

Untreated Tetralogy of Fallot in an Adult Patient Complicated by Acute Pulmonary Valve Endocarditis

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Authors' contributions

This work was carried out in collaboration among all authors. Author HM designed the manuscript paper, wrote the first draft of the manuscript, and managed the literature searches. Author KM, helped drafting the manuscript and managed the literature searches. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

We report the case of a 18 years old male, who presented to the emergency department for prolonged fever, weight loss, with a systolic murmur, echocardiography showed a tetralogy of fallot with multiple vegetations on the pulmonary ejection pathway and pericardial effusion. The diagnosis of certain endocarditis was retained with one major Duke criterion (image of vegetation on the pulmonary pathway) and 3 minor Duke criteria (fever at 39°C, glomerulonephritis and predisposing congenital heart disease). All blood cultures were negative and PCR for mycobacterium tuberculosis was positive in the pericardial liquid. The patient was treated by intravenous antibiotics associating Ceftriaxone and Gentamycin associated to anti-tuberculosis treatment according to the Moroccan national protocol with good evolution.

Keywords: *Infective endocarditis; pulmonary valve endocarditis; tetralogy of fallot; congenital heart disease; complication.*

ABBREVIATIONS

ASD : Atrial septal defect
IE : Infective endocarditis
HIV : Human immunodeficiency virus
PCR : Polymerase Chain Reaction
PDA : Patent ductus arteriosus
PVE : Pulmonary valve endocarditis
TOF : Tetralogy of fallot
VSD : Ventricular septal defect

1. INTRODUCTION

Pulmonary valve endocarditis (PVE) is an extremely uncommon clinical finding comprising less than 1.5-2% of cases for infective endocarditis, even in patients with uncorrected congenital heart disease. "PVE frequency has declined dramatically over the last thirty years "[1]. On the other hand, prosthetic pulmonary valve or conduit infective endocarditis (IE) prevalence is increasing, as is the prevalence of repaired congenital heart disease [2]. "It is a challenging condition to diagnose mainly because of nonspecific signs and symptoms at presentation" [3]. Tetralogy of Fallot (TOF) is the most common cyanotic disease susceptible to PVE [4]. IE usually occur in patients known for their congenital heart disease. Rarely, it can be the first manifestation to reveal an underlying congenital heart disease.

We present the case of a 18 years old patient with prolonged fever caused by a pulmonary IE complicating an unknown and untreated TOF.

2. CASE PRESENTATION

We present the case of an 18 years old young, who presented to the emergency department for fever, aggravating dyspnea and cough, evolving two weeks before. He also had a history of anorexia, weakness, fatigue and weight loss. The patient didn't have any history of drug use or any other comorbidities and the patient didn't have any antibiotics before his presentation.

On clinical examination: The patient had stable hemodynamics with an arterial pressure at 110/75 mmHg and a heart rate at 100 ppm. His oxygen saturation on room air was at 80% and he was feverish with a temperature of 39.5° C. At general examination, he had a cyanosis of the lips and extremities with digital bilateral hypocratism. At cardiac auscultation, there was a left parasternal systolic murmur with a 3/6

intensity as well as a systolo-diastolic murmur at the pulmonary valve auscultation point with a 4/6 intensity. There were slight edemas of the lower limbs with hepato-jugular reflux and jugular veins distention.

Pulmonary auscultation was normal as well as neurologic examination. There were no palpable adenopathies at the cervical, suprasternal and axillary level.

The Chest x ray showed a cardiomegaly with minimal right pulmonary effusion and no obvious lesion in the lung parenchyma (Fig.1). Transthoracic echocardiography revealed a tetralogy of fallot with an overriding aorta with a sub-aortic ventricular septal defect (VSD) measuring 18 mm with a right-to-left systolic shunt and a maximal gradient at 25 mmHg (Fig. 2). The right ventricle was hypertrophied with good systolic function with a tricuspid annular plane systolic excursion (TAPSE) at 21 mm and a tricuspid annulus plane peak systolic velocity (S' wave) at 12cm/s . The RV outflow tract was seat of an infundibular stenosis caused by anterior displacement of the conal septum with a maximal gradient at 45 mmHg. The echocardiography revealed multiple vegetations on the trunk of the pulmonary artery as well as on both of its branches, the biggest vegetation was located on the pulmonary valve measuring 16x12mm it was mobile following the movement of the pulmonary leaflets. It was responsible for a destruction of the pulmonary valve with Severe pulmonary regurgitation without causing pulmonary obstruction. Both right and left pulmonary artery had multiples vegetations (Fig. 3). The pulmonary valve ring had a normal size measuring 19 mm. Pulmonary branches were also of normal size without any obstructions, the right pulmonary artery (RPA) measured 10 mm and the left pulmonary artery (LPA) measured 12 mm.

There was no vegetations located in the RV, the tricuspid, mitral or aortic valves. There was no other VSD nor atrial septal defect (ASD), nor patent ductus arteriosus (PDA). The aortic root was slightly dilated with a moderate central leakage of the aortic valve. The inferior vena cava was dilated at 23 mm and the pericardium was seat of a moderate circumferential effusion measured at 17 mm.

In laboratory investigations, the hemogram revealed an inflammatory microcytic hypochrom anemia with hemoglobin at 9.7 g/dl, hematocrit at

32.4%, an elevated white blood cell count at 12000/mm³. The creatinine level was elevated at 162umol/l (18.4mg/l), the proteinuria was positive at 0.7 g/24h indicating glomerulonephritis and blood culture were all negative for aerobic, anaerobic and fungal micro-organisms, the culture was negative even for slowly growing atypical organisms. By referring to the modified Duke criteria, the diagnosis of certain endocarditis was retained with one major criterion (image of vegetation on the pulmonary pathway) and 3 minor criteria (fever at 39°C,

glomerulonephritis and predisposing congenital heart disease). The patient was treated by intravenous antibiotics associating Ceftriaxone and Gentamycin adjusted to weight and renal function.

No pulmonary nor systemic embolism, nor deep infectious source were detected in the Body CT scan. Serologic tests to hepatitis B, hepatitis C, human immunodeficiency virus (HIV) and syphilis were negative. oral and otolaryngological examination were normal.



Fig. 1. Chest X ray showing a cardiomegaly with a minimal right pleural effusion
**the red sign indicates the right pleural effusion*

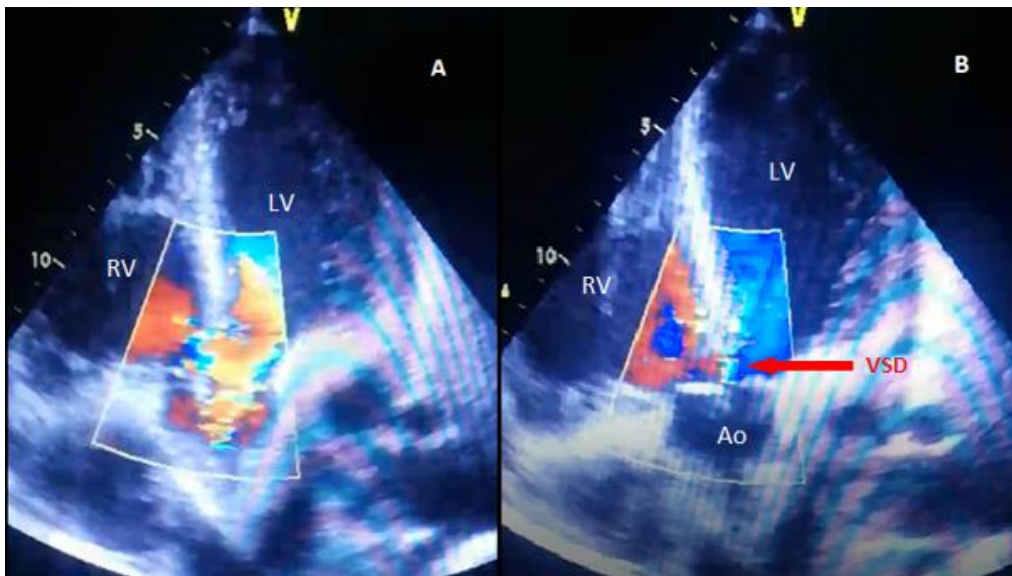


Fig. 2. Transthoracic echocardiography Apical 5 chambers view (A) During systole (B) during diastole, showing an overriding aorta over a sub-aortic ventricular septal defect of 18mm with a right-to-left shunt during systole, with dilated and hypertrophied right ventricle. Aortic root is slightly dilated with a moderate central aortic valve leakage

**LV: Left ventricle; RV: Right ventricle ; Ao : Aorta ; VSD: ventricular septal defect*

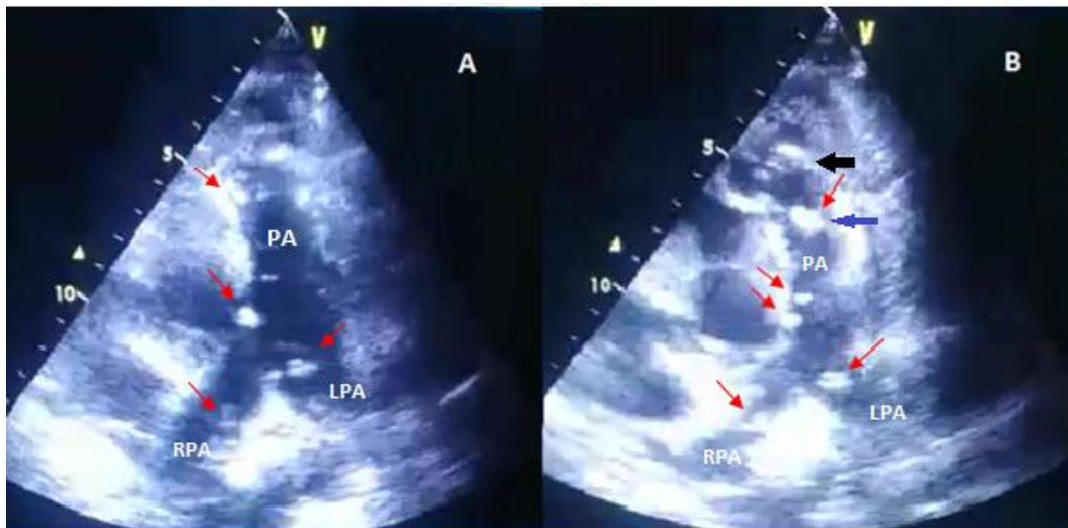


Fig. 3. Transthoracic echocardiography short axis view centered on the pulmonary artery and its division (A) During systole (B) during diastole, showing multiples non obstructive vegetations (red signs) along the pulmonary artery and both right and left pulmonary branches. The biggest vegetation (blue sign) is located on the pulmonary valve measuring 16x12 mm mobile with pulmonary valve movement with a severe regurgitation of the pulmonary valve without obstruction. The black sign indicate the infundibular pulmonary stenosis caused by anterior displacement of conal septum

*PA: pulmonary artery; RPA: Right pulmonary artery, LPA: Left pulmonary artery

In the absence of an identified microorganism in blood cultures, a puncture of the pericardial liquid was carried out with analysis of pericardial puncture fluid showed a lymphocyte predominance (76%) with a protein level of 68g (exudative liquid), the bacterial culture was negative. a Polymerase Chain Reaction (PCR) for mycobacterium Tuberculosis was realized because of suspicion of tuberculosis and returned positive. After these results the patient was put on anti-tuberculosis treatment according to the Moroccan national protocol with 2 months under four anti-tuberculosis agents (Rifampicin, Isoniazid, Pyrazinamide and Ethambutol) followed by two months under two antituberculosis agents (Rifampicin and Isoniazid). Corticosteroid therapy (aiming to reduce the risk of chronic constrictive pericarditis) was not given because of the risk of septic aggravation of endocarditis.

A good clinical evolution was observed afterwards with a disappearance of fever, an improvement in biological parameters of infection and renal function. Regression of the pericardial effusion and a stabilization of the pulmonary vegetations were observed in echocardiographic control. The patient was discharged and kept under anti-tuberculosis treatment. A reassessment after 6 to 9 months of treatment

will be made of his tuberculosis status before proposing a surgical cure for his congenital heart disease.

3. DISCUSSION

Pulmonic valve endocarditis is a rare clinical phenomenon [5]. TOF is the most common complex/ cyanotic congenital heart disease and often requires prosthetic valve implantation. "Patients with TOF are considered at high risk of IE as a result of valvular remodeling (jet impact) with altered hemodynamics in untreated patients and multiple invasive procedures, including pulmonary valve replacement (PVR) in operated patients" [6]. Population-based risk of endocarditis in TOF patients is unknown [7]. "Recent studies have reported increased risk of endocarditis in patients with congenital heart disease compared with the general population, with higher risk in the patients with complex congenital heart lesions and those with prosthetic valves" [8].

"The variety of clinical manifestations and complications, as well as the serious prognosis, makes it mandatory that IE patients need to be treated in experienced hospitals with a collaborative approach between different specialists, involving cardiologists, infectious

disease specialists, microbiologists, surgeons, and frequently others, including neurologists and radiologists” [9].

“Infective endocarditis should be considered in anyone with sepsis of unknown origin, or fever in the presence of risk factors including congenital heart disease” [10]. “The principal predisposing factors for pulmonic valve infective endocarditis has been recognized in adult patients underwent the intravenous medication, central venous catheters, alcoholism, chronic hemodialysis, hepatic transplantation, celiac disease, intravenous drug addicts, congenital heart disease and systemic infection with bacterial agents” [11].

Besides usual IE manifestations, PVE can be responsible for septic pulmonary embolism and pulmonary infections caused by the migration and embolization of pulmonary vegetations [12].

Eighty to 90% of positive blood culture IEs are caused by streptococci, enterococci, and staphylococci. Gram-negative bacilli from the HACEK group (Haemophilus, Actinobacillus, Cardiobacterium, Eikenella, Kingella) account for 5% to 10% of cases of IE. Other Gram-negative and Gram-positive organisms (corinebacterium, Listeria monocytogenes, lactobacilos, Erysipelothrix rhusiopathiae, Bacillus spp) are rare causes of IE. Fungi (mainly Candida spp) represents less than 1% of IE. “Other infectious agents causing IE can be culture negative and require serologic testing, cell culture, or gene amplification for their identification (Coxiella burnetii, Tropheryma whippelii, or Rochalimaea spp) [9]. In our patient all realized blood cultures were negative. PCR for mycobacterium tuberculosis was positive and our patient was diagnosed with pericardial tuberculosis. But is the mycobacterium tuberculosis the micro-organism responsible for the PVE ?. “Valvular endocarditis caused by M. Tuberculosis is extremely rare, and is reported only in the context of miliary tuberculosis. The diagnosis must be made by histology to confirm the causal link. Cope et al. reported a case of disseminated tuberculosis with documented aortic IE which resolved on anti-tuberculosis therapy” [13]. In our case, the patient was treated with standard intravenous antibiotics for 4 weeks besides oral anti-tuberculosis therapy.

4. CONCLUSION

The number of adult patients with untreated TOF has nowadays become a rarity in the well-

developed health systems, however, in developing health systems, the number of people with untreated TOF in adolescents and young adults is still high and thus we still see adult patients presenting with untreated TOF and infective endocarditis. Therefore, we need a new awareness for these patients who will need appropriate therapy for IE and surgical care of TOF when it is possible.

DISCLAIMER

The products used for this research are commonly and predominantly use products in our area of research and country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

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, patients' written consent has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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