a 2/6 systolic murmur on the apex of the heart. He also appeared to be very asthenic and complained of migratory arthralgia on the lower limbs, without joint effusion. Blood tests showed high levels of inflammatory markers (Erythrocyte sedimentation rate (ESR): 68 mm/h; C-reactive protein (CRP): 5.73 mg/dl) and a mild anaemia, with haemoglobin (Hb) at 10.9 g/dl.

Abstract

Rheumatic fever is a systemic disease that affects children with a previous Group A beta-haemolytic streptococcus infection. Clinical manifestations are variable: joints, heart, skin or central nervous system can be involved.

We report a case of a Caucasian 8 year old boy with rheumatic fever, who presented with a stroke as first sign of the disease. Stroke is not a typical manifestation of rheumatic fever and was caused by a cardioembolic event secondary to the carditis.

Key words: Fever, rheumatic, ischemic stroke, paediatric, angio-magnetic resonance
Due to the right sided weakness, MRI with contrast and angio-MRI of the brain were performed, and two hyperintense lesions in the frontal lobe, on the right and left semi-oval centres, were detected, consistent with an ischemic embolic stroke (Figure 1).

Color Doppler echocardiography showed atrial and ventricular enlargement with an abnormal wall motion of basal septum, a severe mitral valve insufficiency and a moderate aortic valve insufficiency. Laboratory tests showed evidence of previous streptococcal infection (elevated Anti-streptolysin titre: 1800 U/I/ml, and anti-DNase B: 2200 U/I/ml, normal values < 200 U/I/ml for both. The diagnosis of acute rheumatic fever was made, and treatment with prednisone (2 mg/kg/day), furosemide (25 mg/day), captopril (6.5 mg 3 times a day) and aspirin (100 mg/day) was started, together with penicillin prophylaxis (1.200.000 U i.m. every 21 days).

The clinical course improved rapidly, neurological symptoms subsided in a few hours, and the patient’s general conditions improved during the hospitalization. Also, the diastolic murmur on the aortic area could not be heard anymore after 3 days of therapy, and the systolic murmur was attenuated. At the follow up after 6 weeks, echocardiography showed a persisting but mild mitral and aortic regurgitation.

**Discussion**

Rheumatic fever is a systemic disease that occurs as a complication of Group A beta-haemolytic streptococcus infection. It mainly affects children between 5 and 14 years of age, and is very rare in developed countries but still a common health problem in developing nations. Classic diagnostic criteria include involvement of skin, joints, heart, and central nervous system. The disease can present with fever and a variable combination of carditis, arthritis, skin manifestations and chorea [1-5].

Sydenham’s chorea is a neurological and psychiatric disorder, characterized by muscle weakness, balance disturbance, involuntary movements, irritability, age-regressed behaviour and obsessive-compulsive disorders. It can be a delayed manifestation of rheumatic fever or a unique one. There are no laboratory or imaging tests to confirm the diagnosis [3]. The neurological symptoms of our patient are not typical manifestations of rheumatic fever, and cannot be considered as a part of Sydenham’s chorea. Instead, they can be classified as a thromboembolic complication secondary to heart disease. Thus, the patient was referred to our hospital because of an indirect sign of rheumatic fever, which was diagnosed upon full physical examination and echocardiography, and after confirmation of a previous Streptococcal infection.

Neuroradiological investigations revealed the ischemic lesions that were causing the boy’s signs and symptoms and were secondary to an embolism from one of the damaged cardiac valves. Although there is extensive literature of strokes caused by cardiac thromboembolism [6,7] to our knowledge there are no reports of acute rheumatic fever with such a presentation. Luckily, follow-up was very favourable, with complete disappearance of all signs and symptoms, but with the persistence of a both mild mitral and aortic insufficiency.

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References