

Complications of Rheumatic Heart Disease: Ischemic Stroke and Infective Endocarditis

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Abstract

The incidence of rheumatic heart disease has been steadily declining throughout the developed world, with the incidence in the United States and other developed now < 2 cases per 100,000 school-aged children [1]. Despite this, worldwide rheumatic heart disease and its complications result in about 233,000 deaths annually with the majority of cases occurring in low-income countries [2]. We described a 15-year old girl with significant history of rheumatic heart disease who presents with left sided weakness, slurred speech and an associated subjective fever.

Keywords: Rheumatic Heart Disease; Infective Endocarditis; Septic Embolic Stroke; hemorrhagic Stroke; Hemiparesis

Introduction

The prevalence of rheumatic heart disease and sequelae of the disease has decreased and is now rare in developed countries due to the appropriate and prompt treatment of Group A Streptococcal pharyngitis. Despite this, it still remains a major cause of cardiovascular disease in developing countries, affecting an estimated 12 million persons world-wide with two-thirds of those who are affected being children between the ages of five and fifteen [4].

Here, we present the case of a 15-year-old female patient with a history of rheumatic heart disease who presented with acute sequelae of the disease, infective endocarditis with septic embolization causing ischemic stroke and splenic infarction. Hemorrhagic transformation of the ischemic stroke precluded early surgical intervention for the cardiac lesion.

The case is of interest as it is now uncommon to see rheumatic heart disease in clinical practice especially causing such significant sequelae affecting morbidity.

Case Report

A 15-year old girl who migrated from Guyana four years prior presented with a two-day history of facial asymmetry, slurred speech and left sided weakness on a background of a two-week history of subjective fever, decreased appetite and one episode of central chest

pain, thigh and hip pain. Pain was relieved by analgesia and anxiolytic. There was a significant past medical history for a prior admission for valvular heart disease, carditis and commencement of anti-failure medication in her country of origin. Since migration, the patient has had no formal follow-up and is no longer on anti-failure medication.

On initial assessment, she was noted to have a left hemiparesis with facial asymmetry with frontal sparing and loss of her gag reflex. She was noted to have a Grade 3/6 pan-systolic murmur heard throughout the precordium, loudest at the apex with radiation to the axilla and louder on expiration. There was no carotid bruit. Three blood cultures taken within an hour revealed Streptococcus. An urgent CT scan and ECHO were performed. The CT brain revealed a right ischemic insular cortex cerebrovascular accident. The ECG revealed normal sinus rhythm with atrial ectopic pacemakers interspersed with normal sinus beats. ECHO revealed infective endocarditis with oscillating vegetation on the A2 segment of the mitral valve with moderate mitral regurgitation and a dilated left atria.

Rheumatic heart disease with major critical emboli; right cerebrovascular accident and infective endocarditis were diagnosed based on clinical history, radiographic findings and echocardiogram. She was managed medically with intravenous antibiotics and intensive, comprehensive physiotherapy with a plan for further surgical intervention for vegetation removal and valve repair. She was monitored closely in the intensive care unit for further complications of her disease.

On day 15 of her ICU admission, the patient was noted to have an acute worsening of her clinical condition; worsening hemiparesis and severe abdominal pain. Urgent repeat imaging was done at this time and revealed haemorrhagic conversion into the area of the previous ischemic stroke but no extension of the insult on CT brain along with a splenic infarct seen on CT abdomen. Clinically, the patient continued to deteriorate over the next 5 days as she developed hypotension, anisocoria, new onset seizures and altered mental status requiring intubation and ventilation. A repeat CT brain done at that time revealed extension of the haemorrhage with extension into the posterior fossa and subarachnoid space with midline shift. Neurosurgery was consulted and a decompressive craniotomy was offered to her parents which was refused. Supported care was offered with inotropic pressor support and mannitol with clinical improvement allowing for extubation on Day 33 of ICU stay. She continued to improve clinically; she is alert and receiving intensive, comprehensive physiotherapy. Her most recent ECHO revealed a decrease in the size of the vegetations with increased stability. Her long-term plan is for surgical intervention a minimum of 6 weeks post her haemorrhagic conversion of her stroke.

Investigations

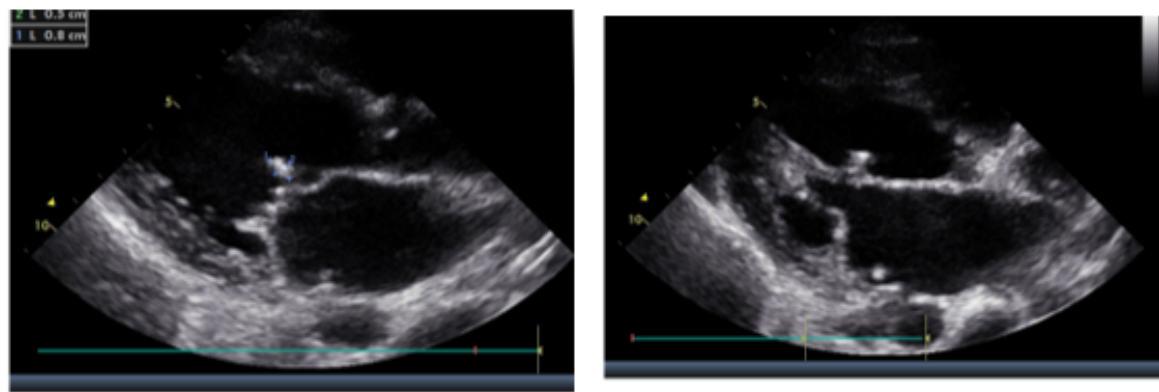


Figure 1 and 2: Initial ECHO: oscillating vegetations on the A2 segment of the mitral valve with mitral regurgitation and a dilated left atria.

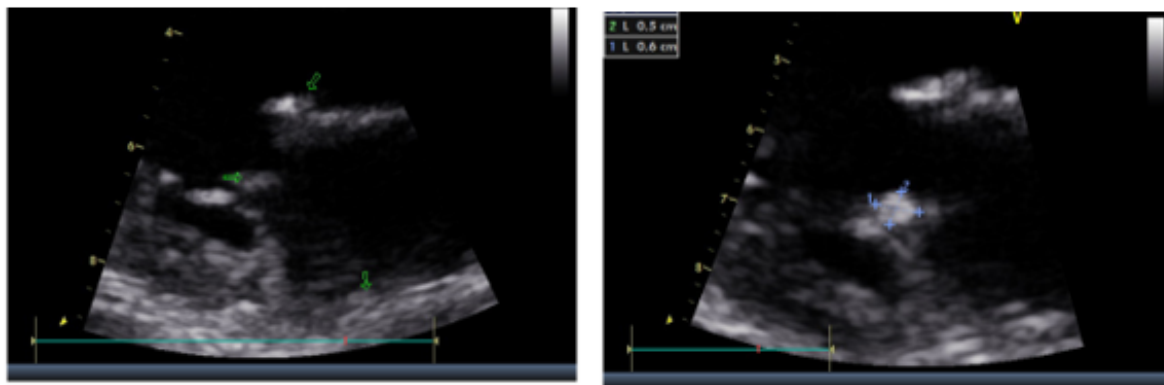


Figure 3 and 4: Day 7 of admission: New vegetations noted on the posterior wall of the left atrial above the posterior mitral valve leaflet 1.2 x 1.4cm mobile and pedunculated along with vegetation on the P2 and the initial vegetation remains.

Repeat CT brain: Hyper-density at previous stroke site. No extension of stroke haemorrhage. CT abdomen no appendicitis or mesenteric ischemia. Impression: small haemorrhage extension at the previous CVA.

Repeat due to hypotension: Thrombus in right MCA, extension of haemorrhage from previous ischemic site into subarachnoid space.

CT abdomen: splenic infarct.

Repeat: Cerebral oedema in R fronto-parietal region with midline shift noted. Subarachnoid haemorrhage appears unchanged.

Discussion

Acute rheumatic fever is a nonsuppurative, immune-mediated consequence of Group A Streptococcal pharyngitis. Acute rheumatic fever typically presents two to three weeks post untreated pharyngitis with arthritis, carditis, chorea, subcutaneous nodules and erythema marginatum. Rheumatic heart disease as is presented in the case above is a sequelae of recurrent or severe acute rheumatic fever which can result in permanent cardiac valve damage and predispose the patient to significant morbidity and mortality. Oftentimes, the first presentation of rheumatic heart disease occurs due to various long-term structural and haemodynamic complications including: heart disease, atrial fibrillation and stroke [3]. Thus, the main objective is to identify and treat acute rheumatic fever early to prevent sequelae of the disease.

Rheumatic heart disease despite being a preventable cause of acquired heart disease, remains a major cause of cardiovascular disease in developing countries though its prevalence has diminished greater in the industrialised world where adequate treatment of Group A pharyngitis has been instituted. Still, it is estimated that 12 million person world-wide are infected and two-thirds of those who are affected are children between the ages of five and fifteen [4]. Rheumatic disease is diagnosed in patients with a history of acute rheumatic fever and pathologic cardiac murmur. The most common lesion found on echocardiogram is mitral regurgitation which can occur alone or in combination with other defects. These patients are managed using periodic clinical and echocardiographic evaluation with the frequency based on the severity to assess for progression and minimise risk of complications. Treatment includes secondary prophylaxis: annual influenza vaccine and meticulous dental hygiene, along with intervention for valvular defects as indicated. Complications of rheumatic heart disease include: arrhythmia, heart failure, endocarditis and stroke.

Strokes and infective endocarditis are both severe complications of rheumatic heart disease with 3% to 7% of all strokes in developing countries directly attributed to rheumatic heart disease [5]. Stroke's are embolic in nature due to dislodgement of emboli formation on the defective heart valve. Emboli can be also be septic embolic when there is co-existing infective endocarditis as is the likely cause in our case.

Infective endocarditis is an infection of the endocardium and/or heart valves that results in the formation of vegetations which damage endocardial tissue and valves. It can arise as a complication of rheumatic heart disease as the microbes can adhere to the damaged valves and form vegetations. In children with underlying heart disease, streptococcus and staphylococcus infections being the most common and most destructive bacteria. The clinical presentation is variable and depends on the extent of the disease and can generally be classified an acute or sub-acute. Children with acute disease presenting with high fevers and severely ill or sub-acute with non-specific complaints of; prolonged low-grade fever, fatigue, myalgia, arthralgia, weight loss, exercise intolerance. Patients may also present with features of embolic events to other organs, i.e., neurological symptoms due to septic emboli in the brain or features of glomerulonephritis. Early diagnosis is paramount given the high mortality rate of 1% to 5% [6].

The diagnosis of infective endocarditis is made in critically ill children based on clinical findings, microbiological findings and ECHO. Duke or modified Duke criteria is used to classify the certainty of diagnosis. In critically ill children, three separate venipuncture for blood cultures should be performed within one hour. Treatment includes: Empiric broad-spectrum antimicrobial coverage followed by culture-directed treatment once available along with symptomatic support. Surgical intervention may be warranted for signs of heart failure, severe valve dysfunction, recurrent embolisation, large vegetations or failure to respond to treatment [7]. The timing of surgery is based on the risk versus the need for urgent surgery. In patients with non-hemorrhagic strokes the recommendation is to delay surgery for 1-2 weeks to reduce the risk of haemorrhage conversion of the infarct and oedema during surgery as the risk of worsening neurological symptoms decreases with increasing time after the initial neurological event. This can be done in stable patients where the risk of additional emboli is deemed low. In patients with hemorrhagic strokes the standard recommendation is to delay surgery for 3 - 4 weeks to reduce the risk of further intracranial bleed during surgery.

Conclusion

With the decline of rheumatic fever globally, it is important to have a high index of suspicion in diagnosing children with high fever and neurological manifestations. Meningitis and encephalitis being the most common diagnoses with such symptoms, in this technological era, a proper history, and physical examination may be able to point the pediatrician to the diagnosis of rheumatic endocarditis with embolic phenomena. This case report is a reminder of the importance of basic clinical skills including history-taking in order to rule out the now rare diagnosis of rheumatic heart disease.

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Conflict of Interest

None of the authors has no conflict of interest.

Informed Consent

Obtained from the all the authors and parent of this case.

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