Extensive Right-Sided Endocarditis in Double-Chamber Right Ventricle Presented with Leukocytoclastic Vasculitis

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Abstract

Congenital heart disease is a risk factor for infective endocarditis. Ventricular septal defects and ventricular outflow tract obstructions are the most common causes of endocarditis in this population. We presented a patient diagnosed with leukocytoclastic vasculitis with renal and pulmonary involvement with right-sided infective endocarditis as an etiology for vasculitis.

Introduction

We report a patient with a Double-chambered right ventricle (DCRV) and extensive right-sided IE with initial presentation of leukocytoclastic vasculitis.

Double-chambered right ventricle (DCRV) is a form of right ventricular outflow tract (RVOT) obstruction caused by anomalous muscular or fibromuscular bundles dividing the right ventricle (RV) into two chambers: A proximal high-pressure and a distal low-pressure chamber. This disorder is commonly associated with membranous ventricular septal defect (VSD). It is shown that VSD and ventricular outflow tract obstructions can predispose patients to infective endocarditis (IE) (1, 2).

Case presentation

A 37-year-old female patient with petechia and purpura from 7 months ago was diagnosed with leukocytoclastic vasculitis confirmed by biopsy. Despite receiving anti-inflammatory treatments, she was admitted to our hospital due to the gradual progression of constitutional symptoms, pancytopenia, and new pulmonary and renal involvement. Diffuse petechia and purpuric lesions were detected, especially in the lower limb. In Table 1, initial lab data results are shown.

Splenomegaly was detected in the abdominal ultrasound. Computed tomography (CT) of the chest showed consolidations in the left upper lobe. Bone marrow aspiration and biopsy showed 40% cellularity with increased erythroid precursors. Megaloblastic changes in both erythroid and myeloid precursors were detected as well. According to these findings, myelodysplastic syndrome secondary to leukocytoclastic vasculitis was the main reason for megaloblastic changes. Megaloblastic anemia was also considered a second differential diagnosis. Due to the history of VSD in childhood, a tans thoracic echocardiogram was done. Multiple mobile masses in RVOT and highly mobile masses at the atrial side of the tricuspid valve and the ventricular side of the pulmonary valve were seen (Figure 1, video). A turbulent flow in RVOT with a peak gradient of about 80 mmHg was detected at the hypertrophied muscle bundle site (Figure 1, video). A transesophageal echocardiogram for further evaluation was performed. The muscle bundle in the right ventricle (RV) was divided it into two chambers: A proximal chamber on the side of the RV inflow and a distal chamber on the side of the pulmonary valve. Many masses were attached to both the muscle bundle and along the distal

chamber involving RV endothelium and pulmonary valve (Figure, video). Multiple masses and destruction of the tricuspid valve leaflets resulted in severe regurgitation (Figure, video).

The patient was diagnosed with DCRV and IE. After consultation with the cardiac surgeon and infectious disease specialist, empiric antibiotics therapy was initiated, and the patient underwent cardiac surgery. Pulmonary and tricuspid valve replacement with bioprosthesis valves was performed. Although VSD was not found in the echocardiogram, probably due to high RV pressure, surgery proved it and was repaired with a pericardial patch. The patient's condition improved after surgery and antibiotics therapy, and she was discharged with normal lab results. The patient was in good clinical and echocardiographic condition and free of any major event in the one-year follow-up.

Discussion

According to previous studies, unrepaired VSD is the most common congenital abnormality associated with the highest risk of infective endocarditis(1), followed by ventricular outflow tract obstruction(2). Both these conditions were present in this patient. Berglund et al. demonstrated that a small and unrepaired VSD could increase the incidence of IE more than 20 times compared with the general population (3).

Based on European Society of Cardiology guidelines, diagnosing right-sided endocarditis is more challenging than the left side due to the lower sensitivity of duke criteria (4), as our patient was diagnosed with right-sided endocarditis with a remarkable diagnostic delay.

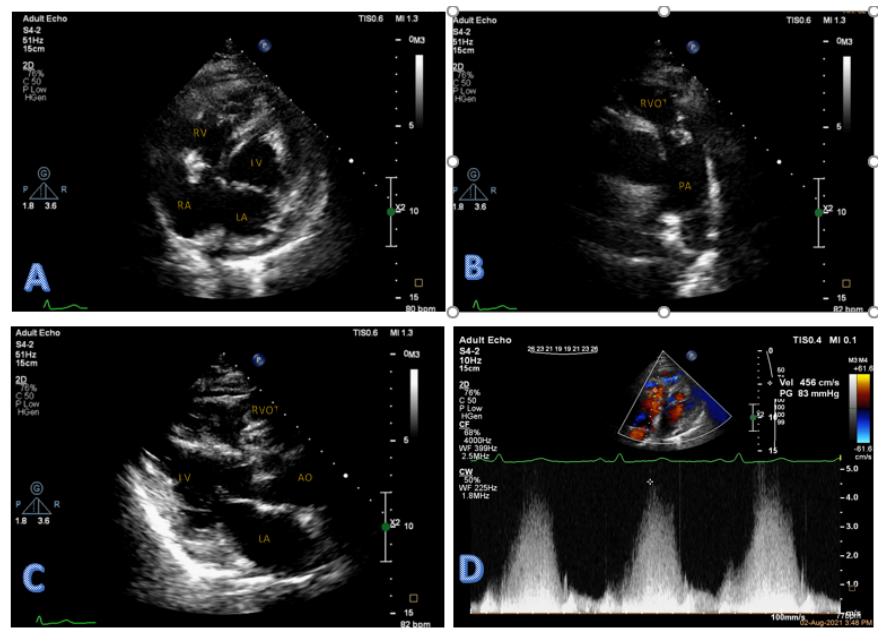
It has been shown that renal infarcts and non-immune complex-mediated glomerulonephritis are the most common renal lesions in right-sided IE by renal biopsy or necropsy examination. Septic embolism was the cause of renal infarcts in more than half of the patients (5).

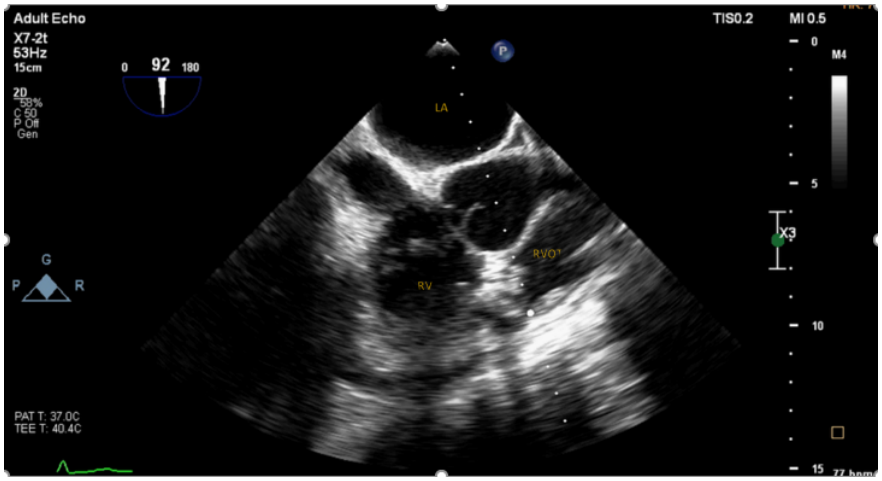
A study in France showed the existence of septic pulmonary emboli (SPE) in up to 70% of right-sided IE(6). Diagnosing SPE can be challenging due to nonspecific presentation and can remain unrecognized.

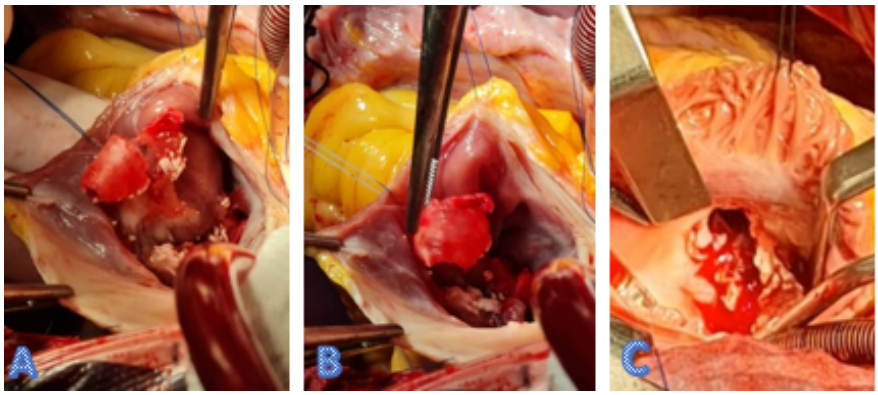
Subacute bacterial endocarditis (SBE) and ANCA-associated systemic small vessel vasculitis have overlapping clinical manifestations. Awareness of false positive ANCA tests in SBE and possible endocardial involvement in idiopathic ANCA-associated vasculitis is important due to different management(7).

Leukocytoclastic vasculitis can be a rare presentation of IE. A retrospective study of 138 biopsy-proven cutaneous leukocytoclastic vasculitis patients showed bacterial infection as the underlying cause in 4 (2.9%), three diagnosed with IE(8).

In conclusion, IE has a wide variety of presentations, imitating connective tissue disorders and vasculitis, so even subtle symptoms should be considered seriously, especially in patients at high risk for endocarditis.







References

1. Fortún J, Centella T, Martín-Dávila P, Lamas MJ, Pérez-Caballero C, Fernández-Pineda L, et al. Infective endocarditis in congenital heart disease: a frequent community-acquired complication. Infection. 2013;41(1):167-74.

2. Li W, Somerville J. Infective endocarditis in the grown-up congenital heart (GUCH) population. European Heart Journal. 1998;19(1):166-73.

3. Berglund E, Johansson B, Dellborg M, Sörensson P, Christersson C, Nielsen NE, et al. High incidence of infective endocarditis in adults with congenital ventricular septal defect. Heart. 2016;102(22):1835-9.

5. Habib G, Lancellotti P, Antunes MJ, Bongiorni MG, Casalta JP, Del Zotti F, et al. 2015 ESC Guidelines for the management of infective endocarditis: The Task Force for the Management of Infective Endocarditis of the European Society of Cardiology (ESC). Endorsed by: European Association for Cardio-Thoracic Surgery (EACTS), the European Association of Nuclear Medicine (EANM). Eur Heart J. 2015;36(44):3075-128.

4. Majumdar A, Chowdhary S, Ferreira MA, Hammond LA, Howie AJ, Lipkin GW, et al. Renal pathological findings in infective endocarditis. Nephrol Dial Transplant. 2000;15(11):1782-7.

6. Hoen B, Alla F, Selton-Suty C, Béguinot I, Bouvet A, Briançon S, et al. Changing profile of infective endocarditis: results of a 1-year survey in France. Jama. 2002;288(1):75-81.

7. Chirinos JA, Corrales-Medina VF, Garcia S, Lichtstein DM, Bisno AL, Chakko S. Endocarditis associated with antineutrophil cytoplasmic antibodies: a case report and review of the literature. Clin Rheumatol. 2007;26(4):590-5.

8. García-Porrúa C, González-Gay MA. Bacterial infection presenting as cutaneous vasculitis in adults. Clin Exp Rheumatol. 1999;17(4):471-3.