Case report

Double outlet right ventricle infective endocarditis: a rare combination and a therapeutic challenge

ABSTRACT

Infective endocarditis involving the right side of the heart occurs rarely and often involves the tricuspid valve. The isolated pulmonary valve infective endocarditis (IPVIE) is a less common condition that occurs in specific population. The double outlet right ventricle (DORV) is an unusual congenital heart disease. The association of DORV and IPVIE darkens the prognosis. We report two cases of the association of DORV and IPVIE. The transthoracic echocardiography (TTE) is the base to the diagnosis. Right sided infective endocarditis in the lack of a guided strategy remains a therapeutic challenge.

Keywords: double outlet right ventricle, infective endocarditis, isolated pulmonary valve infective endocarditis, *pyopneumothorax, pulmonary embolism, empyema.*

1. INTRODUCTION

Double outlet right ventricle (DORV) is an uncommon and complex congenital malformation with many anatomic variations and a poor prognosis [1], [2]. The Infective endocarditis of the right side of the heart in its pulmonary form is even rarer with an incidence of 02% according to literature [3]. We report two rare cases of two patients with untreated DORV who survived until teenage years, but later developed pulmonary valve infective endocarditis.

2. PRESENTATION OF CASES

CASE REPORT 1:

Our patient was an eighteen-year-old male who was diagnosed in childhood with DORV and atrioventricular heart block. He had not undergone corrective nor palliative surgery. He had initially a decreased effort tolerance in the past year. Two months prior to admission, he presented fever with an upper respiratory infection that was treated with poor medication compliance. The fever recurred two weeks after treatment with cough and shortness of breath. The patient was therefore put on antibiotics with no improvement. At his admission, the patient had deteriorated acutely. He presented with acute chest pain and breathlessness, cough with purulent sputum, fever, and asthenia. Following admission, he had a respiratory distress.
On examination, the patient was cyanosed. His blood pressure was 102/49 mmHg, his heart rate at 126 beat per minute and his respiratory rate at 46 breaths per minute. The lung examination found hypersonority on percussion of the left side and bilateral decrease in vesicular murmur, interfering with cardiac auscultation. The patient had signs of right heart failure: edema of the lower limbs reaching the ankles and turgidity of the jugular veins. The patient had hepatosplenomegaly. No facial dysmorphism was noted.

Chest computed tomography (CT) scan of the patient revealed multiple empyema with left pyopneumothorax with purulent mediastinal collection and foci of pulmonary condensation (Fig. 1) without signs of pulmonary embolism.

An urgent strategy of thoracic drainage was realized and 4500 mL of purulent liquid were collected in total.

Microbiology studies were done in conjunction with empiric synergic dual antibiotic: ceftriaxone 02g/day and gentamicine 3mg/kg/day while waiting for the antibiogram. Biology tests found: microbiology of the purulent liquid: white blood cell count of a 1000 with neutrophils predominance, and revealed *pseudomona aeruginosa* resistant to ceftazidim.

Gene xpert was negative and no mycobacterium tuberculosis was found on the cultures. The total white blood cell count was 26 × 103/L (90% neutrophils). C-reactive protein (CRP) level was 200 mg/L and erythrocyte sedimentation rate (ESR) was 76 mm/hr. Two of the five sets of blood cultures were positive with *staphylococcus aureus* resistant to methicillin. Serology test were negatives.

Two-dimensional transthoracic echocardiography (TTE) showed DORV with large sub-aortic ventricular septal defect (VSD) and severe pulmonary stenosis, as well as Large vegetations, noted on the pulmonary valve measuring 27 mm × 19 mm, moderate mitral valve stenosis and a dilated post stenotic pulmonary artery, were associated with dilated unique right ventricle and hypoplasic left ventricle with preserved systolic function (Fig. 2, 3). This confirmed the diagnosis of an active infective endocarditis complicating a necrotizing pneumopathy.

Also, the antibiotic prescription was modified later in link with the antibiogram results and the patient has received a combination of ceftriaxone and gentamicine for fifteen days, followed by twenty-seven days of ceftriaxone. As a total of 42 days antibiotic regimen. The patient presented a clinical improvement with a dried drain, biological improvement: (CRP: 200 to 27 mg/L) and white blood cell count of 6580 elements/mm^3 and a stationary cardiac lesion with declining in size of the pulmonary vegetation. The chest CT scan control showed a marked decline of empyema and no pneumothorax and a decline of pulmonary condensation foci. We decided the ablation of the thoracic drain and to continue 10 days regimen of fluoroquinolone: levofloxacine. Surgery was suggested to the patient after full recovery of the infection. Unfortunately, the patient was lost of sight during the covid and post covid period and consulted later for an intolerance to effort. The TTE showed an aspect of enormous residual vegetation of the pulmonary valve with a dilatation of the pulmonary artery. A bidirectional Glenn procedure was proposed to our patient after an updated chest CT scan.

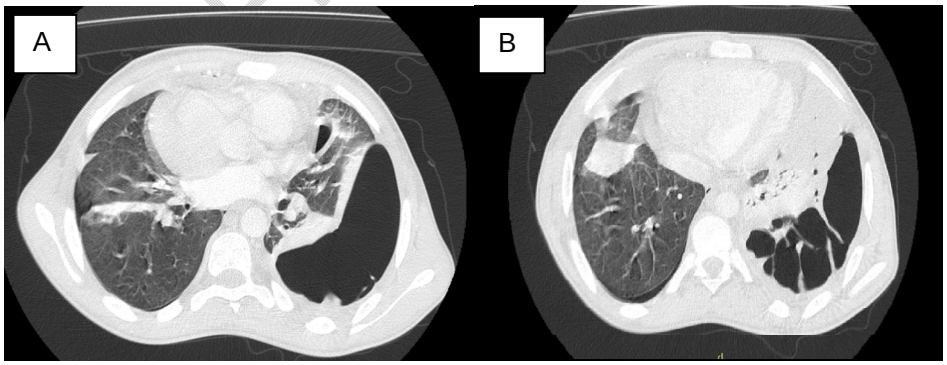

Fig. 1. (A)+(B) Chest CT scan images of pulmonary empyema and markedly enlarged right side heart.

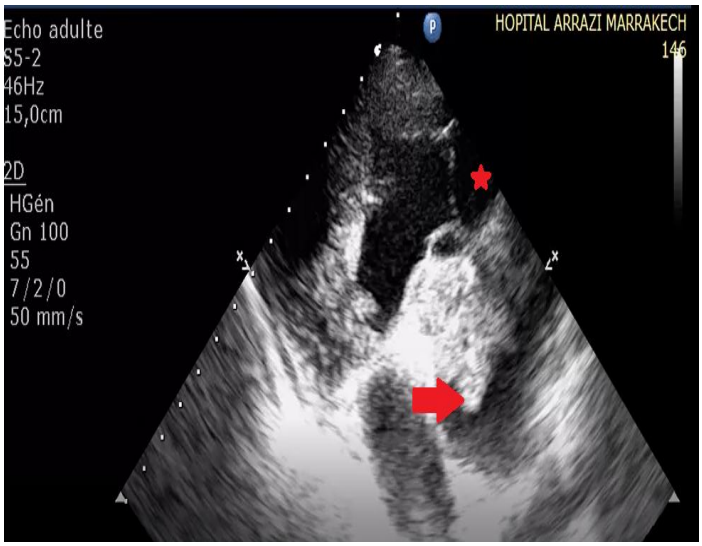

Fig. 2. A 05-chamber view of echocardiography showing:
Red arrow: image of pulmonary artery and huge pulmonary valve vegetation ; red star: both the aorta and pulmonary artery are emerging from the right ventricle.

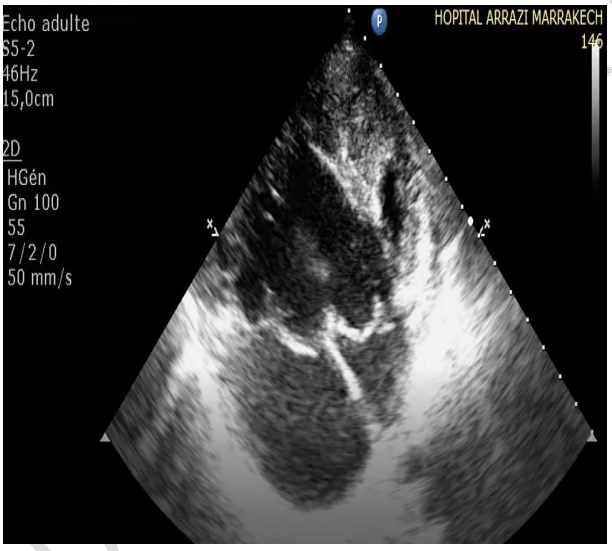

Fig. 3. A 4-chamber view of echocardiography showing: a unique right ventricle with a large ventricular septal defect measuring 27 mm and hypoplasic left ventricle.

CASE REPORT 2:

Our second case is about a twenty-seven-year-old non consanguineous man with a history of sudden death in sibling, who was diagnosed in childhood with DORV. He had not undergone corrective or palliative surgery. He presented to the emergency for poly-arthralgia, palpitations, breathlessness, asthenia, and fever.
At his admission the physical examination found. A blood pressure at 115/58 mmHg, irregular heart rate at 145 beats per minute, a respiratory rate at 26 breaths per minute, SpO2 at 89%, temperature at 38.7°C. The cardiac auscultation found wheel radius

systolic murmur examination. No signs of right nor left heart failure. The patient had hepatosplenomegaly. No facial dysmorphism was noted.
Two-dimensional TTE showed DORV with large sub-aortic ventricular septal defect (VSD) and severe pulmonary stenosis, as well as large vegetations on the pulmonary valve measuring 20 mm × 17 mm, resulting into a severe pulmonary regurgitation and stenosis, and a dilated post stenotic pulmonary artery, were associated with dilated unique right ventricle and hypoplasic left ventricle with preserved systolic function (Fig. 4, 5). Electrocardiogram showed atrial fibrillation.
Microbiology studies were done in conjunction with empiric synergic dual antibiotic: ceftriaxone 02g/day and gentamicine 3mg/kg /day; while waiting for the antibiogram.
Two of the three sets of blood cultures were positive of staphylococcus aureus.
The total white blood cell count was 10 × 10^3/L (90% neutrophils), C-reactive protein (CRP) level was 60 mg/L and erythrocyte sedimentation rate was 56 mm/hr.
Proteinuria was negative and no embolic events were noted.
This confirmed the diagnosis of an active infective endocarditis.
Under the antibiotic regimen the patient presented a clinical improvement with no fever over 15 days and also a biological improvement: (CRP: 60 to 24 mg/L) and white blood cell count at 7506 ele/mm3. The patient is still in follow-up and surgery is to be planned after.

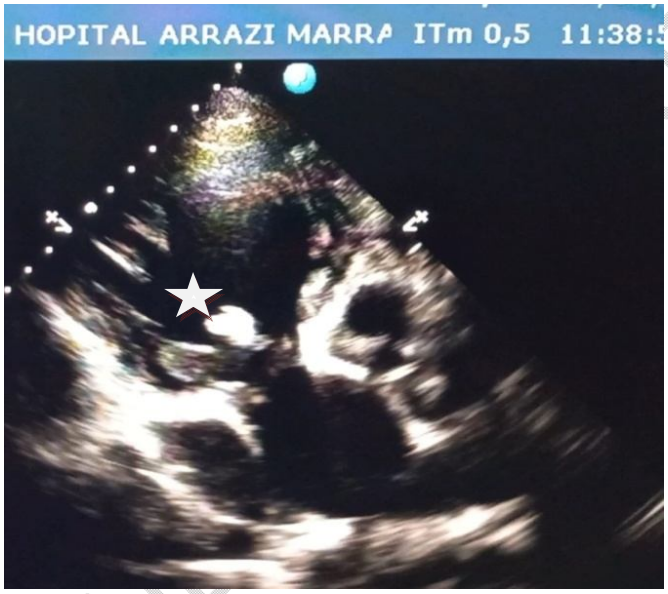

Fig. 4. Echocardiography of 05 chamber view showing: white star: image of dilated pulmonary artery and pulmonary valve vegetation, and both the aorta and pulmonary artery are emerging from the right ventricle.

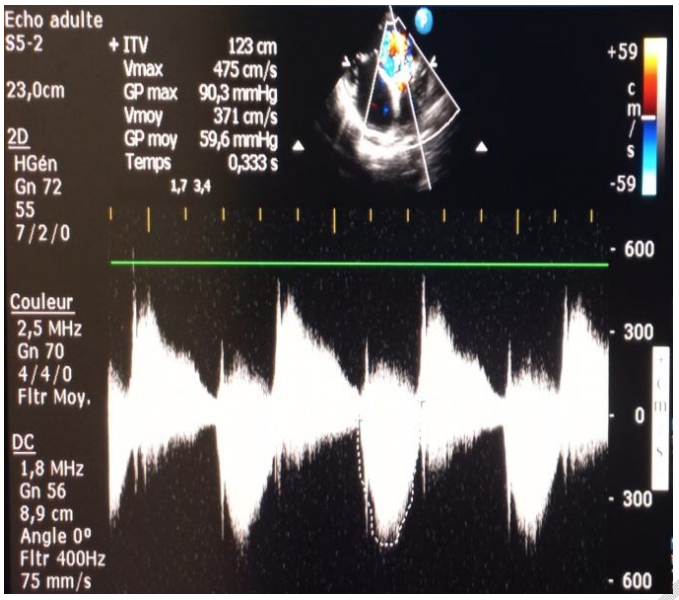

Fig. 5. Echocardiography of continuous Doppler on pulmonary artery image of a severe valvular pulmonary stenosis gradient max 90.3 mmHg and severe pulmonary regurgitation.

3. DISCUSSION

DORV is a congenital cardiovascular malformation in which both great arteries arise entirely or predominantly from the right ventricle [4]. DORV is a rare condition with poor prognosis. DROV as congenital heart disease is considered a risk factor of right sided infective endocarditis [7]. PVIE is an uncommon condition with an incidence of 02% of right sided infective endocarditis. The survival of the patients presenting with DROV at one month age is around 25% and only few patients reach adulthood [6]. Association of both entities darkens the prognosis [5].

Patients present typically with a respiratory distress and pulmonary embolism septic or cruoric in PIVE. However, due to the large VSD in DROV, systemic embolic events might occur especially cerebral stroke with severe life-threatening complications [2], [8]. As a result, the invasiveness of right sided infective endocarditis lies in the other clinical pictures that a patient may present: septic shock, obstructive shock compromising the hemodynamics of the patient with a massive pulmonary embolism [9]. The diagnosis is based on duke criteria. However, the microbiology profile of right sided infective endocarditis is different from the left sided endocarditis with a predominance of *staphylococcus aureus* in 90% and an increasing prevalence of pseudomonas aeruginosa with fungal microorganisms to be associated to a higher mortality, and that is related to our cases [10].

In all infective endocarditis, the antibiotic regimen is compulsory, must be dual to triple antibiotic therapy, bactericide and synergic, initially empiric but later adapted to the antibiogram [7].

There is a gap in the management of infective endocarditis of the right heart, particularly the pulmonary valve, in the guidelines compared to the left sided endocarditis. However, in the recent literature, indications of surgery were established and we cite: persistent bacteremia for more than 07 days despite adequate antibiotic as recurrent pulmonary embolism with or without right heart failure on the tricuspid valve, an enormous vegetation > 20mm, right heart failure secondary to a severe regurgitation and abscess [10].

The surgery in a simpler case would focus on the vegetation removal debridement of the valve and a repair or bioprotheses of the pulmonary valve. Other authors would suggest

percutaneous angio-aspiration of the vegetation prior to surgery, in order to reduce the embolic events. Metanalysis concluded to the safety and efficiency of the AngioVac devices, but the use of the technique must be determined on a case-by-case basis, in the collaboration of a heart team [11].
The association of an advanced not corrected DORV till adulthood with PVIE in our case added more complexity to the surgery decision. As in our cases, the patients present with severe cyanosis with decreased pulmonary blood flow, the bidirectional Glenn shunt seems to be an option for our patient as described in the European Society of Cardiology (ESC) guidelines of congenital heart disease [7].
The emphasis of the new guidelines from the American Heart Association (AHA), and European society of congenital heart disease focused on the maintenance of oral and cutaneous hygiene and prophylactic antibiotics especially in cyanotic congenital heart disease in procedures with infectious risks, as it is believed that this group is at high risk of developing infective endocarditis. Moreover, the majority of infective endocarditis cases could be avoided in special population when hygiene measurements are respected [7], [12]).

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