Epistaxis as the initial presentation in a case of rheumatic heart disease

Monika Pathania¹, Vyasa Kumar Rathaur², Archana James Pattupara¹, Swati Arya³, Shashank Bansal⁴, Nidhi Kaeley¹

¹Department of Medicine, AIIMS, Rishikesh, ²Department of Pediatrics, GDMC, Dehradun, ³Department of Anesthesiology, AIIMS, Rishikesh, Uttarakhand, ⁴Dr. Barooah Cancer Institute, Guwahati, Assam, India

Abstract

Rheumatic heart disease (RHD) remains prevalent in developing nations and shows varied presentations, causing diagnostic challenge. Here, we report a case of RHD in a 13-year-old boy who presented with recurrent epistaxis as the initial symptom. On detailed work-up, echocardiography revealed the incidental finding of involvement of mitral and aortic valves but the patient never had any symptom as per the diagnostic criteria for RHD. This report highlights the clinical and epidemiological significance of atypical presentations as such cases might go undiagnosed and untreated, seeking medical attention in advanced stages, which would have otherwise contributed to the actual prevalence of the disease in the population.

Keywords: Aortic regurgitation, mitral regurgitation, rheumatic epistaxis, valvular heart disease

Introduction

Majority of the cases of rheumatic heart disease (RHD) are diagnosed with the help of modified Jones criteria. But atypical presentations of the entity cause diagnostic dilemma and can be easily missed. It becomes extremely important for primary care physicians to have a bird’s-eye view so as to correctly and timely diagnose and refer these cases for further management. Prolonged or recurrent episodes of bleeding should alarm the clinician about the causes of epistaxis like bleeding disorders or rarer causes like RHD, as in this case. This case report emphasizes the importance of a forgotten, nonspecific, and unusual presentation of RHD.

Case Report

A 13-year-old boy presented with undocumented on and off low grade fever since last 15 days. He also experienced three episodes of sudden onset, moderate amount of epistaxis since then, subsiding within 15–20 minutes of onset. There was no history of bleeding diathesis, trauma, or anticoagulant intake. General examination of the patient revealed pallor. The pulse of the patient was collapsing. The blood pressure was 140/10 mmHg, with wide pulse pressure. He was afebrile and the respiratory rate was 14 per minute. Cardiovascular examination revealed hyperdynamic apex at the sixth intercostal space in the anterior axillary line. S1 and S2 were normal but a high-pitched early diastolic murmur along the left sternal border was heard. The peripheral signs of aortic regurgitation (AR) including de Musset’s sign, Quincke’s sign, and Hill’s sign were positive. The patient denied any cardinal cardiac symptoms. He had history of recurrent sore throats over the last 4 years. The patient had no history of chest pain, nausea, vomiting, palpitations, diarrhea, rash, arthralgia, myalgia, hematuria, or headache. There was no bleeding, discharge, or congestion on nose and throat examination. The findings of respiratory and abdominal examinations were within normal limits. RHS and infective endocarditis were close differential diagnosis.

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Further investigations revealed mild anemia (9.5 g/dl) with normal leucocyte and platelet count, raised antistreptolysin O titer (453 IU), and raised erythrocyte sedimentation rate of 45 mm per first hour. Liver and kidney function tests were within normal limits. Three samples of blood culture showed no growth after 48 hours of incubation. Chest X-ray revealed biventricular enlargement. Electrocardiogram was significant for left ventricular hypertrophy with strain pattern and echocardiography showed left ventricular muscle hypertrophy [Figure 2], thickening and calcification of aortic cusp along with significant regurgitation across the aortic and mitral valves [Figures 1]. No vegetation was observed on the heart valves as suggested by echocardiographic evaluation [Figures 1-3]. A diagnosis of RHD with acute rheumatic fever was made. He was started on prednisolone (in view of acute rheumatic fever with carditis) and penicillin prophylaxis. His parents were counseled regarding valve replacement.

Outcome and follow-up
The patient was symptomatically better, with no further episodes of epistaxis. His parents did not agree for valve replacement due to financial constraints. The patient continues to be on penicillin prophylaxis.

Discussion
RHD is a major public health concern in the developing nations with a mean prevalence of 2.1/1000 among school going children (5–15 years of age) and contributing to 6.6% of cardiovascular admissions.[1,2] Recurrent attacks of acute rheumatic fever are more common in the adolescent and younger age groups, while RHD shows its peak prevalence between 25 and 40 years.[3] RHD has a female preponderance of about two times as in males.[3] Even though the disease is very rare in developed countries, they do encounter atypical presentations of the disease. The modified Jones criteria for diagnosis of rheumatic fever effectively diagnose the condition in most cases but varied presentations might lead to undiagnosed/misdiagnosed cases. In the original Jones criteria (1944), the minor criteria included epistaxis, abdominal pain, and pulmonary findings which were later removed due to the lack of specificity of these symptoms.[3]

Epistaxis is a usual presentation in emergencies, ear nose throat/ otorhinolaryngology and medicine outpatient settings. About 60% of the population experience nose bleed at least once in their lifetime, of which only 6–10% seek medical care.[5,6] As found in a German study, the hospitalized patients were just 6% from children less than 15 years.[7] The common causes being trauma (nose picking, facial injury), hypertension, bleeding diathesis, dry climate, and idiopathic causes, to site a few. Recurrent episodes call for further examination to look for causes including polyps or nasopharyngeal diseases like juvenile angiofibroma especially in children.[8,9] Bleeding could be anterior from the Kiesselbach’s plexus of vessels or posterior from the branches of the sphenopalatine artery or the woodruffs plexus.[10] Epistaxis can be controlled in most of the cases by pinching the nose and applying pressure.

There is a 4% probability of developing epistaxis in a patient with acute rheumatic fever which increases to 9% in those who had recurrence of RHD.[11] The probable pathogenesis of this presentation can be attributed to the increased susceptibility of the capillaries owing to rheumatic heart associated vasculitis.
and fibrinoid degeneration of the fibrous tissue which prevents effective platelet aggregation and hemostasis.[1,2]

The key in unlocking this atypical presentation was the collapsing pulse and widened pulse pressure which called for further detailed examination and investigations. As defined by the World Heart Federation for echocardiographic diagnosis of RHD in individuals less than 20 years, the involvement of both the mitral and aortic valve gave the diagnosis.[3]

The involvement of heart valves shows varied pattern. The most commonly involved is the mitral valve, followed by aortic and rarely tricuspid and pulmonary valves. As observed in a study on the pattern of valvular involvement, combined lesions are found in more than 55% of the cases where combination of mitral stenosis and regurgitation with AR is the commonest.[3] About 20% of the cases show mitral and AR, as seen in our case. Chronic AR causes gradual left ventricular volume overload that leads to left ventricular enlargement and eccentric hypertrophy. Pure single valve lesion is less frequent, mitral stenosis being the commonest.[13]

This case report is significant as it shows a male with RHD at an early age with initial presentation as epistaxis, which is infrequent and not among the diagnostic criteria. This patient had multivalvular disease without any cardinal symptoms of the cardiovascular system, which was peculiar. It is possible that such atypical presentations are being missed or undiagnosed and remain “subclinical” in the population of both developing as well as developed countries. They can present with cardiac failure later, as it would be undetected in the initial stages especially when any cardiac symptomatology is absent. Clinicians should be aware of such presentations and appropriate prophylaxis and treatment should be initiated.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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