Cardiology in the Young

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Isolated pulmonary valve endocarditis leading to right ventricular outflow obstruction

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Brief Report

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Abstract

Isolated native pulmonary valve endocarditis is rare. We present a rare case of isolated native pulmonary valve endocarditis resulting in severe right ventricular outflow tract obstruction in an immunocompetent patient with surgically repaired ventricular septal defect caused by Burkholderia cepacia.

Introduction

Congenital Heart Disease (CHD) or prior cardiac surgery is becoming a more prevalent risk factor as opposed to rheumatic heart disease for Infective Endocarditis (IE). Historically, IV drug abuse has been associated with right heart infective endocarditis but isolated pulmonary valve involvement is rare.[1] We present a rare instance of isolated native pulmonary valve endocarditis resulting in severe right ventricular outflow tract obstruction in an immunocompetent patient with surgically repaired ventricular septal defect caused by *Burkholderia cepacia*.

Case report

A 23-year-old female presented with high-grade fever, cough, and pleuritic chest pain for the past three months. At the age of three, she had undergone surgical closure (using a pericardial patch) for a small doubly committed and juxta arterial ventricular septal defect causing aortic regurgitation. There was no history of IV drug abuse, prolonged intravenous therapy, or indwelling catheter placement; however, she had received blood transfusion for severe anaemia, a month prior to the current admission. Her examination revealed high-grade fever and sinus tachycardia (110 beats/minute). She was normotensive, acyanotic (oxygen saturation on room air 97%), and all peripheral pulses were palpable. She was pale but showed no skin rash, ulcers, nail changes, or other stigmata of IE. A grade IV/VI harsh ejection systolic murmur was heard in the pulmonary area.

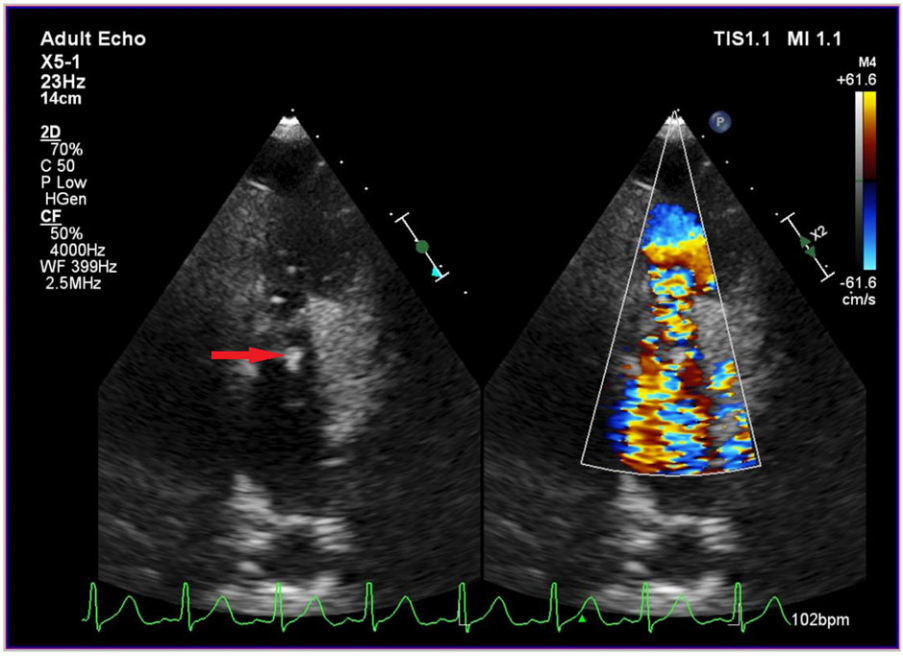

Figure 1. Still image of video 1 showing transthoracic echocardiography in modified short axis view showing a large vegetation (red arrow) attached to pulmonary valve.

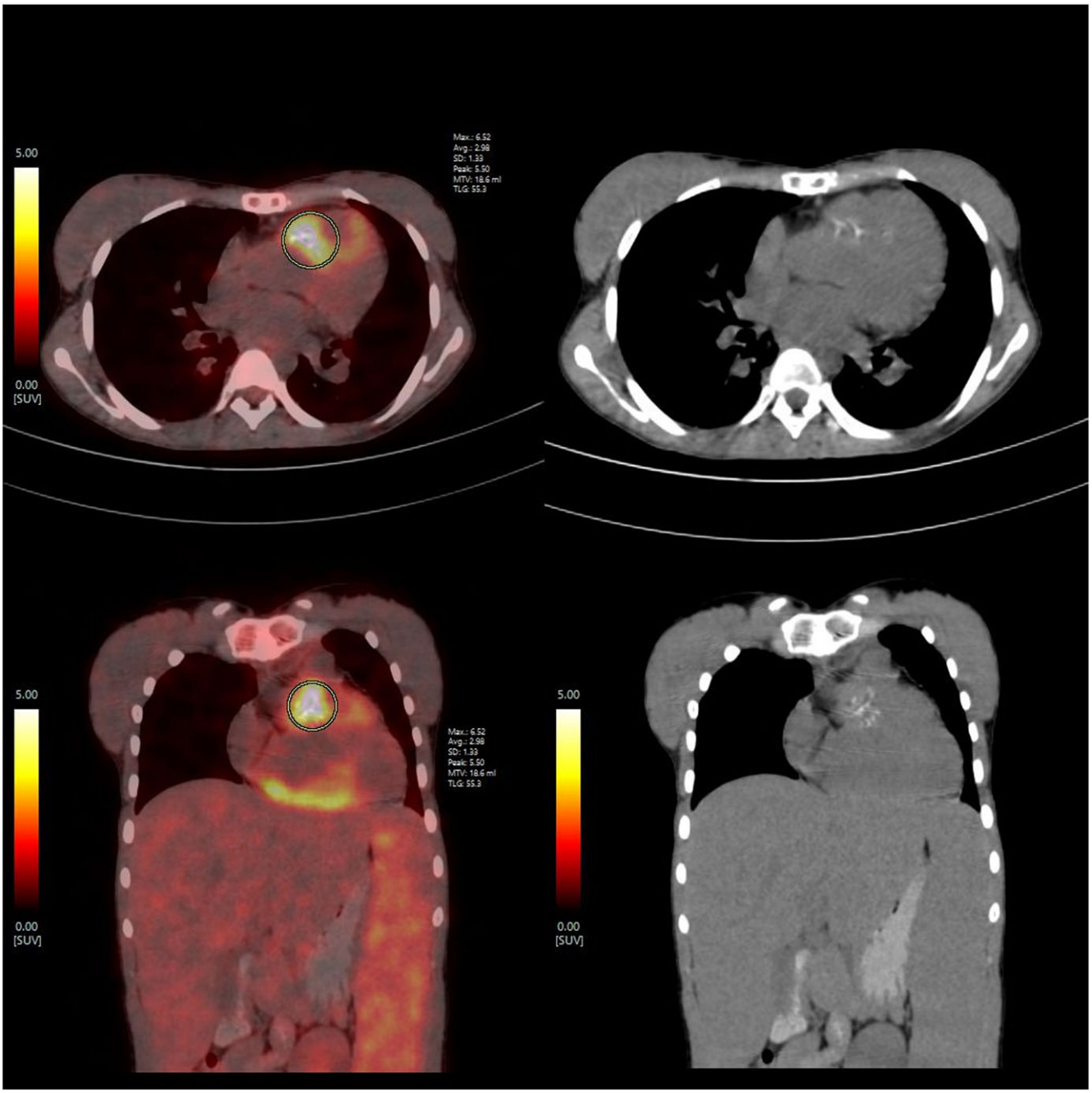

Figure 2. Whole body FDG-PET-CT image showing increased uptake in right ventricle outflow tract and subjacent myocardium.

Investigations showed severe anaemia with a haemoglobin of 6.8 gm%. Although her total lymphocyte count (8200 per mm^3) was normal, the markers of inflammation, such as erythrocyte sedimentation rate 70 mm/hour and C-reactive protein 114mg/dL, were elevated, suggesting infection. Rheumatoid factor was negative and urine analysis showed 12 pus cells and 4 red blood cells per high power field and proteinuria of 180 mg/dl in 24 hours sample. Work-up for cause of anaemia revealed an elevated lactate dehydrogenase 457 IU/L and indirect hyperbilirubinemia. Blood cultures sent from three sites grew gram-negative bacilli-*Burkholderia cepacia* sensitive only to ceftazidime, minocycline, and levofloxacin. Thus, suspecting IE an echocardiography was performed, which showed a large mobile pedunculated mass attached to the pulmonary valve with degeneration of the valve leaflets causing severe right ventricle outflow tract obstruction with peak systolic gradient of 90 mm Hg (figure 1). The surgical patch was intact. Whole body positron emission tomography showed high-grade uptake in patch area near pulmonary valve and subjacent right ventricular myocardium with small consolidatory areas of uptake in bilateral lower lung lobes suggesting septic pulmonary embolism (figure 2).

She was started on ceftazidime and levofloxacin in accordance with the sensitivity report and also received blood transfusions to correct anaemia. Despite four weeks of targeted antibiotic therapy, there was persistent fever, increasing titre of C-reactive protein, blood culture remaining positive for same organism, and her echo showed further degeneration of the pulmonary valve with the

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development of mild pulmonary regurgitation along with right ventricle outflow tract obstruction. The patient was therefore referred for surgery, in view of persistent sepsis with a virulent organism and pulmonary valve degradation.

Discussion

Isolated endocarditis of pulmonary valve is rare and accounts only for 2% of cases. Literature search did not reveal any other case of right ventricle outflow tract obstruction due to isolated pulmonary valve endocarditis. Common risk factors are intravenous drug abuse, prosthetic heart valves and conduits, pacemaker leads, indwelling central venous catheter, immunocompromised state, alcoholism, and congenital heart lesions as in this case.[1–3] The agent *Burkholderia capecia* is a virulent gram-negative bacillus that inhabits moist environments and can cause life-threatening infection owing to its intrinsic resistance to several antibiotics. Patients with cystic fibrosis, other granulomatous lung diseases, prosthetic heart valves, or immune-compromised states are at high risk for Burkholderia infections. To date, only a few case reports of *Burkholderia species* causing infective endocarditis have been reported.[4]

Usual complications of endocarditis involving the pulmonary valve include valvar regurgitation due to leaflet perforation, intramyocardial abscess, and/or septic pulmonary emboli.[1,2] However, in this case, the vegetation is large enough to cause right ventricle outflow tract obstruction in a ball valve fashion with septic embolism to the lower lobes of both lungs. There was no infundibular stenosis causing obstruction.

The current European Society of Cardiology Guidelines for diagnosis and management of infective endocarditis give a Class IA recommendation to Flurodeoxyglucose positron emission tomography (FDG-PET) as a modality to detect peripheral lesions in symptomatic patients. Our patient showed involvement of surgical patch and surrounding myocardium as well as septic pulmonary embolism. There was no evidence of systemic embolism.[5] Intravenous antibiotic therapy in accordance with the culture report remains the cornerstone for management of all cases of endocarditis. Specific indications for surgery, especially in right heart involvement, include difficult to eradicate organisms, large vegetations (≥1 cm), right-sided heart failure, persistent bacteraemia despite adequate anti-microbial therapy, recurrent pulmonary emboli, and intramyocardial abscess.[1]

Our patient had pulmonary valve endocarditis with a rare and difficult-to-treat organism viz. *Burkholderia capecia* and a large vegetation, which persisted despite one month of appropriate antibiotic therapy and caused septic pulmonary embolism. Hence, she was referred for surgical management. Potential challenges for surgical intervention include a redo-midline sternotomy procedure, which itself is a high-risk surgery, need for surgical patch excision, along with vegetation, and pulmonary valve replacement in an infected and inflamed milieu.

Conclusion

This case is most noteworthy for rare pulmonary valve endocarditis caused by rare and difficult to treat organisms.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951124026015.

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Competing interests. None.

Ethical standards. Patients informed consent was taken.

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