Pulmonary Edema in the Acute Stage of Rheumatic Fever Treated with Double-Valve Replacement in a Pediatric Patient

Yilmaz Yozgat[1] Selcuk Uzuner[2] Osman Yesilbas[2] Mustafa Ogur[2] Kahraman Yakut[1] Can Yilmaz Yozgat[3] Hafize Otcu Temur[4] Yasin Ay[5]

[1] Department of Pediatric Cardiology, Bezmialem Vakif University, Istanbul, Turkey
[2] Department of Pediatric Critical Care, Bezmialem Vakif University, Istanbul, Turkey
[3] Faculty of Medicine, Bezmialem Vakif University, Istanbul, Turkey
[4] Department of Radiology, Bezmialem Vakif University, Istanbul, Turkey
[5] Department of Cardiovascular Surgery, Bezmialem Vakif University, Istanbul, Turkey

Address for correspondence Yilmaz Yozgat, MBBS, MD, Department of Pediatric Cardiology, Adnan Menderes Bulvarı, Vatan Caddesi, Bezmialem Vakif University, Istanbul 34093, Turkey (e-mail: yozgatyilmaz@yahoo.com).

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Abstract

Cardiogenic pulmonary edema (CPE) is a rare clinical condition of acute rheumatic fever (ARF) in the early stage. Generally, CPE can be convalesced by steroid and anticongestive treatment. Herein, we describe a case of a 14-year-old boy with ARF presenting with bilateral pulmonary edema secondary to acute mitral and aortic insufficiency. In this case, the pulmonary edema of ARF was successfully managed by combined surgical replacements of both valves.

Keywords

- bilateral pulmonary edema
- acute rheumatic fever
- childhood

Introduction

Acute rheumatic fever (ARF) is prevalent in developing countries. Bilateral pulmonary edema is a rare clinical manifestation of ARF in the early stage. Herein, we describe a case of a 14-year-old boy with ARF presenting with bilateral pulmonary edema secondary to acute mitral and aortic insufficiency. Initially, the case did not respond to antipulmonary edema and anti-inflammatory therapies. During the acute stage of ARF, pulmonary edema was successfully managed with aortic and mitral valve replacement.

Case Report

Herein, we describe a case of a 14-year-old male who was born and raised in Syria and recently took refuge in Turkey. The patient was presented to the emergency clinic with lethargy and shortness of breathing. Before the patient was admitted to the emergency department, the patient had these complaints since about 15 days, but, recently, the complaints had progressed in severity. On physical examination, the patient was ill-looking, pale, and lethargic. The patient's weight was 38 kg (<3 percentile). He was tachycardic with a heart rate of 150 beats per minute with S4 gallop, and tachypnea with 32 breaths per minute. The patient had a weak peripheral pulse, jugular vein distention, and mild hypotension (blood pressure: 80/36/50 mm Hg). The patient's oxygen saturation was 94% at room temperature and had a long capillary refill time (CRT) (CRT > 5 seconds). During the respiratory system examination, the patient was in the orthopneic position, and there were intercostal and subcostal withdrawals and nasal flaring. Clinical examination of the lungs showed bilateral coarse crepitations.

On cardiac examination, we detected a holosystolic murmur with the severity of 4/6 at the apex, which thrilled upon palpation. A chest X-ray revealed patchy infiltrate, which was predominantly located in the left and lower lobes (bilateral pulmonary edema). The chest X-ray also demonstrated prominent pulmonary conus, closed left costophrenic sinus, and disappearance of the heart borders (►**Fig. 1**). A subsequent echocardiogram showed the existence of first-degree atrioventricular block (PR: 0.22), and V5, V6, and T were negative. An echocardiographic inspection revealed the left

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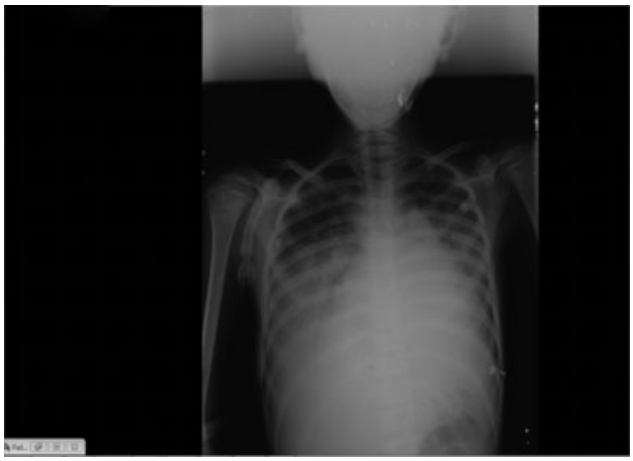

Fig. 1 Chest X-ray demonstrating bilateral pulmonary edema.

atrium and the left ventricle (LV) to be extremely large. Also, the inspection disclosed heavy mitral and aortic insufficiency. Another intriguing finding on the echocardiogram was the detection of ruptured chordae tendineae of the anterior mitral valve and incomplete coaptation of the aortic valve (►**Videos 1** and **2**). The patient's LV systolic function was normal. Laboratory investigations showed hemoglobin of 10 g/dL, white blood cells of 11 × 103/μL, renal impairment (urea: 88 mg/dL; creatinine: 2 mg/dL), C-reactive protein of 25.5 mg/L, and erythrocyte sedimentation rate of 35 mm/hour. Antistreptolysin O titer was >1,300 IU/mL (reference: <200). The first sets of cardiac enzymes were as follows: 3.5 IU/mL troponin I, 450 IU/mL creatine kinase, 56 IU/L CK-MB (creatine kinase myocardial band) isoenzyme. Growth of group A β-hemolytic streptococci in throat culture was negative. Our team thought that the patient had bilateral pulmonary edema presenting with ARF with carditis due to severe heart valve insufficiency.

Video 1

Echocardiography showing a ruptured chordae tendineae of the anterior mitral valve. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0039-3399580.

Video 2

Echocardiography demonstrating incomplete coaptation of the aortic valve. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0039-3399580.

Our team took a preemptive measure and transferred the patient to the pediatric intensive care unit (PICU). The patient was started on noninvasive ventilation (NIV) and medical treatment, including milrinone, dopamine, dobutamine, and Lasix IV infusion. Primary prophylaxis of ARF was given with intramuscular (IM) benzathine penicillin (1.2 × 106 units/kg). Two sets of blood cultures from two separate veins were obtained to rule out the possibility of infective endocarditis. The patient was prescribed oral steroid for the treatment of ARF with carditis (four doses of prednisolone 2 mg/kg/day). On the fifth day in the PICU, the general patient condition was unchanged. Even though the patient was under the substantial treatment for pulmonary edema and orthopnea, the conditions were not convalesced by the treatment. No further growths on blood culture were detected. The patient's refractory pulmonary edema with underlying ARF was discussed in a council. The council was composed of a heart surgeon, two pediatric cardiologists, and two pediatric critical care specialists. There had not been any significant improvement in the clinic situation of the patient throughout his stay in the PICU. That is why the council decided that surgical valve replacement would be the optimal treatment option. However, our team faced a significant obstacle from the patient's parents. The parents did not consent for the surgical operation immediately.

On the 10th day in the PICU, our team performed the mechanical replacement of the mitral valve and aortic valve (►**Figs. 2** and **3**). After general anesthesia induction, a pediatric cardiologist performed a transesophageal echocardiography (TEE) and evaluated valve pathologies. Median sternotomy, aortic arterial, and bicaval venous cannulation were performed. Myocardial arrest and protection were achieved with antegrade and retrograde blood cardioplegia. After exploration of the left atriotomy, mitral valve leaflets were fibrotic and chordae had been thickened. There was

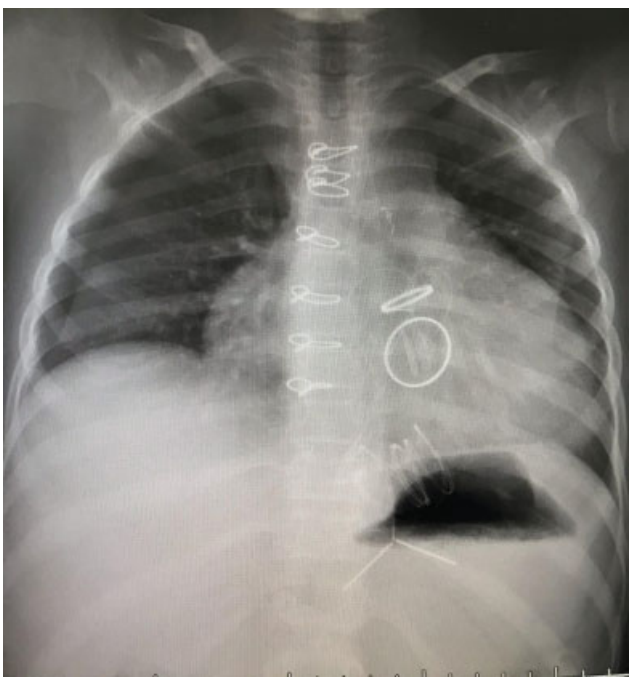

Fig. 2 Chest X-ray demonstrating the mechanical replacement of the mitral valve and the aorta valve after cardiac surgery.

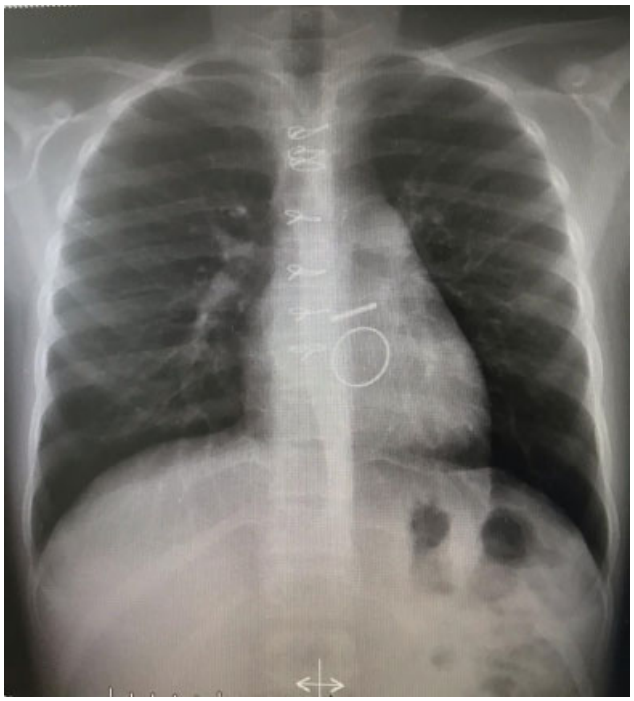

Fig. 3 Chest X-ray showing the heart 6 months later after the surgery.

rupture of an anterior chordae in the leaflet and fusion in both commissures. Oblique aortotomy was performed, and the aortic valve leaflets were fibrotic, thick, and retracted, and fusion was present in all three commissures. Aortic valve structure required total replacement, and mitral valve repair had a low chance of success. Both valve leaflets were resected. Aortic annulus #21 St. Jude Medical mechanical heart valve and mitral annulus #27 St. Jude Medical mechanical heart valve were implanted. Aortotomy and left atriotomy were primarily sutured. After cross-clamping, the heart worked in spontaneous sinus rhythm, and no cardiac block was observed. Cardiopulmonary bypass was performed with low-dose inotropic support (dobutamine 5 µg/kg/minute). Both valve and ventricular functions were checked with TEE. Following decannulation, bleeding control was performed, drainage tubes were placed, and the sternum was closed with steel wires. The patient was transferred back to the PICU. Aortic cross-clamp time was 86 minutes, and cardiopulmonary bypass time was 106 minutes. There was no hemodynamic problem throughout the intensive care unit follow-up, and a total of 150-mL drainage was performed in the first 24 hours. Postoperative mechanical ventilation time was 6 hours. The inotropic support was terminated at the fourth postoperative hour. Chest tubes were taken on the second postoperative day. Warfarin and low molecular weight heparin (LMWH) were prescribed with the first postoperative day. LMWH was stopped when the patient reached sufficient warfarin levels.

The patient's general condition ameliorated significantly during postoperative follow-up, and steroid treatment was completed in a month. Steroid treatment was slowly reduced and eventually stopped. At the same time, aspirin treatment was started. The patient was discharged from the hospital on the 40th day since the admission into the emergency department. A month after the patient was discharged from the hospital, his follow-up echocardiography disclosed standard prosthetic valves function without displacement. The patient continues to be attended at our outpatient clinic and is still under treatment for prophylaxis with IM benzathine penicillin (1.2× units/kg) on a half-month basis.

Discussion

ARF usually occurs by an autoimmune response to throat infection due to *Streptococcus pyogenes*. Cardiac involvement during ARF can result in rheumatic heart disease that may be the primary reason behind the cause of heart failure and premature mortality. The acute phase of pancarditis conspicuously shows itself with inflammation in pericardium, myocardium, and endocardium. Pericarditis and myocarditis do not cause any short-term significant hemodynamic problem at childhood. Valvulitis might result in severe hemodynamic deterioration. Endocardial involvement might cause edema, macrophage, and fibroblast infiltration in the valves, and formation of small thrombosis, verrucous vegetation, develop the closure surfaces of mitral and aortic leaflets. Annular dilatation growths in the valves and coaptation deteriorate the chordae tendineae in the mitral valve due to distention, which eventually results in valvular insufficiency. The chordae tendineae may also rupture.[1,2] The case presented with an acute stage of ARF and both insufficiencies of the aorta and mitral valve due to the ruptured chordae tendineae of the anterior mitral valve and incomplete coaptation of the aortic valve. The clinic manifestation of the disease revealed itself to be pulmonary edema. On echocardiography, we detected the chordae tendineae to be ruptured. Valvuloplasty or valve replacement during an acute attack of the disease is not a suitable treatment option. Valve replacement may become necessary for the long-term follow-up in symptomatic patients who have severe valve insufficiency and progressive left ventricular dilatation.[3]

In our case, throughout his stay in the PICU, the patient was put under treatment for NIV and severe intensive pulmonary edema, although his condition had not regressed since the admission. Our team was able to correct the patient's pulmonary edema by a double-valve replacement at the acute phase of rheumatic fever (RF), and at the same time, the patient continued his steroid treatment on his 10th day in the PICU. There have been few reports in the literature about combined surgical replacements of both valves with mechanical prostheses in adult patients with RF.[4–6] However, none of the reported cases are in acute stages of the disease. To the best of our knowledge, this will be the first pediatric case reported in the literature on the development of pulmonary edema in the acute phase due to RF with carditis that improved significantly after the surgical operation of double-valve replacement. Another crucial factor that should be considered is the patient has lived almost all of his life in a war-torn country, and

therefore it is right to say that the patient's health might have been affected because of silent carditis. Clinical manifestation of this condition might be a form of recurrence of the disease.

Our team had a tremendously difficult time to decide which type of valve should be used on the patient for his double-valve replacement. It is a known fact that biological valves are less durable than mechanical valves and that the patient would be likely to go through reoperation later in his life. Even though mechanical valves are durable, still there would be severe repercussions of long-term anticoagulation.[4] In this case, mechanical valve replacement was performed on both valves, and the patient was not responding to the intensive medical treatment. In patients with pulmonary edema secondary to RF, the disease can be convalesced by steroid and anticongestive treatment. Even in the acute stage, successful aortic and mitral valve replacement can be performed to alleviate pulmonary edema.

Conclusion

Bilateral pulmonary edema is a rare clinical manifestation of ARF in the early stage in pediatric patients. According to the literature, the disease can be convalesced by steroid and anticongestive treatment. If a patient does not respond to intensive medical treatment of pulmonary edema in the acute stage, combined surgical replacements of both valves become an optimal treatment option and can be performed safely and successfully while under steroid treatment.

Authors' Contributions

All authors participated in creating the content of the manuscript, editing, and providing final approval for submission. No undisclosed authors contributed to the manuscript.

Note

Permission was granted by the parents and patient to publish the case report.

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Conflict of Interest

None declared.

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