

Case study

ISOLATED PULMONARY VALVE ENDOCARDITIS ON AN UNDIAGNOSED CONGENITAL HEART DISEASE IN A YOUNG ADULT: A RARE CLINICAL ENTITY

Abstract

Background: Isolated pulmonary valve endocarditis (PVE) is a rare condition that accounts for 1.5–2% of all reported cases of endocarditis. Herein, we describe a rare case of isolated pulmonary valve endocarditis with a fortuitous discovery of a congenital heart disease in a young adult subject. Unlike other cases of right-sided endocarditis, we treated our patient both medically and surgically.

Case presentation: The patient was diagnosed with an isolated pulmonary valve endocarditis after blood cultures confirming the presence of *Abiotrophia defectiva*, a germ difficult to cultivate whilst the echocardiographic revealed a vegetation mass measuring 8mm long alongside with the discovery of a severe pulmonary valve stenosis and a large atrial septal defect (ASD) of 39mm wide. Septic pulmonary emboli were the first clinical manifestation in our patient. Both medical and surgical treatment was indicated based on dual antibiotics, removal of the vegetation, valvulotomy and closure of the ASD.

Conclusion: Both medical and early surgery therapy should be considered in patient with right-sided endocarditis associated with congenital heart disease for better outcome.

Keywords: Isolated pulmonary valve endocarditis, pulmonary valve stenosis, Atrial septal defect, *Abiotrophia defectiva*, Transthoracic echocardiography.

Introduction

“Right-sided infective endocarditis represents less than 10% of all infective endocarditis cases” [1] whereas Isolated pulmonary valve endocarditis (PVE) is a rare condition that accounts for 1.5–2% of all reported cases of endocarditis [2]. “It shares demographic, clinical, and microbiological features with tricuspid valve endocarditis” [3]. “Risk factors for developing PVE include intravenous drug abuse, central venous catheters and prosthetic valves. It is a challenging condition to diagnose mainly because of its non-specific signs and symptoms. Common clues for suspecting PVE include a new-onset of pulmonary valve insufficiency or recurrent lung infections due to septic emboli in high-risk individuals. Moreover, echocardiographic views used in the evaluation of pulmonary valves are limited, and as a result, vegetation on pulmonary leaflets can easily be missed” [2]. We describe a rare case of PVE without tricuspid involvement on an undiagnosed congenital heart disease revealed by septic pulmonary emboli in a young adult subject with no medical history who presented at the emergency department with an acute respiratory syndrome.

Case Presentation

A 35-year-old young woman with no personal or family history was admitted for NYHA stage III dyspnea accompanied by episodes of hemoptysis in a context of impaired general health, asthenia and weight loss amounting to 10Kg over the last 3 months.

The patient was initially hospitalized in the medical intensive care unit for acute respiratory distress in which a chest computed tomography scan performed (CT scan) showed an organized pneumonitis with low-abundance pericardial effusion. Patient also presented several episodes of febrile peak at 38.5°C for which she was transferred to the cardiology department for suspected endocarditis.

On somatic examination findings, patient was stable with a blood pressure (BP) of 120mmHg systolic blood pressure and 70mmHg diastolic blood pressure, tachycardia at 112 beats/minutes, febrile at 38.8°C and saturation of 98% in ambient air. The cardiovascular and lung clinical examination found bronchial rales in the lower right lung associated with spontaneous pleuritic pain on the same side. Cardiac auscultation found a systolic-diastolic murmur at the pulmonary focus whilst the rest of the clinical examination in search of infectious entry side was unremarkable except for a poor oral hygiene. The electrocardiogram (ECG) findings showed a regular sinus rhythm, right axis deviation associated with electrical signs of hypertrophic right ventricle (R/S ratio greater than 1) and negative T waves at the inferior leads.

(Figure1)

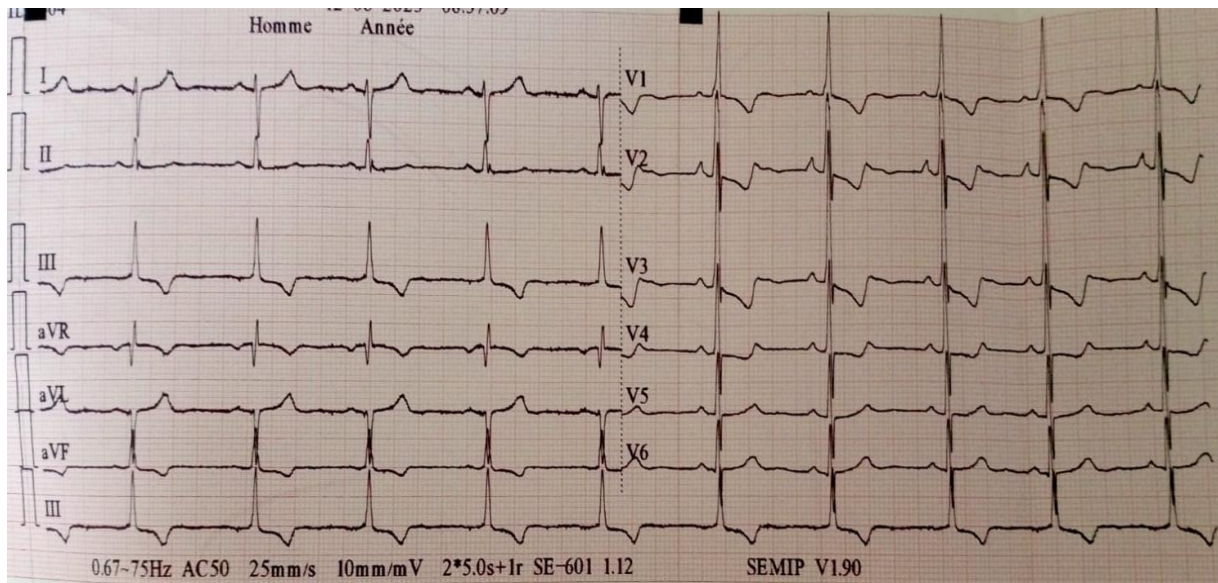


Figure 1: Electrocardiogram (ECG): showing a regular sinus rhythm, right axis deviation associated with hypertrophic right ventricle and negative T waves at the inferior leads.

In front of this clinical scenario, a chest CT angiogram was performed, revealed a right pulmonary embolism associated with pulmonary infarction and the presence of pulmonary nodule and micronodule of non-specific appearance (Figure 2).

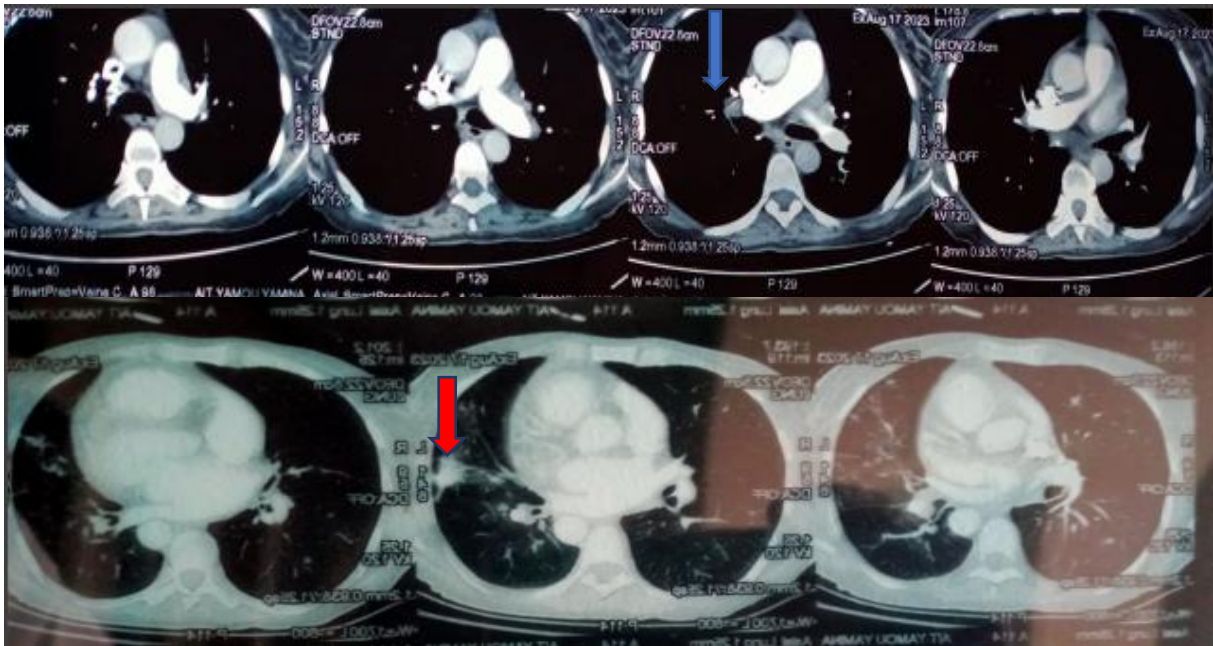


Figure 2: ChestCT angioscan: parenchymal and skeletal view: showing a right pulmonary septic emboli,(blue arrow) associated with pulmonary infarction of the right lung (red arrow) and pulmonary nodule and micronodule of non-specific appearance.

The transthoracic echocardiography done (TTE) showed a non-dilated left ventricle (TDDLv: 40mm indexed at 26mm/m²), non-hypertrophic with a preserved left ventricle ejection fraction (LVEF= 60%) and a dilated right atrium with a surface area of 27.8cm². Absence of mitral and aortic valves abnormalities, dilated right ventricle (RV)(BDRV: 42mm), hypertrophic (PWRV: 8mm) with preserved systolic longitudinal function (S'RV: 16cm/s and TAPSE: 23mm) (figure 3). Also, a severe pulmonary valvular stenosis (Gmax: 152mmHg, VmaxP: 6.3cm/s) associated with the presence of a vegetation at the level of the pulmonary valve measuring 8mm (Figure 4) was objectified. We also noted the presence of a large atrial septal defect (ASD), ostium secundum type (ASD measuring 39mm wide) with a bidirectional shunt (Figure 5). Measurement of the initial segments of the aorta was normal. The inferior vena cava was non-dilated and compliant without signs of pulmonary venous return. A low abundance pericardial effusion was visualized next to the right side of the heart.

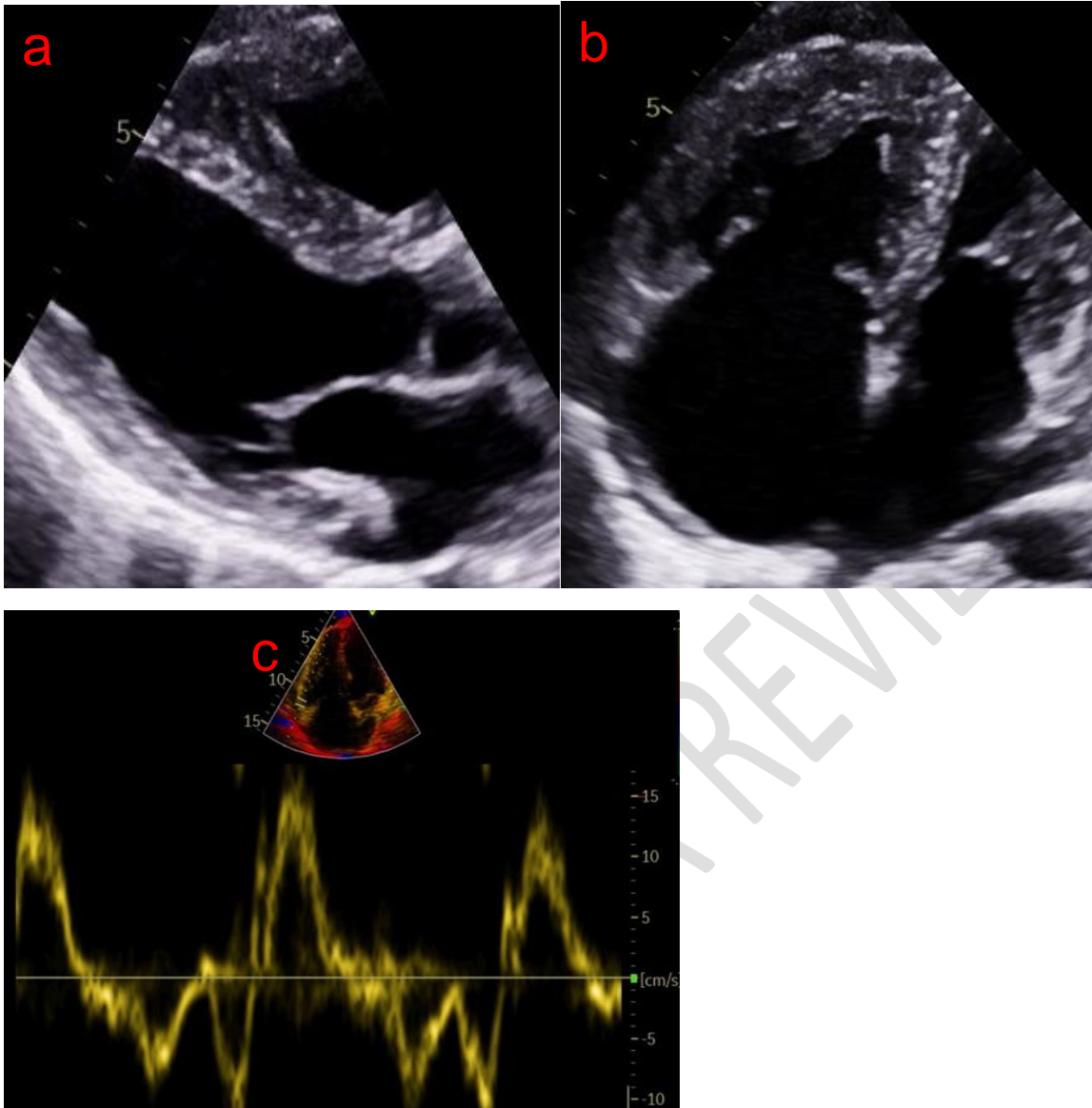


Figure 3: Transthoracic echocardiography (TTE): **a**-Parasternal long axis view: non-dilated and non-hypertrophic left ventricle (TDDL_V: 40mm indexed at 26mm/m²), **b**-Apical 4 chambers view: dilated right atrium with a surface area of 27.8cm² associated with a dilated right ventricle (RV) and hypertrophy of the posterior wall of the right ventricle measuring 8mm thick.

c-Tissue doppler of the tricuspid valve ring: showing a good longitudinal contraction function of the right ventricle with a Tricuspid annular plane systolic excursion value of 14.8m/s.

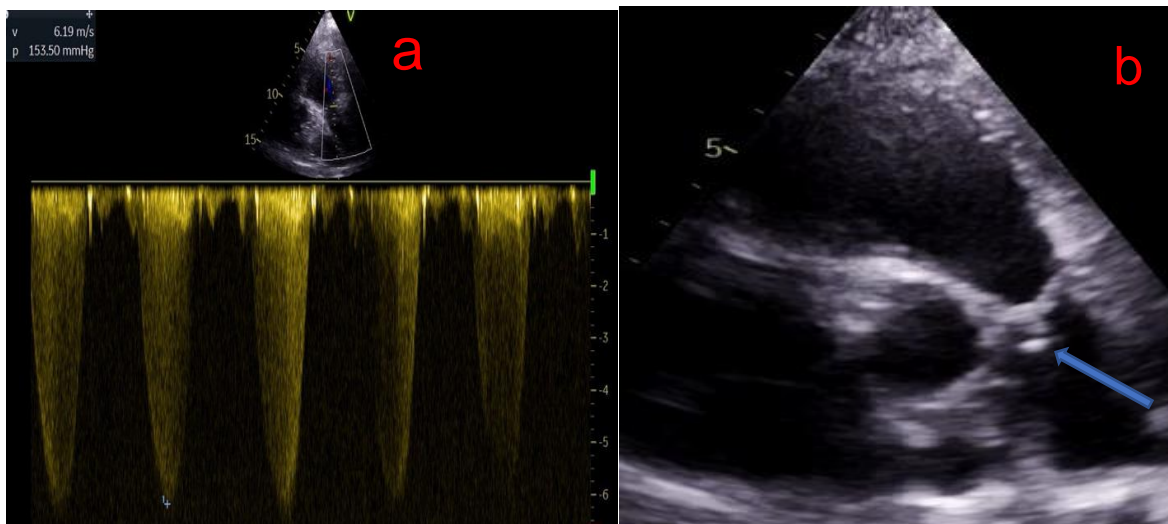


Figure 4 :TTE :Parasternal short axis view centered on the pulmonary valve; contiuous doppler (CW): **a**-flux of a severe pulmonary valvular stenosis (Gmax: 152mmHg, VmaxP: 6.3cm/s), **b**-2D;presence of a vegetation at the level of the pulmonary valve measuring 8mm at the level of its arterial side (blue arrow).

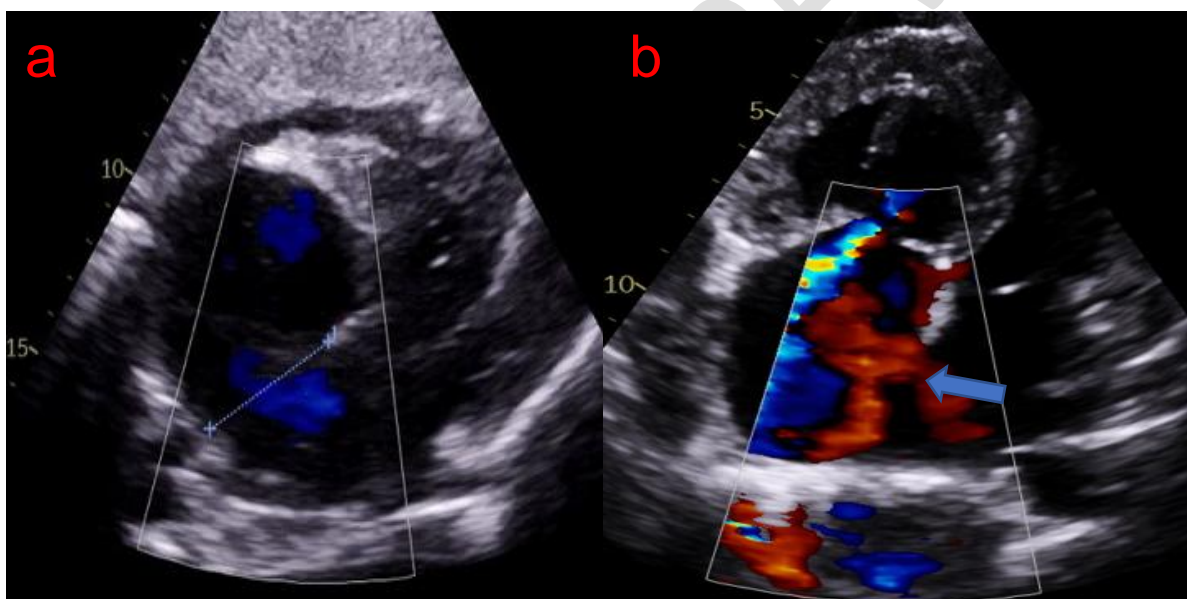


Figure 5 :**a**-Subcostal 4 chambers view showing a large atrial septal defect ostium secundum typemeasuring 39mm wide, **b**- Apical 4 chambers view showing a bidirectional shunt caused by the ASD.

A transesophageal echocardiography (TOE) was done for complementary information, objectified the presence of a mobile pedunculated hyperechoic image attached to the pulmonary valve measuring 9mm in length on the arterial side associated with pulmonary valve stenosis. The bubble test was positive confirming the presence of a large atrial septal defect (ASD) with bidirectional shunt (Figure 6). The rest of the exploration was unremarkable with a normal rate of emptying and filling of the auricle chamber.

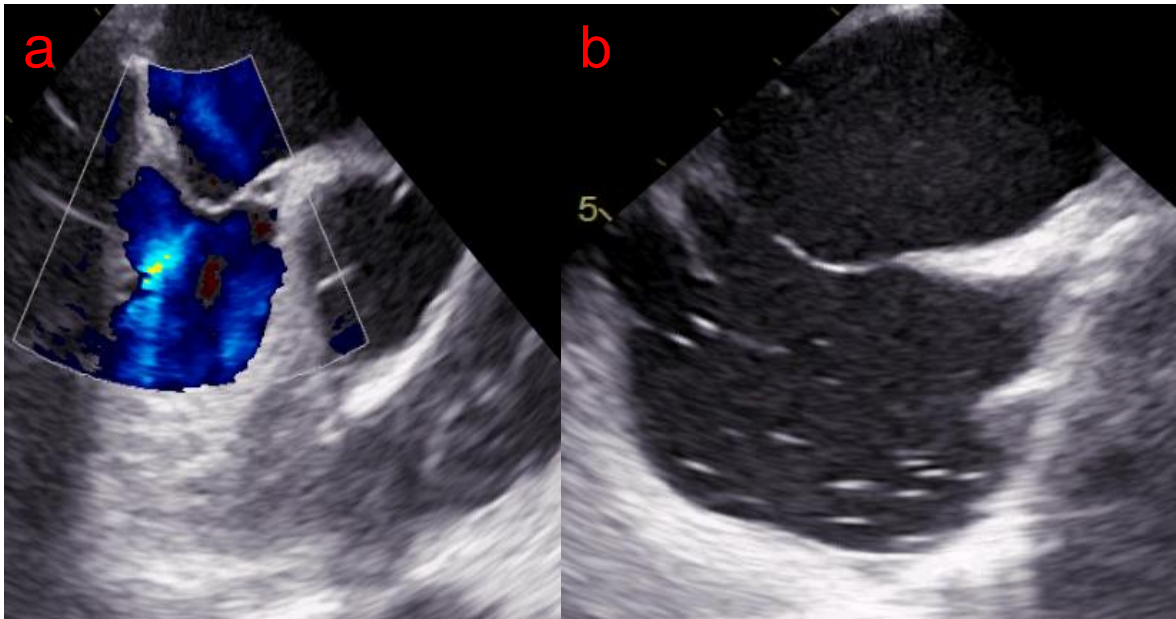


Figure 6: **a-**TOE angled 45° centered on the pulmonary valve:showing a mobile pedunculated hyperechoic image attached to the pulmonary valve measuring 9mm in length on the arterial side associated with pulmonary valve stenosis, **b-** TOE angled 120° centered on the inter-atrial septum:positive bubble test confirming the presence of a large ASD.

The biological assessment objectified a slight microcytic hypochromic anemia at 10g/dl, with a disturbed infectious assessment;hyperleukocytosis at 11890/ μ l predominantly polynuclear, C reactive protein (CRP) raised at 161mg/l and positive procalcitonin at 1.1ng/l.

At this stage, the diagnosis of isolated PVE without tricuspid involvement on an undiagnosed congenital heart disease was retained. The blood cultures carried out were positive for a gram-positive cocci bacterium, *Abiotrophiadefectiva* sensitive to a large spectrum of antibiotics.

An endocarditis extension assessment was carried out including an immunology assessment (RF, Ac Anti DNA, etc.) viral serologies (HBV, HCV, HIV, TPHA/VDRL) and body scan, which was without abnormalities.

The patient was put on dual antibiotics therapy based on vancomycin 30mg/kg/day for 4 weeks and gentamycin 1mg/kg/day for 2 weeks. The surgical indication of ASD closure, removal of the vegetation and valvulotomy was indicated.

A good clinical evolution was marked in the patient after 4 weeks of dual antibiotics therapy with negative blood culture assessment control but without regression of the size of the vegetation. The patient underwent a surgical operation after the end of medical therapy for ASD closure, removal of the vegetation and valvulotomy without valve prosthesis (Figure 7). The postoperative follow-up was without complications.



Figure 7: Surgical specimen of a resected pulmonary valve: Destroyed pulmonary valve leaflet and vegetation.

Discussion

“Right-sided endocarditis comprises less than 5% of all endocarditis; blood-borne infections travelling from the right atrium to the pulmonary artery have to pass through the tricuspid valve, resulting in tricuspid colonization. Therefore, involvement of only the pulmonary valve without tricuspid valve endocarditis is a rare occurrence, accounting only for 1.5–2% of all reported cases of endocarditis” [4]. Risk factors for the disease include intravenous drug use, central venous catheters, cyanotic congenital heart disease, and degenerative valve lesions [5]. In our case, an undiagnosed adult congenital heart disease (severe pulmonary valve stenosis and a large atrial septal defect (ASD) was discovered concomitant with the PVE which explained the clinical manifestation of acute respiratory syndrome presented by the patient.

A study carried out by Isabella Zwiener and al. on 3840 cases of infective endocarditis (IE), 1.6% involved PV: 30 native IE and 30 prosthetic IE. The mean age was significantly higher in patients with native as opposed to prosthetic IE, and they were also more frequently male. Patients with native IE had more extracardiac comorbidities, with a higher age-adjusted Charlson score than patients with prosthetic valve IE. They also received immunosuppressive treatment more frequently and had a higher prevalence of intravenous drug abuse [6]. In our case, the subject is a female and had an isolated PVE on a native valve with no medical history.

In a prospective cohort study, the main pathogenic microorganism isolated from blood culture was gram's bacteria (83%), of which ***Staphylococcus aureus*** accounted for 31% [7]. The most common pathogenic microorganism in North America is ***Staphylococcus aureus*** [7,8], which is consistent with the patient's history and blood culture. In this case a gram cocci bacterium ***Abiotrophia defectiva*** was isolated which can be present in nasopharyngeal, digestive or genital flora. The entry point for IE in our patient was due to poor dental hygiene.

As the AHA (American Heart Association) guidelines [9] and European society of cardiology 2023 guidelines [10] recommend, both TTE (transthoracic echocardiography) and TOE (transesophageal echocardiography) are indispensable in many patients with IE during initial evaluation and subsequent follow-up, and they provide complementary information. It is estimated that the sensitivity and specificity

of TTE are 30–63% and 91–100%, respectively, and those of TOE are 87–100% and 91–100% respectively [11]. Additional information was obtained after TOE in patients especially the dimension of the vegetation, its extension to the trunk and branches of the pulmonary arteries and anatomy of the pulmonary artery. Cardiac CT can also show the full spectrum of right-sided endocarditis cardiopulmonary features including manifestations that cannot be demonstrated by echocardiography [12]. In our patient, diagnosis was confirmed by TTE and TOE without the need for Cardiac CT scan.

The AHA guidelines [9] recommend that right-sided IE should be treated as conservatively as possible, and nonrandomized trial data from a single center experience [13] and international collaboration [14] support that early valve surgery may not be beneficial for all primary patients with primary IE caused by *Staphylococcus aureus*. In this case due to severe pulmonary valve stenosis, large ASD and septic emboli, surgery was absolutely necessary. Even the new ESC guidelines on IE did not explain the role of surgery in isolated pulmonary endocarditis [10]. Witten JC et al. found in a 13-year retrospective study of right-sided IE [8] if surgery was performed at an early stage, the surgical risk was low. In total, right-sided endocarditis has a better prognosis than left-sided endocarditis [15]. Given the 15-year single-center experience from Liekiene D et al. [16], removal of vegetation by preserving the valve is the most beneficial at the early stage of PVE [16, 17, 18, 19]. In our case, valvulotomy and vegetectomy was done to treat pulmonary valve stenosis as well as the removal of the vegetation.

The postoperative results were favorable and patient was educated on the risk of recurrence and preventive measures such as dental health was also emphasized at discharge, as stipulated by the new ESC guidelines 2023 [10].

Conclusion

In summary, our patient presented a septic embolic in conjunction with PVE due to *Abiotrophia defectiva* on an undiagnosed congenital heart disease (severe pulmonary valve stenosis and ASD). This highlights that right-sided IE is rare and transthoracic echocardiography should be done carefully in order to avoid misdiagnosis due to the location of the pulmonary valves. Surgery should be considered in case of associated congenital heart disease. Clinicians should advise patients on preventive measures to avoid possible recurrence.

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

Consent

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

Abbreviations

BDRV: Basal diameter of the right ventricle

Gmax: Maximum gradient

HBV: Hepatitis B HCV: Hepatitis C

HIV: Human Immunodeficiency Virus
NYHA: New York Heart Association dyspnoea stages
PWRV: Posterior wall of the right ventricle
RF: Rheumatoid factor
TDDL: Tele diastolic diameter of the left ventricle
TPHA/VDRL: Blood markers for Syphilis

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