Pulmonary valve infective endocarditis: A case series

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ABSTRACT

Background and Objectives : **Infective endocarditis (IE) involving the native pulmonary valve (PV) is extremely rare, with no data in Indian literature. The objective of this communication is to describe the clinical and diagnostic characteristics, underlying risk factors, microbiological features, and management of PVIE.**

Methods : **This is a retrospective analysis of 8 cases of PVIE managed in a tertiary care center from 1992 to 2020.**

Results : **PVIE was observed in 8 patients with underlying congenital cardiac malformation (Group A, 6 Patients) and in patients with central venous catheter (Group B, 2 patients). All the patients had prolonged febrile illness accompanied by right heart failure 4 (50%), septic pulmonary emboli 2 (25%), and pulmonary regurgitation 3 (37.5%). Trans-thoracic echocardiography demonstrated the vegetations, whereas computed tomography of chest diagnosed pulmonary emboli in 2 (25%), and pulmonary artery aneurysm in 1 (12.5%) patient. The early mortality was extremely high (5, 62.5%). Delayed diagnosis, fulminant septicemia, and multi-organ failure resulted in unfavorable outcomes.**

Conclusions : **IE of the native PV is a rare and potentially lethal illness. Diagnosis should be considered in any febrile patient with an underlying congenital defect, central venous line, bacteremia, and comorbidities. Multi-modality imaging should be utilized to enhance the diagnostic yield and detect complications promptly.**

Keywords : Infective endocarditis, mycotic pulmonary artery aneurysm, native pulmonary valve endocarditis, pulmonary valve

INTRODUCTION

Right heart infective endocarditis (IE) mostly involving tricuspid valve accounted for only 5%–10% of cases in the recent European Society of Cardiology data.[1] Endocarditis affecting the native pulmonary valve (PV) is extremely rare and ranges between 1% and 2%.[2,3] The infective process may be isolated to PV or concomitantly involve other valves.[4-6]

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There are reports of PVIE from India in international journals.[7-10] The aim of this study is to report the clinical features, management, and outcome data of 8 patients of native PVIE seen over the last 28 years.

METHODS

The records of consecutive cases of PVIE seen in a tertiary care teaching institution form the material of this study.

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From October 1992 to October 2020, 8 cases of PVIE were diagnosed, treated, and followed up. Diagnosis of PVIE was guided by modified Dukes criteria.[11] Patients were categorized on the basis of predisposing factors. Group 1 consisted of patients with an underlying congenital cardiac malformation (CCM) (n = 6) and Group 2 consisted of IE related to an indwelling central venous catheter (CVC) (n = 2).

Clinical, investigative, management, and follow-up information was analyzed. Relevant laboratory investigations and blood cultures were studied. Electrocardiogram, Skiagram chest, echocardiography, high-resolution computed tomography (HRCT) of chest,[3] pulmonary angiography,[1] and management data were analyzed.

RESULTS

Table 1 summarizes the clinical, investigative, and outcome data. Group 1 includes 6 patients (all male) aged 4–22 years with an underlying CCM. All had prolonged (6–16 weeks) febrile illness and had received multiple antibiotics prior to referral to our center. Case 1 had features of congestive cardiac failure (CCF) and pulmonary regurgitation (PR). This patient was considered too sick for surgery and died due to septicemia. Patient 2 had staphylococcus endocarditis and responded favorably to antibiotics and underwent a successful balloon PV dilatation 6 months after the index episode. The clinical picture of patient 3 was complicated by septic pulmonary emboli (SPE). Two-dimensional echocardiography (2DE) in short axis view revealed vegetations attached to the PV. The vegetations were protruding into the pulmonary artery (PA) and suggested a flail valve [Figure 1a and b]. The family was unwilling for surgery due to guarded prognosis. Case 4 was transferred to the pediatric intensive care unit with febrile illness of 4 months duration unresponsive to anti-tubercular treatment and multiple antibiotics. Physical examination of this sick, cachexic child (weight 8 kg) revealed anemia, tachypnea, tachycardia, ejection click, and a grade 3/6 ejection systolic murmur along the left sternal border. Multiple blood cultures and fungal cultures were negative. Skiagram of the chest posterior-anterior view revealed a homogenous shadow occupying the right lung field [Figure 2a]. This shadow has remained unaltered in several X-rays. HRCT of the chest revealed a large enhancing vascular structure with surrounding low-density thrombus in relation to the right PA (RPA). This structure extended upon the right paracardiac region and measured approximately 6.1 cm × 5.0 cm [Figure 2b]. A 2DE with Doppler interrogation revealed thickened and mildly stenotic valve with a trans-PV gradient of 24 mm of Hg. Diagnosis of PVIE with mycotic aneurysm of PA was entertained.

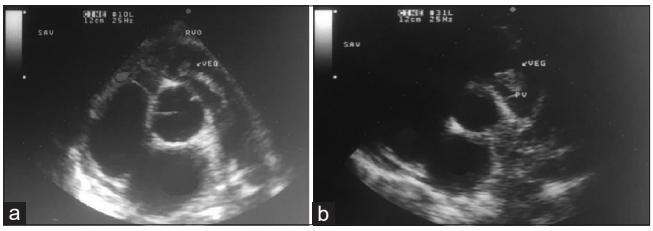

Figure 1: (a and b) Two-dimensional echocardiography short axis view shows vegetation (veg) on pulmonary valve. The excessive mobility into RVO and PA raise the possibility of a flail leaflet. AO: Aorta, RA: Right atrium, LA: Left atrium, TV: Tricuspid valve, RVO: Right ventricular outflow, PA: Pulmonary artery

Table 1: Clinical, echocardiographic, microbiological data and outcome

Age (year), sex	Underlying CCM/Risk factor	2DECHO	Infecting organism	Outcome
		Group 1		
17, male	TOF	Mobile mass on PV, PR, TR	Polymicrobial organisms	Died of septicemia
12, male	PVS	Mobile mass on PV	*S. aureus*	Improved on antibiotics, balloon dilation of PV after 6 months
18, male	VSD and PVS	Mobile mass on PV, protruding in PA (flail PV), PR	Culture negative	Patient unwilling for surgery (died)
4, male	PVS	Thickening of PV	Culture negative	Rupture of pseudoaneurysm and death
7, male	VSD	Thickened PV, vegetations	*Streptococcus viridans*	Closure of VSD and pulmonary valvectomy (alive)
22, male	TOF	Mobile mass on PV, mild PR	*S. aureus*	Intracardiac repair (VSD-patch and RVOT - patch repair), alive
		Group 2		
56, female	CKD, DM, CVC	Mobile mass on PV, protruding in PA, free PR	*Klebsiella, C. albicans*	Considered too sick for surgery (died)
45, female	RA, DM, CVC	Mobile vegetations on PV	*Pseudomonas, Candida*	Considered too sick for surgery (died)

CCM: Congenital cardiac malformation, CVC: Central venous catheter, DM: Diabetes mellitus, PR: Pulmonary regurgitations, PV: Pulmonary valve, PVS: PV stenosis, RA: Rheumatoid arthritis, RVOT: Right ventricular outflow tract, TOF: Tetralogy of Fallot, TR: Tricuspid regurgitation, VSD: Ventricular septal defect, 2DECHO: Two-dimensional echocardiography, CKD: Chronic kidney disease, PA: Pulmonary artery, *S. aureus: Staphylococcus aureus, C. albicans: Candida albicans, S. viridans: Streptococcus viridans*

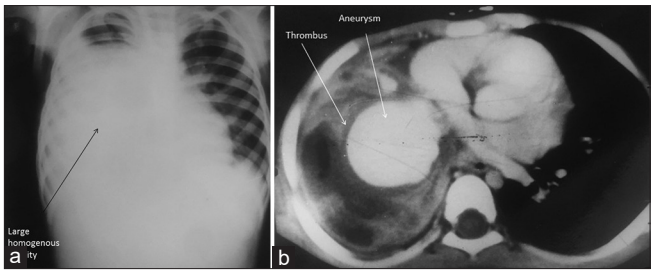

Figure 2: Skiagram chest pulmonary artery view shows homogenous shadow occupying large portion of right lung (Panel a). High resolution computed tomography of chest revealed a large (6.1 cm × 5.0 cm) enhancing vascular structure with surrounding low density thrombus in relation to right pulmonary artery interpreted as aneurysm (Panel b)

Pressure data (mm of Hg) at cardiac catheterization revealed: Aorta 100/60 mean 85, left ventricle 100/8, main, left, and RPA 20/8, right ventricle (RV) 50/6, trans PV gradient 30 and right atrium (RA) mean 6. Selective RPA angiography revealed a large round aneurysm with marked stasis of contrast occupying the entire right hemi-thorax. A narrow neck communicated between middle lobe branch of RPA and the aneurysm. Cardiothoracic surgery was refused due to moribund condition of the child. Partial percutaneous closure was achieved by multiple coils however, further attempts to close the residual aneurysm proved fatal.

Cases 5 and 6 presented with a febrile illness with blood cultures showing streptococcus viridans and *Staphylococcus aureus*. Both underwent successful surgical procedures after medical stabilization.

Patient 2 was doing well up to 5 years follow up, patients 5 and 6 followed up only for 2 years.

Group 2 includes two female patients aged 45 and 56 years. These patients were extremely sick and displayed features of fulminant septicemia. Both had CVC inserted for different reasons. Additional risk factors included diabetes mellitus, chronic kidney disease, rheumatoid arthritis, and history of immunosuppressive therapy [Table 1]. Both the patients succumbed to multi-organ failure secondary to fungal and Gram-negative septicemia.

DISCUSSION

IE of native PV is rare. There are reports published by Indian authors in international journals.[7-10] Our experience of diagnosing and managing 8 cases of PVIE during the last 28 years is reported. There has been continued progress in the diagnostic abilities and the management of IE over the years. However, the recognition of this entity remains a challenge due to its rarity and varying clinical presentation. In an autopsy series from India, PV involvement was observed in 5.6% of cases.[10]

Symptoms and signs of infection are invariably present. Most had prolonged febrile illness and have received at least 2–3 weeks of antibiotics prior to referral to a tertiary care. The dominant etiology in this series is underlying CCM and included tetralogy of Fallot, ventricular septal defect, and PV stenosis [Table 1]. Almost all the variety of congenital lesions can be complicated by PVIE.[4-6,8] As reported previously, the diagnosis of PVIE should be entertained in the presence of fever, SPE, PR with or without CCF.[8] Heart failure in 4 (50%), SPE in 2 (25%) and murmur of PR in 3 (37.5%) supported the diagnosis. Signs of right-sided CCF depend on degree of PR which occurs as a sequel to the valve destruction. The most challenging presentation was of a 4-year-old moribund child (case 4) seen 2 decades back and was referred as persistent consolidation despite multiple rounds of antibiotics and a course of ATT. The pediatric resident recorded a murmur and sought a cardiac evaluation. Multiple chest X-rays showed a persistent homogenous opacity which was misinterpreted as nonresolving consolidation. Echocardiography revealed a stenotic PV with vegetation and HRCT of chest provided a clue to aneurysm of RPA. PA aneurysm (PAA) is a rare vascular anomaly which can occur due to a variety of conditions.[12] Mycotic PAA has been described after right heart endocarditis and is attributed to septic emboli or a jet hitting the wall of the PA as in patent ductus arteriosus.[13,14] The management is challenging as they are inherently unstable and can rupture. It is also important to achieve eradication of infection by intensive antibiotic therapy. Definitive removal or obliteration of the aneurysm is best achieved by surgical procedures such as aneurysmectomy, aneurysmorrhaphy, banding, or lobectomy.[13,14] These aneurysms have been treated by detachable coils[15] and stent-graft.[16] Percutaneous closure was preferred as surgery was considered very high risk; however, the child succumbed to rupture of the aneurysm. Modern-day hardware and experience certainly promise a better outcome.

During the last 5 years, we encountered two middle-aged patients presenting with fulminant septicemia. One had severe PR and other multiple SPE which were diagnosed as recurrent pneumonia. Both had CVC inserted for dialysis and pharmacotherapy. A wide spectrum of risk factors ranging from intravenous drug addiction, cardiac implantable electronic devices (CIED), central lines, immunosuppressive therapy, and several health care associated procedures have been associated with PVIE.[17-19] These cases highlight the need to recognize PV endocarditis in presence of CVC and extra-cardiac comorbidities. Infection of PV can occur even in individuals without any known illness.[20] There are fewer patients with CCM being encountered in recent PVIE series and this change is attributed to early surgical treatment of these defects.[18,19] On the contrary, there is an increase in conduit and prosthetic PV endocarditis.[19]

The Duke's criteria utilize clinical, echocardiographic, and blood culture findings for the diagnosis of IE.[11] The applicability of these criteria in PVIE is unproven. Anemia, leukocytosis, and elevation of inflammatory markers though nonspecific raise a suspicion of IE in the presence of underlying risk factors. The yield of blood cultures has improved due to refined techniques and organisms could be isolated in six out of eight cases (75%). A favorable outcome was observed in patients who grew either staphylococcus (25%), or streptococcus viridans (12.5%). The remaining with poly-microbial (12.5%), Gram-negative and fungal (25%), and culture-negative (25%) endocarditis did not survive. Fungal, poly-microbial, and culture-negative endocarditis pose diagnostic challenges and a poor prognosis.[21,22]

TTE remains a key modality for the diagnosis. Vegetations cause thickening of valve, shaggy/fluffy echoes, or mobile mass [Table 1 and Figure 1]. PV is extremely difficult to be visualized especially in adults, and it is quite likely that endocarditis involving this valve is missed in real-world scenario. M-mode and 2-DE identification of PV vegetations have been well reported.[7,23] Parasternal short-axis view is the most useful and along with parasternal long axis, RV inflow and subcostal views provide diagnosis in 87%–91%.[18] Better visualization of valve and Doppler assessment of PR is possible on TTE than trans-esophageal (TEE) with experienced operators. TEE of the anteriorly located PV can be challenging and is superior in evaluating CIED lead involvement.[18]

It is suggested that adjunctive imaging techniques are utilized to complement the information derived from the echocardiography. HRCT diagnosed SPE in two patients (25%) and PAA in one patient (12.5%) in this study. This modality diagnoses pulmonary complications (SPE, abscess, infarction, effusion) and large (>10 mm, mobile) vegetations which usually cause embolization.[24,25] CT angiography and cardiac magnetic resonance imaging provide high-resolution images and can help in the diagnosis of pseudo-aneurysm, fistulas, valve perforation, abscess and valvular dehiscence.[26] 18F-fluorodeoxyglucose positron emission tomography/(PET/CT) has been utilized for diagnosis of right-sided endocarditis.[27,28]

High mortality was reported in an earlier communication over 3 decades ago.[8] Unfortunately, the mortality trends remain unchanged in this study, despite better diagnostic and therapeutic facilities. This may be partly attributable to the inclusion of cases of group 2 who succumbed to septicemia. The cases referred to tertiary care are often too sick to be considered for surgery. Only three patients (37.5%) (2 with surgical and one with initial conservative treatment) survived. High mortality ranging from 20% to 66% has been observed.[19] Early surgery should be considered in the presence of highly mobile vegetations, recurrent SPE, and failure of intensive medical therapy.[29]

PVIE is a rare entity. A high index of clinical suspicion is needed for an early diagnosis. If the diagnosis is delayed, the outcome may be less than satisfactory.

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Conflicts of interest

There are no conflicts of interest.

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