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 discover of a congenital heart disease in a young adult subject. Unlike other cases of right sided endocarditis, we treated our patient both medical and surgically.

Case presentation: The patient was diagnosed with pulmonary valve endocarditis with blood cultures showing *Abiotrophia defective,* a germ difficult to cultivate and echocardiographic revealing a mass measuring 8mm long 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: Pulmonary valve endocarditis, *Abiotrophia defectiva*, transthoracic echocardiography, pulmonary valve stenosis, Atrial septal defect, valvulotomy.

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 microbiologic features with the more common tricuspid endocarditis [3]. Risk factors for developing PVE include intravenous drug abuse, indwelling catheters and prosthetic valves. It is a challenging condition to diagnose mainly because of nonspecific signs and symptoms at presentation. Common clues for suspecting PVE include a new-onset 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 on an undiagnosed congenital heart disease reveled 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 Report

We report the case of a 35-year-old young woman with no personal or family history 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, the chest computed tomography scan (CT scan) of which showed organized pneumonitis with low-abundance pericardial effusion. Faced with an episode of febrile peak at 38.5°C, the patient was transferred to the cardiology department for suspected endocarditis.

Somatic examination finds a stable patient with a blood pressure (BP) of 120mmHg systolic pressure and 70mmHg diastolic pressure, tachycardia at 112 beats/minutes, fever at 38.3°C, 98% saturated in ambient air. The cardiovascular examination is normal, while the pleuropulmonary examination finds bronchial rales in the lower right lung with pleuritic pain on the same side. Cardiac auscultation found a systolic-diastolic murmur at the pulmonary focus and the rest of the examination in search of the portal of entry was unremarkable except for an altered dental 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. (Figure 1).

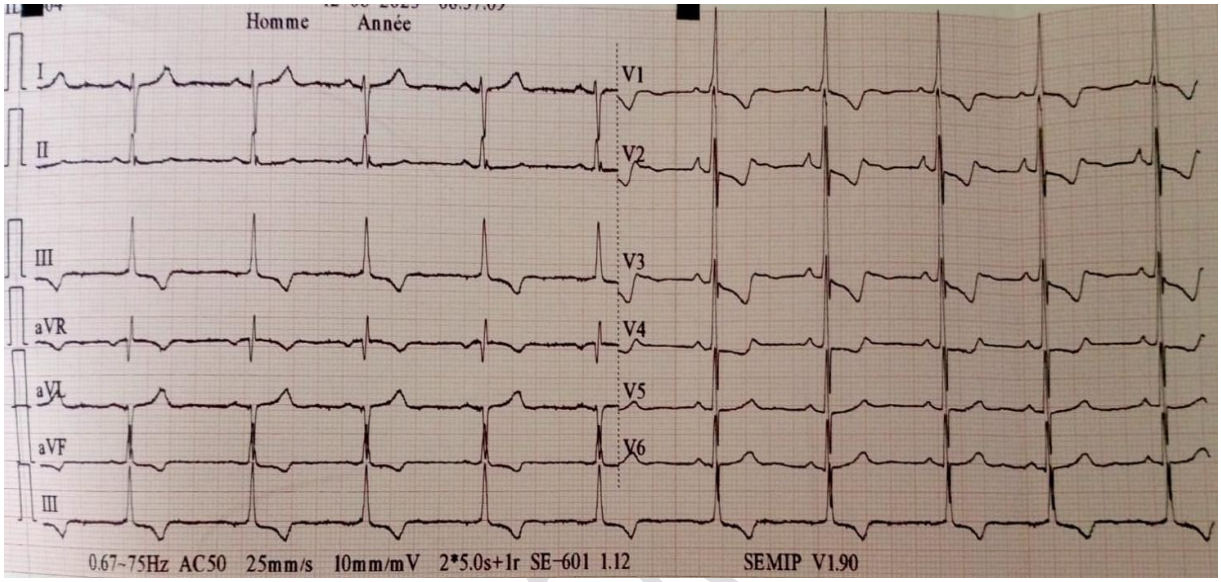

Figure 1: Electrocardiogram (ECG): showing a regular sinus rhythm, right axis deviation associated with hypertrophic right ventricle and negative T waves at the inferior leads

In front of this clinical scenario, the thoracic angioscan was made which revealed a right pulmonary embolism with a pulmonary infarction and pulmonary nodule and micronodule of non-specific appearance (Figure 2).

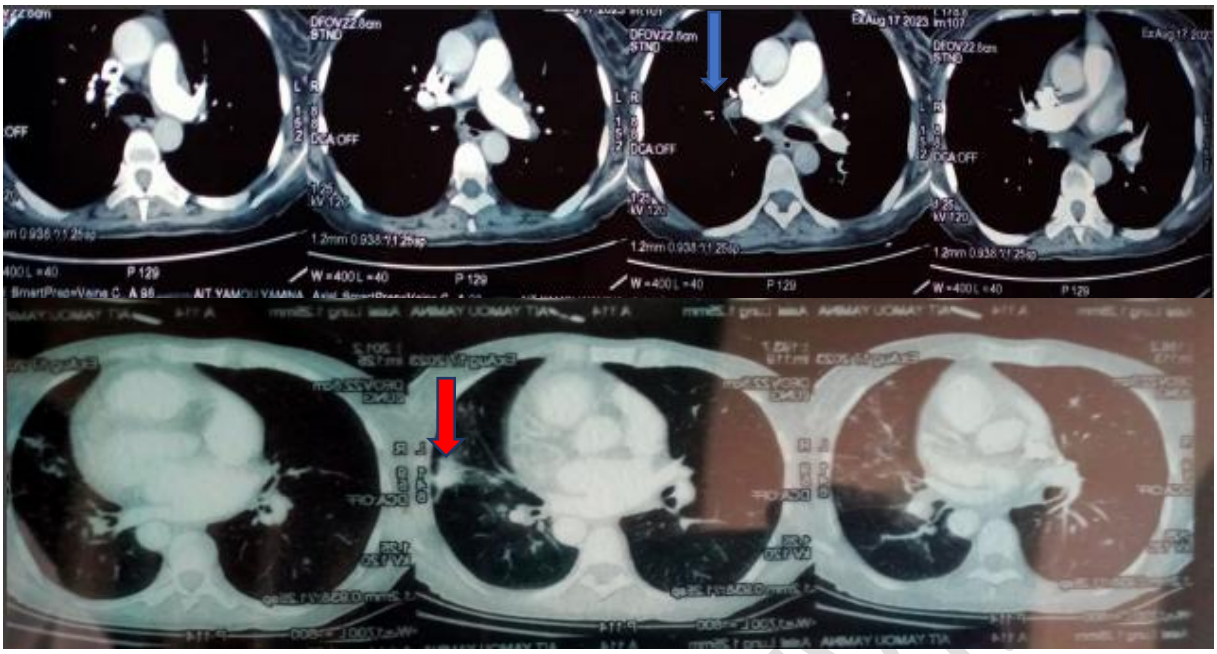

Figure 2: Thoracic angioscan: parenchymal and skeletal view: showing a right pulmonary septic emboli (blue arrow) complicated with a pulmonary infarction (red arrow) and pulmonary nodule and micronodule of non-specific appearance.

The transthoracic echocardiography (TTE) showed a non-dilated left ventricle (TDDLV: 40mm indexed at 26mm/m2), non-hypertrophic with good global contractility and segmental, left ventricle ejection fraction (LVEF) at 60%, dilation of the right atrium with a surface area of 27.8cm2. Absence of mitral and aortic valves abnormalities with dilated right ventricle (RV) (BDRV: 42mm), hypertrophic (PWRV: 8mm) with good longitudinal systolic function (S'RV: 16cm/s and TAPSE: 23mm) (figure 3). Presence of severe pulmonary valvular stenosis (Gmax: 152mmHg, VmaxP: 6.3cm/s) with presence of vegetation at the level of the pulmonary valve measuring 8mm at the level of its arterial version (Figure 4). We also note the presence of a large atrial septal defect (ASD), ostium secundum type (ASD measuring 39mm) with a bidirectional shunt (Figure 5). Initial aorta of normal caliber, non-dilated and compliant inferior vena cava without signs of pulmonary venous return, pericardial detachment facing the right cavities.

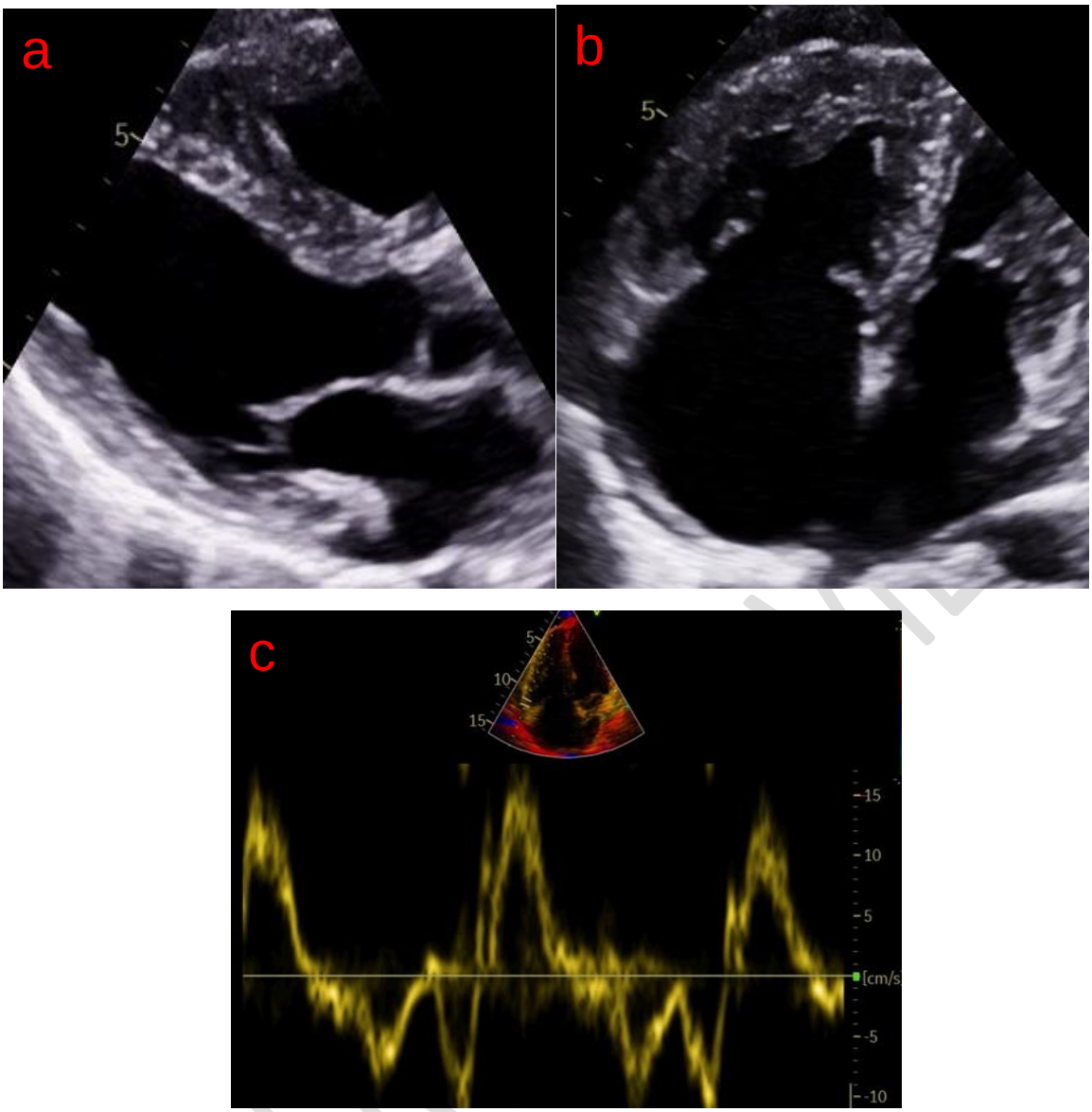

Figure 3: Transthoracic echocardiography (TTE): a-long axis view: non-dilated and non-hypertrophic left ventricle (TDDLV: 40mm indexed at 26mm/m2), b-4 chambers apical view: dilation of the right atrium with a surface area of 27.8cm2 with dilated right ventricle (RV) and hypertrophy of the posterior 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.

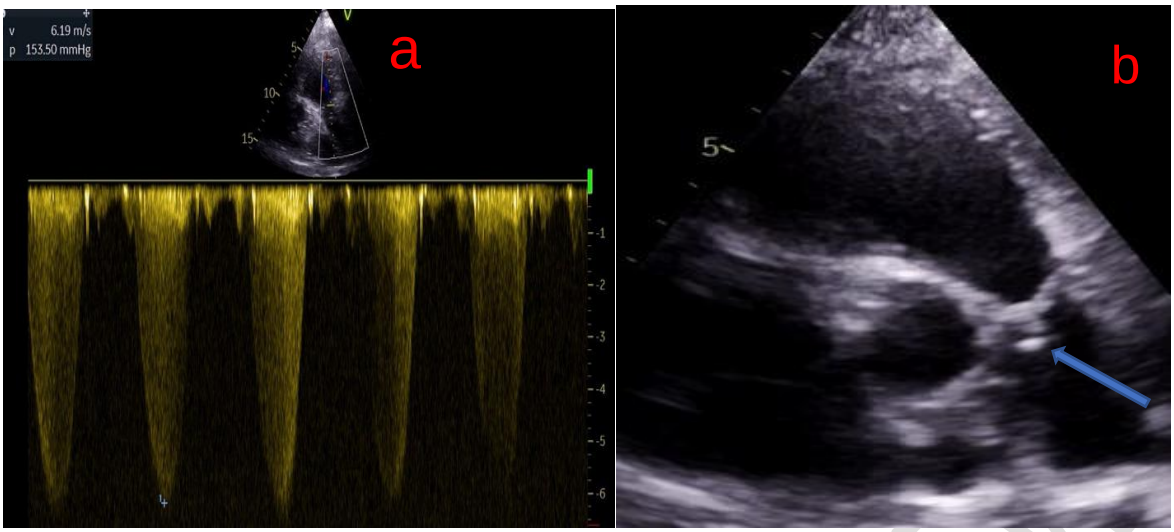

Figure 4 :TTE :short axis view centered on the pulmonary valve: **a**-Severe pulmonary valvular stenosis (Gmax: 152mmHg, VmaxP: 6.3cm/s), **b**- with presence of vegetation at the level of the pulmonary valve measuring 8mm at the level of its arterial version (blue arrow)

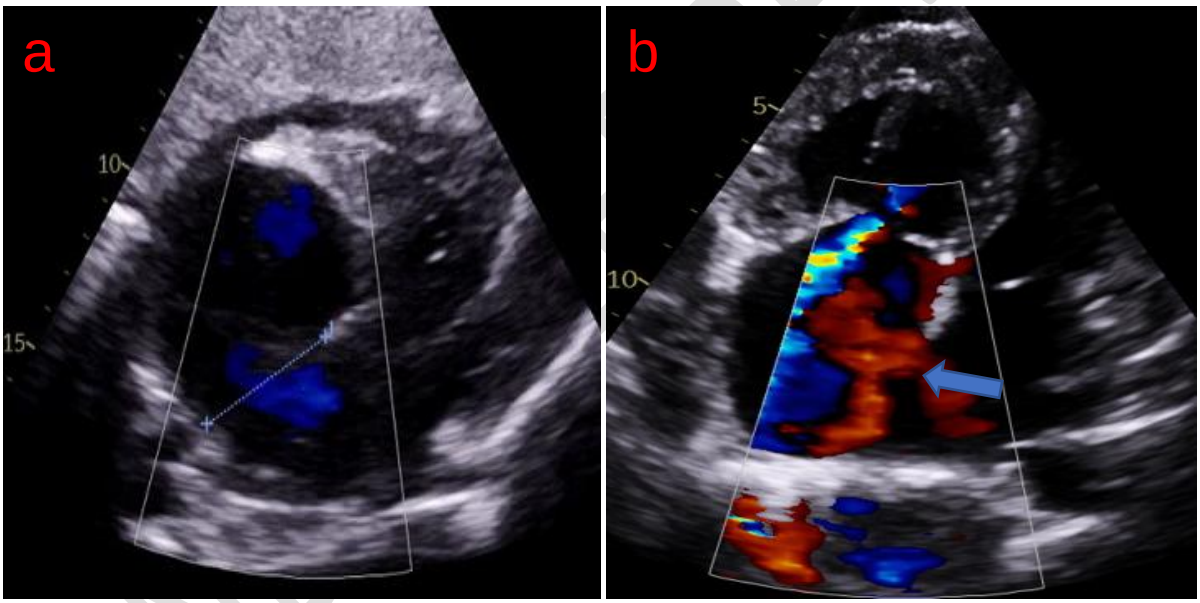

Figure 5 : a- under costal view showing a large atrial septal defect ostium secundum type measuring 39mm, **b**-4 chambers view showing a bidirectional shunt caused by the ASD

A transesophageal ultrasound was done to better visualize the vegetation objectifying 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 with the presence of 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.

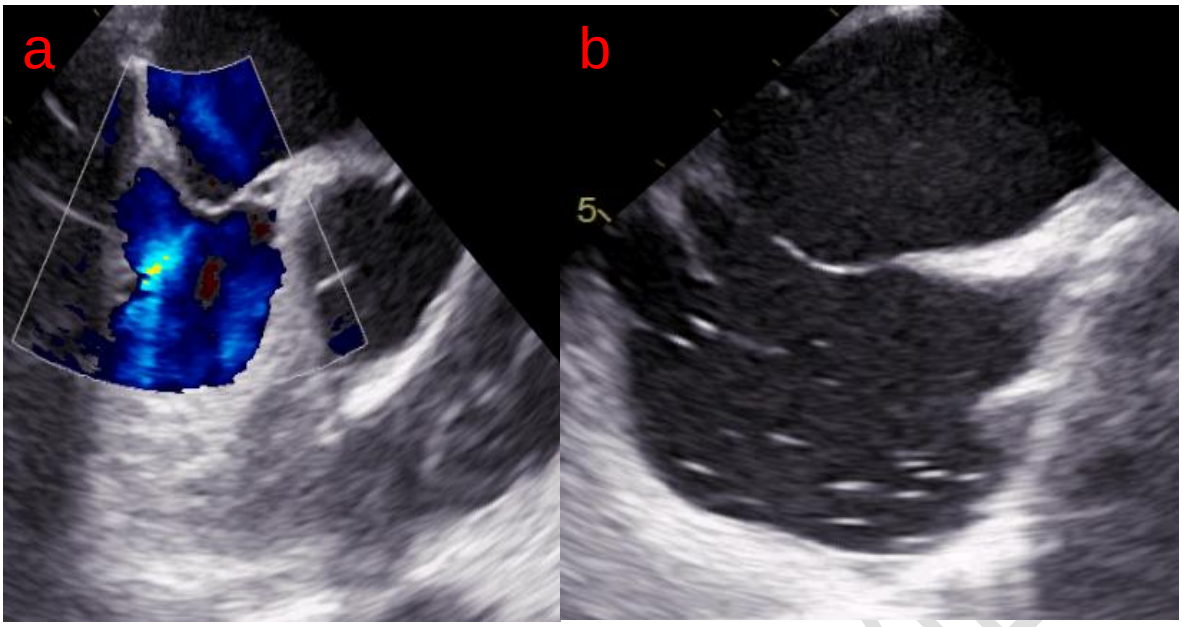

Figure 6: a-view centered on the pulmonary valve: mobile pedunculated hyperechoic image attached to the pulmonary valve measuring 9mm in length on the arterial side associated with pulmonary valve stenosis, **b**-view centered on the inter-atrial septum: positive bubble test with the presence of large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

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

In view of the clinical, ultrasound and biological findings, infective endocarditis with isolated involvement of the pulmonary valve on undiagnosed congenital heart disease was retained, of which the blood cultures carried out proved positive by the presence of a gram-positive cocci bacterium***, Abiotrophia defectiva*** multi type sensitive to antibiotics.

The endocarditis extension assessment was completed by an immunology assessment (RF, Ac Anti DNA, etc.) and viral serologies (HBV, HCV, HIV, TPHA/VDRL) and body scan which turned out to normal.

The patient is put on dual antibiotic therapy based on vancomycin 30mg/kg/day for 4 weeks and gentamycin 1mg/kg/day for 2 weeks. The surgical indication was posed on the patient for a closure of the ASD, removal of the vegetation and valvulotomy in the first stage.

A good clinical evolution was marked in the patient after 4 weeks of dual antibiotic therapy with a negative control blood culture but without regression of the size of the vegetation. The patient was later operated by closure of ASD, removal of the vegetation and valvulotomy without valve prosthesis (Figure 7). The postoperative follow-up was without complications.

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 a 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 a 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 TEE (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 TEE 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 IPE [16, 17,18]. In our case, valvulotomy and vegectomie 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 with emphasis on dental health 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 mise diagnosis 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.

Abbreviations

TDDLV: Tele diastolic diameter of the left ventricle
BDRV: Basal diameter of the right ventricle
PWRV: Posterior wall of the right ventricle
RF: Rheumatoid factor
HBV: Hepatitis B HCV: Hepatitis C
HIV: Human Immunodeficiency Virus
TPHA/VDRL: Blood markers for Syphilis

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