Rheumatic Heart Disease: Presentation and Management Dilemmas

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ABSTRACT

Rheumatic Heart Disease (RHD) is a major public health concern in developing countries. Late presentation and diagnosis, difficulties in accessing medications in remote areas, and loss to follow-up patients adequately may plausibly explain the suboptimal clinical outcomes. RHD is commonly encountered during childhood and young adulthood and is a major cause of disabilities and preventable deaths.

The objective of this case report is to highlight on the occurrence of RHD and some challenges to care of RHD cases presenting to health facilities in resource-limited settings. This may also serve to alert authorities and medical practitioners of the need to ensure the early and adequate diagnosis of RHD given the availability of resources appropriate for its management. Further, although early diagnosis is key, preventive measures need to be put in place, medications made available, and reliable patient follow-up teams established as a holistic package of care.

Keywords: Rheumatic Heart Disease, Diagnosis, Valvulopathy, Management, Resource Limitations.

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Introduction

Rheumatic Heart Disease is a systemic immune process that is the sequelae to group A beta haemolytic streptococcal infection of the pharynx. It results from either single or repeated attacks of rheumatic fever that result in rigidity and deformity of valve cusps, the fusion of the commissures, or shortening and fusion of the chordae tendineae.[1]

RHD is highly prevalent in resource-limited settings where access to health care is inadequate and exposure to group A beta-haemolytic streptococcus is unchecked leading to the cause of the most cardiovascular morbidity and mortality especially among vulnerable and marginalized groups including children, adolescents, pregnant women, and poor and indigenous populations.[1,2] It is still prevalent in many parts of developing countries, including sub-Saharan African countries where the prevalence is 1 to 3 for every 1000 school children and other regions such as Oceania and South Asia. Moreover, recent studies have shown that globalization, migration, and refugee crises have led to the

evolution of RHD in developed countries making RHD a global health problem.[3]

The substantial burden of RHD has led to an expansion of health policy, programming, and research aimed at combating the disease in endemic regions4 hence, concerted efforts are underway in various parts of the world, including sub-Saharan Africa, to scale up health workers' capability to satisfactorily manage patients that present with a sore throat.[5,6]

Typically, RHD presents as left-sided valvulopathy, mitral (35%), and aortic valves (12%). However, this does not exclude an involvement of the tricuspid and pulmonary valves and may further complicate atrial fibrillation, stroke, heart failure, pulmonary hypertension, and infective endocarditis.[7]

This case report describes a presentation of rheumatic valvulopathy involving right-sided valves, in particular, the pulmonary valve with arrythmias and syncope.

We present a case of a female 17 years old, a resident of a suburban community in Zambia, and the firstborn child in her family currently attending senior secondary school.

The patient came in as a referral from a first-level hospital to the University Teaching Hospital for further management of recurrent fainting episodes. At the point of referral, the patient was unconscious and was initially resuscitated in the emergency department before being transferred to the medical filter section.

Clinical Assessments

History: The patient came in with complaints of two episodes of a sudden loss of consciousness and generalized body weakness for a day. She had no overt symptoms until around noon of that eventful day when she experienced a sudden loss of consciousness. The circumstances surrounding the event were that the patient was eating when she suddenly felt dizzy and lost consciousness. She did not report having experienced palpitation, headache, or dizzy spells before the event and further added that she did not experience any nausea or vomiting prior. The patient lost consciousness for about 10 minutes at home and once again just before reaching the hospital. There was no report of any jerking movements, tongue biting, or bladder incontinence during the unconscious phase. The patient was oriented after the episode but could not recall what happened and for how long she had lost consciousness. The patient also reported having experienced similar events about 3 years prior and had been investigated and examined but to no avail. The patient reported no history of seizure disorder. There were no mood changes, weakness of any limbs, and no tingling sensation was noted.

Systemic Review

Gastrointestinal System: The patient's last meal was just before the event; her appetite was not altered. She had no history of painful oral ulcers, difficulties/pain in swallowing, abdominal pain, nausea, and/or vomiting. The patient was passing stool which was soft and non-bloody. Of note, the patient reported having experienced one fainting episode in the past after she was straining at stool.

Respiratory system: She had no history of running nose, cough, difficulties in breathing, and/or chest pain. The patient had never expectorated bloody sputum before but had reported having chronic sore throats before which she was treated for. The patient had no alteration in voice and no noisy breathing was reported.

Genital urinary system: Patient was able to pass urine without difficulty or experiencing pain. No abnormal vaginal discharge was reported and her menstrual cycles and flow were normal.

Musculo-Skeletal and Skin: The patient reported no joint pains or swelling, muscle aches, rashes, hypopigmentation, and/or desquamation of the skin.

Past Medical History: Patient had never been admitted to the hospital for any medical cause nor did she have any surgeries done on her. The patient was not diabetic, not epileptic and reported no epileptiform seizures in the past. She was non-asthmatic, had no known TB exposure and her HIV status was negative (verbal report). The patient had been managed in the past several times for tonsillitis and syncope episodes but no definitive diagnosis was made.

Drug History: The patient was not on any medications currently and denied any use of herbal medication in the recent past. She had no known food or drug allergies.

Family History: She had a positive family history of hypertension in the father but had no known history of epilepsy, asthma, diabetes and tuberculosis. There was no history of similar presentations in other family members but there was a history of a sudden cardiac death that occurred to her uncle (paternal).

Social History: She is a high school pupil in her eleventh grade and firstborn in a family of 4. She is single and Christian by religion. She reported no history of alcohol consumption or smoking.

Physical Examination

We examined a female client who was lying supine on a propped-up bed at about 30 degrees. Generally, the patient was alert, stable, and oriented to time place, and person. On closer examination, the client had no finger clubbing, no Jane way lesions, or Osler's nodes where noted. The capillary refill rate was less than 3 seconds. The patient was cold to touch and had a cannula in situ. Her pulse was 110 b/min regularly irregular full volume non-collapsing with a blood pressure of 100/68mmhg. There was no presence of axillary or cervical lymphadenopathy. The patient was plethoric with hyperemic conjunctiva but there was no scleral jaundice. The oral cavity was clear with no ulcers and no thrush and no central cyanosis.

On Chest Examination: there were no obvious deformities, no scars, no spider naevi, or distended veins. Further examination revealed equal chest expansion with each inspiration and resonant chest in both anterior and posterior aspects. There was equal air entry bilaterally with vesicular breath sounds.

The precordium had normal activity with the apex beat palpable in the left anterior axillary line and no thrills but there was a left

parasternal heave. The first and second heart sounds were auscultated with a wide splitting S_2 and P_2 was soft. There was a systolic murmur (Graham Steell murmur) heard over the left parasternal area.

Abdominal Examination: The abdomen was flat, with no flank fullness, there were no scars and no stretch marks of note. There was no abdominal tenderness and the liver span was 10cm. There were no palpable masses and no organomegaly.

Cranial nerve exam: All the cranial nerves were intact with no dysfunctions.

Limb Examination: Both the upper and lower limb's power was 5/5 with no atrophy or increased muscle bulk. There was no spasticity or rigidity in the flexor and extensor myotomes. Deep tendon reflexes were normal.

Differential Listing

1. Pulmonary regurgitation with cardiac syncope in a patient with recurrent sore throat.
2. Pulmonary stenosis with cardiac arrhythmias and syncope.
3. Arrhythmogenic right ventricular cardiomyopathy with cardiac syncope in RHD.

Diagnostic Focus and Assessment

- History of recurrent sore throat and tonsillitis, dizziness spells, and loss of consciousness.
- On examination, there was a plethoric conjunctiva. The apex beat was palpable in the left midaxillary line, presence of a left parasternal heave and left parasternal systolic murmur.
- Investigations which include electrocardiogram, echocardiogram, chest x-ray, and full blood count/differential count.
- Admitted to cardiology unit for consult with cardiologist.

The thought process was in the line with the history of recurrent episodes of sore throat and tonsillitis making a high index of suspicion of Rheumatic fever which had complicated to RHD which further affected the heart valves predominantly affecting the right side of the heart.

Test Results of Note

ECG: Atrial flutter with right ventricular hypertrophy and right atrial hypertrophy.
Echo: results were not seen.
Chest x-ray: Cardiomegaly and downward displacement of the diaphragm with increased pulmonary vascular markings.
FBC/DC: It showed polycythemia of $18x10^3$cells/µL. At this point, a diagnosis of Pulmonary Regurgitation with syncope in RHD with atrial flutter and cardiomegaly was made.

Therapeutic Focus and Assessment

Anti-heart failure medications with anti-coagulation were prescribed and administered. Further, the Consultant recommended an echocardiogram to detect valvular lesions and assess ejection fraction and chamber dilatation, and put the patient on prophylactic penicillin to reduce the risk of recurrences that may further exacerbate the ongoing problem. The patient was scheduled for follow-ups and adherence

counseling toward medications and medical reviews was offered.

Discussion

Summary of the Case and Results: This is a case of female/17 years old, who presented with two episodes of syncope and malaise for a day preceded by dizziness. Additionally, the patient reported having recurrent episodes of sudden syncope about 6 years ago and recurrent episodes of sore throat despite treatment. This presentation is more suggestive of the RHD secondary to post-streptococcal throat infection affecting the heart valves causing circulatory compromise. However, there was no history of headache, blurred vision, heart palpitations, cough, dyspnea, orthopnea, chest pain, anorexia, nausea, vomiting, fever, body rash, mood changes, and/or epilepsy.

On examination, the patient was alert, oriented, and afebrile to touch. There was a regularly irregular tachycardic pulse suggestive of arrhythmia with normal blood pressure. There was a plethora suggesting increased blood viscosity which may result from chronic anoxia or a myeloproliferative disorder. Precordium was normoactive with the apex beat in the 5th intercostal space midaxillary line suggesting cardiomegaly and a left parasternal heave pointing to right ventricular hypertrophy. S_1 and S_2 were heard with a wide splitting S_2 and a soft P_2, and a diastolic murmur over the left upper parasternal area indicating pulmonary valve incompetence. However, no finger clubbing, Jane Way lesions, Osler's nodes, palmar pallor, central cyanosis, and the patient's oral hygiene was good ruling out the possibility of infective endocarditis.

The full blood count showed polycythemia which may have resulted from chronic anoxia hence increased erythropoiesis leading to mismatch between plasma and haematocrit concentration. Chest X-ray revealed enlarged cardiac shadow and downward displacement of the diaphragm with increased pulmonary vascular markings and the ECG depicted an atrial flutter with features of right ventricular hypertrophy and right atrial enlargement. This resulted from increased end-diastolic blood volume with strain on the right ventricle and eventually the right atrium.

Importance of the case

Rheumatic heart disease is the most important long-term sequela of acute rheumatic fever due to its potential to cause disability or death.[8] and the incidence of the disease is higher in children and adolescents between the ages of 5 and 25 years.[9] The finding is consistent with this case, as the patient is an adolescent with a history of recurrent episodes of sore throat and tonsillitis which have been there for over five years. Females have a higher risk of having a poor outcome from RHD as shown by the patient in discussion. This is further supported by a study conducted,[3] and suggesting that sex differences have also shown to be risk factor for RHD with a female predominance in a ratio of 2:1 and in another showing that females have a worse prognosis compared to males.[9]

RHD is more prevalent in resource-limited settings[1] of which Zambia a sub-Saharan country is categorized. Lack of routine screening and inconsistent medical supplies due to resource constraints in addition to the poor social-economic standing of the patients may present challenges to the care of the

RHD patients leading to poor clinical outcomes.

In chronic rheumatic heart disease, the most common finding on ECG is sinus tachycardia, heart blocks, and left atrial enlargement secondary to mitral stenosis.[7] The findings in this case presentation suggest otherwise. The ECG findings for our patient revealed features consistent with atrial flutter, right ventricular hypertrophy, and right atrial enlargement. Further, rheumatic heart disease commonly presents with left-sided valvulopathy and is the predominant cause of mitral stenosis.[10] But this case describes a relatively atypical presentation of RHD involving right-sided valves with arrhythmias and syncope.

Conclusion

RHD is still a global public health burden especially in a younger population and regions of low socio-economic status. It has the potential of causing high morbidity and mortality as it impairs the functionality of the heart. This case write-up brings to light the clinical relevance of RHD and its diverse course and presentation. The case further highlights the importance of employing thorough history, physical assessment, and investigations to establish the diagnosis definitively and institute appropriate treatments to achieve good clinical outcomes. Patient education on adherence to medication and the early presentation should be emphasized to help mitigate fast deterioration, recurrences, and complications associated with RHD

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References

1. Dass C., Kanmanthareddy A (2020). Rheumatic Heart Disease. Michigan State University. https://pubmed.ncbi.nlm.nih.gov/30855870/
2. World Health Organisation (2018). Rheumatic Fever and Rheumatic Heart Disease. Seventy-first world health Assembly. https://apps.who.int/gb/ebwha/pdf_files/WHA71/A71_R14-en.pdf
3. Mutagaywa R.B., Wind A.M., Kamuhabwa A et al (2020). Rheumatic heart disease anno 2020: Impacts of gender and migration on epidemiology and management. European Journal of Clinical Investigation. https://dx.doi.org/10.1111%2Feci.13374
4. Zuhlke L., Engel E., Karthikeyan G., Rangarajan S., Mackie P., Cupido B., Mauff K., et al.[2015].Characteristics, complications, and gaps in evidence-based interventions in rheumatic heart disease: The Global Rheumatic Heart Disease Registry (the REMEDY study). https://doi.org/10.1093/eurheartj/ehu449
5. Watkins D., Zuhlke L., Engel M., Daniels R., Francis V., Shaboodien G., Kango M, et al (2016). Seven key actions to eradicate rheumatic heart disease in Africa: the Addis Ababa communique. Cardiovasc J Afr. https://doi.org/10.5830/cvja-2015-090
6. Musuku, J., Lungu, J.C., Machila, E. et al. Epidemiology of pharyngitis as reported by Zambian school children and their families: implications for

demand-side interventions to prevent rheumatic heart disease. BMC Infect Dis 17, 473 (2017). https://doi.org/10.1186/s12879-017-2563-x

7. Sika-Paotonu D., Beaton A., Raghu A., Steer A., Carapetis J (2017). Acute Rheumatic Fever and Rheumatic Heart Disease. Centre for International Child Health, Australia. https://pubmed.ncbi.nlm.nih.gov/28379675/
8. Shulman S.T., Bisno A.L (2015). Nonsuppurative poststreptococcal sequelae: Rheumatic fever and glomerulonephritis. Philadelphia (PA): Elsevier. https://doi.org/10.1016/B978-1-4557-4801-3.00200-9
9. Chin K.T (2019). Paediatric Rheumatic Heart Disease. Cardiac Disease and Critical Care Medicine.
10. Noubiap, J.J., Agbor, V.N., Bigna, J.J. et al. Prevalence and progression of rheumatic heart disease: a global systematic review and meta-analysis of population-based echocardiographic studies. Sci Rep 9, 17022 (2019). https://doi.org/10.1038/s41598-019-53540-4

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