Case Report

A case report of isolated rheumatic tricuspid regurgitation and pericarditis

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Abstract

Rheumatic fever (RF) is a complex syndrome in which the human body develops antibodies against β-haemolytic streptococcus, and triggers inflammation in various organs. RF valvulitis mostly affects the mitral valve (MV) and, to a lesser extent, the aortic valve (AV). Isolated rheumatic tricuspid valve (TV) disease and pericarditis is extremely rare and is not well described in the RF guidelines. The current case demonstrates the echocardiographic findings of TV valvulitis of RF and describes the presentation and electrocardiogram (ECG) changes in pericarditis. We present the case of a 16-year-old male patient who developed upper respiratory tract infection followed by typical pericarditis chest pain. The patient had no history of joint pain or swelling, but was found to have a tricuspid regurgitation (TR) murmur upon examination. Laboratory investigations revealed an elevated erythrocyte sedimentation rate, and elevated C-reactive protein and antistreptolysin O titres. ECG showed a wide-spread 1.5 mm upward concave ST-segment elevation. In echocardiography, the TV opened well with markedly thickened leaflets and severe TR, while the MV and AV were normal in both structure and function. The diagnosis of RF was established and treatment with high-dose aspirin and antibiotics was initiated. The treatment led to resolution of the chest pain. Our case highlights that the physical and lab findings of rheumatic TR are similar to those of rheumatic mitral regurgitation, with the exception of a high-velocity jet. Similarly, the presentations of rheumatic pericarditis are similar to other types and may also respond to high-dose aspirin. Finally, physicians should be familiar with both the common and rare complications of RF because the guidelines have placed Middle East region among the high-risk countries.
Introduction

Rheumatic fever (RF) is an autoimmune disorder caused by group A β-haemolytic streptococcal infection. It is considered one of the most common diseases worldwide, particularly in developing countries. The manifestations of RF were described by Jones in 1944, who divided them into two categories (known as Jones criteria), major and minor. The major criteria include carditis, arthritis, chorea, erythema marginatum, and subcutaneous nodules. The minor criteria include fever, arthralgia, elevated erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP), and prolonged P-R interval on electrocardiogram (ECG). A diagnosis of acute RF can be made if a patient has two major or one major and two minor Jones criteria, along with evidence of a preceding streptococcal infection.

Rheumatic heart disease (RHD) has been reported in 40–80% of patients with RF. The endocardium becomes involved in the process of inflammation early in the process of RHD, and valve incompetence occurs subsequently. Later, fibrosis develops, which leads to valvular stenosis. RF predominantly affects the left-sided valves. The aortic valve (AV) is infrequently involved in mitral valve (MV) diseases in patients with RF. The most common AV lesions are regurgitation (AR), followed by mixed AR and stenosis (AS), and finally, isolated AS.

Rheumatic tricuspid valve (TV) lesions are seldomly encountered and mostly in association with left side-valvular disease. The prevalence of organic rheumatic TV disease is approximately 7%, but 99.3% of these patients have concomitant MV disease. Given its rarity, the echocardiography findings of isolated rheumatic TV disease are not well established. Similarly, acute rheumatic pericarditis is extremely rare. It has been estimated that 5% of patients with acute RF will go on to develop clinically significant pericarditis.

This case describes the clinical presentations of two extremely rare manifestations of RF, which, although historically described, are poorly documented in the current guidelines. We demonstrate that the echocardiographic findings of isolated severe rheumatic tricuspid regurgitation (TR) are similar to those described for rheumatic MR. Additionally, we found that the presentation of rheumatic pericarditis is similar to other forms, and may respond well to treatment with high-dose aspirin. These cases can enlighten physicians about the unusual complications of RF in high-risk geographical areas so they can be readily recognised and treated.

Case presentation

A 16-year-old male patient presented to the cardiology clinic with chest pain that had persisted for 2 weeks. He was a healthy high school student with no past history of medical or surgical diseases or prior hospitalisation for any illnesses. Three weeks earlier, he developed flu-like symptoms that were associated with a sore throat and a fever of 38.2 °C, which were managed with antipyretic and pain killer. Ten days later, he started to feel gradual chest pain that increased in severity. He experienced pain all over the chest but predominantly on the left side. It felt like pressure pain, which increased with lying flat and improved with sitting and standing. This pain was not radiating, and it became more severe until he sought the opinion of cardiologists. There was no history of joint pain, stiffness, or swelling.

On physical examination, his vital signs were within the normal range. The musculoskeletal examination was intact, and the respiratory and abdominal examinations were normal. The estimated central venous pressure was 4 cm above the sternum, with no lower limb oedema. Pericardial examination revealed a nondisplaced apex beat with no heaves or thrills. The auscultation of the heart showed normal first and second heart sounds, with loud pansystolic murmur heard all over the cardiac areas but more so in the lower sternal border; the murmur increased with inspiration and there was no pericardial rub.

Laboratory data revealed a white blood cell (WBC) count of 14.2 × 10^9/µL, with predominant neutrophils as was CRP, which was at 83 mg/L (0–10 mg/L). The chest radiography on presentation was unremarkable.

A 12-lead ECG showed 1.5 mm diffuse concave up ST-segment elevation with no evidence of reciprocal changes and a normal P-R interval (Figure 1). The antistreptolysin O (ASO) titre was elevated at 420 IU/mL (<200 IU/mL). The echocardiography showed markedly thickened TV leaflets which opened well with severe TR by colour Doppler, seen in both the 4-chamber and parasternal short access views. The vena contracta width was 2 cm and TR occupied most of the right atrium. Continuous wave assessment across TR revealed a dense semi-triangular jet, which peaked early in systole, with a velocity of 2 m/s. The right ventricle and right atrium were not dilated, but the inferior vena cava measured 2.2 cm, which collapsed by >50% with inspiration. The estimated right ventricular systolic pressure was 30 mmHg. The pulmonary valve, MV, and AV were normal in structure and function, and the left ventricle was normal in size and function. No pericardial effusion, or intra-atrial or intra-ventricular septal defects were observed (Figures 2 and 3).

The patient was diagnosed with RF since he fulfilled one major (carditis) and two minor components (fever, elevated ESR, and CRP) of the Jones criteria, with a proof of preceding streptococcal infection evident by the elevated ASO titre.
Figure 1: A 12-lead electrocardiogram showing widespread concave up ST-segment elevation with no reciprocal changes and normal P-R interval.

Figure 2: (a) The mitral valve (MV) opens well with no evidence of leaflets thickening or stenosis. The aortic valve (AV) is normal in structure. (b) Compared to MV leaflets, the tricuspid valve (TV) leaflets are markedly thickened, particularly at the tips.

Figure 3: (a) Severe tricuspid valve (TV) regurgitation by colour Doppler; the jet is central and its ratio to the right atrium (RA) is >50%, with a vena contracta of 2 cm. No significant MV regurgitation was detected by colour Doppler. (b) Continuous wave Doppler across the TV showed a pansystolic, dense, semi-triangular shaped jet that peaked early, with a peak velocity of 2 m/s. The inferior vena cava (IVC) was 2.2 cm, but collapsed well with inspiration. The right ventricular systolic pressure (RVSP) was 30 mmHg.
Treatment with co-amoxiclav and high dose aspirin were initiated. The patient’s chest pain improved significantly after being administered with 14 days of aspirin, after which, he was placed on secondary prophylaxis with monthly intramuscular benzathine penicillin. He will undergo frequent echocardiography surveillance to assess the need for surgical intervention.

Discussion

This report illustrates the case of a patient who initially presented with upper respiratory tract infection symptoms and fever, and subsequently developed typical pericarditis chest pain. Physical examination revealed a murmur, which is not typically seen in patients with RF. He fulfilled one major Jones criteria (carditis) and two minor criteria (fever and prolonged ESR/CRP), with an elevated ASO titre. ECG showed widespread concave up ST-segment elevations, and ECG confirmed isolated severe TR and thickened leaflets, with normal structure and function of the MV and AV. Isolated severe TR and pericarditis are rare manifestations of RF that are not well described in the literature, and, to the best of our knowledge, this case is the only available published evidence in KSA, and probably in the Middle East.

In recent years, the prevalence of RF has declined significantly in developed countries. However, in developing countries, it remains a major challenge that consumes significant health resources. The annual incidence of RF is estimated to be <0.5/100,000 in developed countries, and >100/100,000 in developing countries.10,11 Because of these significant differences in distribution, the Jones criteria underwent a major revision in 2015, which further divided cases into low-risk or moderate-high-risk groups. This is based on geographical distribution; Low risk patients are those in countries where the incidence of RF is ≤2 per 100,000 school-aged children, or an all-age prevalence of ≤1 per 1,000 population year. Patients in countries with a higher incidence are considered to be in the moderate-high-risk group. Most Middle East counties, including KSA, are within the moderate-high-risk geographical area. This change in the guidelines was introduced to avoid over-diagnosis of RF in developed countries, as well as to prevent its under-diagnosis in moderate-high-risk counties.12

The revision involved changes in the definitions of several Jones manifestations. In the major criteria, the number of joints involved in the arthritic process differentiates the moderate-high-risk and low-risk groups. In the moderate-high-risk group, monoarthritis can be considered a major criterion; however, in moderate-high-risk patients, the inflammation has to be present in more than one joint (polyarthritis). In the minor criteria, the differences between the groups relate to fever and ESR. In low risk populations, fever is defined as a temperature ≥38.5 °C, and elevated ESR has to be ≥60 mm within the first hour. However, in the moderate-high-risk group, a fever ≥38.0 °C and an ESR ≥30 mm in the first hour are considered diagnostic. This indicates that the diagnosis of RF in moderate-high-risk geographical areas is now less strict to increase early detection and prevent late complications.

The prevalence of children with RHD in KSA is higher than the global rate. In 1990 a cohort of 9,418 schoolchildren aged 6–15 years estimated a prevalence of 3.1 per 1,000 children.13

RHD is a common cause of cardiovascular disease and death. Indeed, it is responsible for approximately 30–40% of all cardiovascular disease-related hospitalisations, and is a leading indication for cardiac surgery in most developing countries.14,15

RHD predominantly affects the left-sided valves, causing regurgitation, stenosis, or mixed pathology. Valvulitis of the MV happens frequently, while valvulitis of AV occurs less frequently.16 An early study described that 74% of patients with RHD will go on to develop MR.17 Although isolated MR is common, it can be concomitant with MS, causing a mixed valvular lesion.18 It has been shown that mixed MR and MS is seen in 27.9% of patients with RHD.19,20 Although isolated MS occurs less frequently, with a prevalence between 7 and 24% among patients with RHD,21 RF is considered leading the cause of MS in 95–99.3% of individuals aged <50 years.22,23

AV disease is seen in 19.5% of the cases. It is usually seen in association with MV lesions.24 Isolated AV disease is rare with an overall prevalence of 8%.25,26

Rheumatic disease of the TV in the absence of MV involvement does occur, but it is exceptionally rare. In a 30-year single centre experience including 328 consecutive patients who underwent TV surgery for RHD, isolated TV disease was observed in 12 patients, 199 patients had triple-valve disease (MV, AV, and TV), 114 patients had double valve disease (MV and TV), and 3 patients were diagnosed with AV and TV disease.27 Moreover, the Euro Heart Survey, which was conducted in 92 centres of 25 countries, showed that isolated TV lesions are very rare, with an estimated prevalence of only 1.2% in a sample of 5,001 patients.28

Four case reports of isolated rheumatic TV disease have been published in peer-reviewed journals; two patients had isolated TV stenosis,29,30 one patient had isolated severe TR,31 and one patient was diagnosed with mixed pathology.32

In KSA, the pattern of valvular lesions in patients with RHD in the different geographical areas is not significantly different from what is known worldwide. In the central area, two studies explored the prevalence of RHD. In the first study, MR was the most common valvular lesion, followed by mixed MV and AV disease, and finally isolated AV lesions.33 Similar findings were observed in the second study, but with a longer observation period of 9 years. However, this second study identified two patients with TR concomitant with MR.34 In a cohort of 162 patients with evidence of RHD in the Almadinah Almunawwarah region, left-sided lesions were detected in all patients.35 Furthermore, over an 11-year observation period in Damman, left-sided lesions were diagnosed in all patients.36 In
the southern area, 40 patients with rheumatic carditis were identified, and 6.7% of them who developed TR were found to have concomitant MR. Isolated rheumatic TV has never been reported in KSA.

The guidelines that describe the echocardiographic features of rheumatic carditis are only focused on the aortic and MV lesions, and do not describe rheumatic TV disease. This case is showing that the findings of isolated rheumatic TR in echocardiography are similar to rheumatic MR with regards thickening of the leaflets, its presence in at least two views, and pansystolic jet by continuous wave Doppler, however, with the exception of high-velocity jet, which is likely explained by the low pressure in the right-side of the heart.

The pathology of pericarditis in RF has been described in the past, but is poorly addressed in the current guidelines, and information on its prevalence and significance is limited. Pericarditis of RHD is characterised by fibrous sterile exudates and the formation of fibrous strands in the pericardial space, it is observed in 5% of patients with RHD and is extremely rare in the absence of valvulitis. Despite extensive fibrinous deposition into the pericardial space, it usually resolves without complication.

There are few published case reports regarding pericarditis in RF, and these patients present with pericardial effusion or its complication (tamponade). This case is unique because the patient presented with typical pericarditis pain and did not have pericardial effusion; however, it also existed with carditis. The 12-lead ECG showed the typical ST-segments changes of pericarditis. Although the patient responded well to high-dose aspirin, a severe form of constrictive pericarditis was described, this suggests that early recognition and aggressive therapy may prevent late pericardial complications in patients with RF pericarditis. Previous studies that examined the prevalence and distribution of RHD in KSA did not report or describe pericarditis.

Conclusion

This case is the only available evidence in the middle East region that isolated TR and pericarditis can occur in patients with acute RF. Furthermore, it enlightens physicians about these findings since this issue is poorly addressed in the guidelines. Rheumatic pericarditis has a similar presentation to other forms of the disease and can be treated with high-dose aspirin. The echocardiographic findings of rheumatic TR are similar to those described for rheumatic MV, with the exception of the high-velocity, which is likely attributed to lower pressure in the right ventricle compared to the left. In high-risk geographical areas like KSA, RF can be under-diagnosed in patients presenting with these two exceptionally rare manifestations and a high index of suspicion is required to prevent long-term complications.

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Conflict of interest

The author has no conflict of interest to declare.

Ethical approval

The patient understood that his clinical data is going to be utilized for publication purposes and he agreed on that. The patient gave consent to use his data for publication. All his information remained confidential and efforts were made to conceal his identity. This study was approved by the Scientific Research Ethics Committee of The College of Medicine at Taibah University (study ID: 015-1442) dated 04 October 2020.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jtumed.2020.11.007.

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