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Case Report

Cardiovascular accident unveiling rheumatic heart disease

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ABSTRACT

Infection with group A *Streptococci* causes an autoimmune reaction known as acute rheumatic fever (ARF) which leads to chronic cardiac condition called rheumatic heart disease (RHD). Rheumatic involvement of the cardiac valves is one of the sequelae of ARF. The valvular lesions begin as verrucae along the borders of heart valves and tend to resolve leaving a scar once the inflammation subsides. Cardio embolism is seen as the main pathophysiological mechanism of ischemic stroke in RHD.

Keywords: Autoimmune, Rheumatic heart disease, Valvular lesions, Stagnant blood flow, Thrombus, Cardioembolism

INTRODUCTION

Rheumatic heart disease (RHD) is a common consequence of acute rheumatic fever (ARF) in developing countries.[1] Cardio embolism is seen as the main pathophysiological mechanism of ischemic stroke in RHD, mainly associated with mitral valve lesion, in which left atrial (LA) enlargement predisposes stagnant blood flow, thrombus formation and consequently, embolic events. Wolf et al reported that the rate of stroke in patients with RHD and AF was nearly 18-fold higher than an age, sex, and hypertension matched population without AF (based on just 7 events among 154 patients).[2] In this case report, we present a 17-year-old girl who presented with quadriparesis (left more than right) diagnosed later with RHD. We would like to bring into light, cardiovascular accident (CVA) being an initial presentation of RHD.

CASE REPORT

17-year-old female child brought with complaints of difficulty in performing daily activities due to weakness of all four limbs for 20 days and inability to speak since 15 days. No significant relevant history. On admission child was vitally stable. Central nervous system (CNS) examination revealed motor aphasia, right sided upper motor neuron (UMN) type of facial palsy, all four-limb weakness with left sided weakness more than right, hyperreflexia in all four limbs, bilateral babinski positive, cardiovascular system (CVS) examination revealed pansystolic murmur grade III on mitral area. Routine investigations done hemoglobin (Hb) 9.3 g%, hematocrit (HCT) 27.6, white blood cells (WBC) 9730/mm^3, platelet count (PLC) 2,04,000 /mm^3, C-reactive protein (CRP) 27.61 mg/l, coagulation profile prothrombin time (PT)/activated partial thromboplastin time (aPTT)/international normalized ratio (INR) 11.2/36.8/0.85, erythrocyte sedimentation rate (ESR) 26 mm/hour, antistreptolysin O (ASO) <75, on day 2 of admission 2D ECHO done suggestive of RHD, severe MR, mild TR, pulmonary hypertension, dilated LA and LV, aortic valve normal, normal biventricular function, electrocardiography within normal limits (ECG WNL). As per cardiologist advise, started on aspirin in antiplatelet dose with angiotensin converting enzyme (ACE) inhibitors. On day 3 of admission, magnetic resonance imaging (MRI) brain done suggestive of subacute infarcts in bilateral cerebral hemispheres likely Vasculitis. However, MR Angiogram Brain done showed normal study. Antinuclear antibody (ANA) profile sent was

negative. As per neurologist advise dual antiplatelet therapy started and diagnosed as chronic RHD with CVA secondary to cardio embolism and discharged on day 7 of admission. Speech being normalized with improvement in motor component. Child is planned to be operated for severe MR with mitral valvuloplasty.

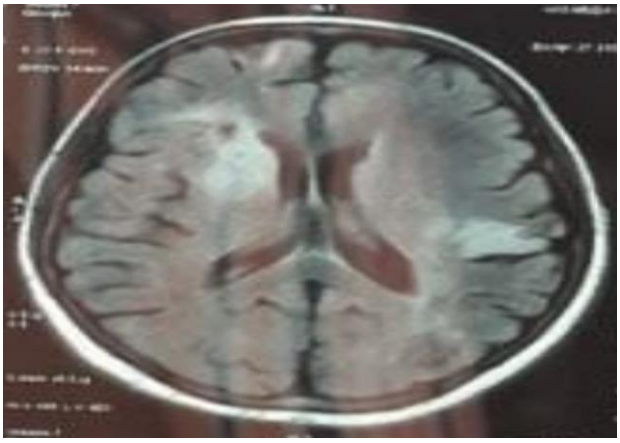

Figure 1: MRI brain T2 sequence showing subacute infarcts in bilateral cerebral hemispheres.

DISCUSSION

Worldwide, rheumatic heart disease remains the most common form of acquired heart disease in all age-groups, accounting for up to 50% of all cardiovascular disease and 50% of all cardiac admissions in many developing countries. The global, regional, and national burden of RHD from 1990 to 2015, as part of the 2015 global burden of disease study, was reported in a 2017 publication. The study found persistence of high rates of RHD in poor regions of the world where RHD remains endemic.[3]

RHD typically affects left-sided valves, with greater affinity and consequence for the mitral valve. Characteristic acute mitral valvulitis shows mitral annulus dilatation, chordal elongation, and anterior leaflet prolapse, with varying degrees of MR and rarely chordal rupture. Isolated aortic disease occurs in 2% of cases.[4] Right-sided valve disease is not infrequent, typically affects the tricuspid valve (as primary valvulitis or as the result of deleterious hemodynamic consequences of left-sided valve disease), and rarely affects the pulmonic valve. Acute rheumatic valvulitis manifests as valvular regurgitation, but over time, chronic inflammation leads to valve stenosis from commissural fusion with or without associated regurgitation in a subset of patients. MS from commissural fusion, with variable degrees of involvement of other parts of the mitral valve apparatus, is the hallmark lesion of the later stages of RHD.[5]

In RHD, emboli formation could happen as a consequence of the calcification process that is undergone by the rheumatic valves. When released, it might flow to the brain resulting in a cardioembolic stroke, which is responsible for cerebral infarction. RHD usually affects mitral and aortic valves.

Our case presented a rare manifestation in which the tricuspid valves were affected, resulting in a trivial tricuspid regurgitation. Also in our case, the child initially presented with features of upper motor neuron lesion, possibly cerebrovascular accident. Cardiac cause was suspected due to clinically evident murmur. 2D ECHO was suggestive of RHD. Also considering the age and sex as a risk factor for autoimmune disease, Angiogram and ANA profile were done and ruled out vasculitis. Though supportive imaging could not be established with 2D ECHO, the child presenting days later after the symptoms, support the possibilities of microthrombi and embolism. Complete motor improvement following dual antiplatelet therapy, supports the cardiac aetiology.

A relevant case report on hemorrhagic transformation of embolic stroke in pediatric rheumatic heart disease says risk of stroke is 18 times higher in cases with RHD. Emboli occurs as a possible consequence of calcification in the rheumatic valves.[6]

CONCLUSION

Through this case we would like to bring to light that it is a rare case of chronic RHD which initially presented with CNS complications of embolism. Cardio embolism is seen as the pathophysiological mechanism of ischemic stroke in RHD, mainly associated with mitral valve lesion.

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