

Perforated cuspal aneurysm of aortic valve following infective endocarditis presenting as complete heart block: a case report and review of literature

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Received 6 March 2022; first decision 20 April 2022; accepted 22 April 2022; online publish-ahead-of-print 27 April 2022

Aortic cuspal aneurysm is a rare clinical entity and often occurs as a complication of infective endocarditis. We report a case of a 30-year-old male with no prior comorbid conditions who presented with fever, acute onset shortness of breath, and chest pain along with multiple episodes of syncope. Electrocardiogram revealed complete heart block while two-dimensional echocardiogram was suggestive of perforated aortic cuspal aneurysm with aortic regurgitation. Blood cultures were positive for *Streptococcus viridans*. The patient was initiated on broad spectrum antibiotics, temporary pacemaker implantation, and subsequently underwent aortic valve replacement followed by permanent pacemaker implantation after 6 weeks. A diagnosis of perforated aortic cuspal aneurysm subsequent to infective endocarditis was made. This was based on clinical presentation, echocardiographic evaluation, blood cultures, and surgical as well as histopathological findings.

Keywords

Aortic valve • Infective endocarditis • Heart block • Perforation • Aortic regurgitation

Learning points

- Aortic cuspal aneurysms are rare clinical entity.
- Most of the aortic cuspal aneurysms occur secondary to infective endocarditis.
- Clinical presentation is that of acute aortic regurgitation and treatment is surgical aortic valve replacement.

Introduction

Cardiac valvular aneurysm is a rare clinical entity with the mitral valve being more commonly involved than the aortic.¹ Most of these valvular aneurysms are due to the complication of infective endocarditis.

Other possible aetiologies include rheumatoid arthritis and Bechet's disease.^{2,3} The clinical presentation of aortic valvular aneurysm is often that of acute heart failure. We present a young patient with complete heart block (CHB) and perforated aortic cuspal aneurysm subsequent to infective endocarditis.

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Handling Editor: Amardeep Ghosh Dastidar

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Timeline

Time	Events
1 month prior	<ul style="list-style-type: none"> Fever with chills
2 days prior	<ul style="list-style-type: none"> Sudden onset retrosternal chest pain, tearing type with profuse diaphoresis Progressive shortness of breath (NYHA Classes III and IV) Multiple episode of syncope Fever
Day 1	<ul style="list-style-type: none"> Fever: temperature of 38.5°C Blood counts revealed neutrophilic leucocytosis (total leucocyte count: 17 000/mm³) Raised erythrocyte sedimentation rate (45 mm/h) and high-sensitivity C-reactive protein (11.2 mg/dL). Positive procalcitonin Electrocardiogram revealed complete heart block with infra-Hisian ventricular escape rhythm. Temporary pacemaker was inserted via right femoral vein approach. Transthoracic echocardiogram: <ul style="list-style-type: none"> Dilated left ventricle (LV) with normal LV ejection fraction—60%, saccular aneurysmal sac arising from the non-coronary cusp (NCC) of aortic valve (AV) and prolapsing into the left ventricular outflow tract (LVOT) without aortic root dilatation, two defects at the apex of aneurysmal sac were noted through which the regurgitant colour jet was appreciated from aorta to LVOT. Serial blood cultures were collected to identify the organism and its sensitivity to antimicrobials. Provisional diagnosis of infective endocarditis (IE) was made and empirical antimicrobials were initiated. Inotropic infusion was given as patient was in shock.
Day 2	<ul style="list-style-type: none"> Transoesophageal echocardiography showed prolapse of the NCC that resembled a wreath bobbing chaotically into the LVOT during diastole and into the aortic root during systole. Jet of severe aortic regurgitation entered the LV through a defect on the NCC.
Day 3	<ul style="list-style-type: none"> Blood culture revealed growth of typical IE organism, i.e. <i>Streptococcus viridans</i> which was sensitive to penicillin and ceftriaxone IE diagnosis was confirmed and patient was stabilized with antibiotics and inotropes.
At 2 weeks	<ul style="list-style-type: none"> Surgical AV replacement with metallic prosthesis
At 3 weeks	<ul style="list-style-type: none"> Histopathological examination of the excised AV revealed microscopically, thick fragments of hyalinized collagenous tissue with foci of myxoid degeneration, occasional foci of calcification and varying infiltrates of lymphocytes/plasma cells without evidence of infection or granulomatous lesions. Culture of the excised valvular tissue grew <i>Streptococcus viridians</i>.
At 6 weeks	<ul style="list-style-type: none"> Completed antibiotics regime Complete heart block persistent Permanent pacemaker implanted on left pectoral side
Follow-up 1 year	<ul style="list-style-type: none"> Asymptomatic Compliant to anticoagulant Normal aortic prosthesis function Normal pacemaker interrogation

Case report

A 30-year-old male presented to the emergency department with complaints of sudden onset retrosternal chest pain which was tearing in nature along with profuse diaphoresis 2 days back. Following this episode of chest pain, he developed progressive shortness of breath along with orthopnoea and paroxysmal nocturnal dyspnoea. Additionally, he also had multiple episodes of syncope since the past 2 days. Prior to presentation, he had a history of fever with chills for the 1 month. On presentation, he had a toxic appearance with cold and clammy extremities. Physical examination revealed the presence of purplish-pink tender, nodules in the finger pads of both hands suggestive of Osler nodes (Figure 1A). His heart rate

was 36 b.p.m., blood pressure was 70/20 mmHg, temperature of 38.5°C, and a respiratory rate of 36 per minute. Cardiac examination revealed a Grade III/VI diastolic regurgitant blowing murmur at the fourth intercostal space on the left sternal border along with bilateral basal inspiratory crepitations. Routine blood investigations revealed neutrophilic leucocytosis (total leucocyte count: 17 000/mm³). Liver and renal function tests were within normal limits. He had a raised erythrocyte sedimentation rate (45 mm/h) and C-reactive protein (11.2 mg/dL) along with positive procalcitonin. Thyroid function tests were normal while a vasculitis profile was negative. Electrocardiogram on presentation revealed CHB with infra-Hisian ventricular escape rhythm (Figure 1B). Chest radiography was suggestive of cardiomegaly with left ventricle (LV) type of apex.

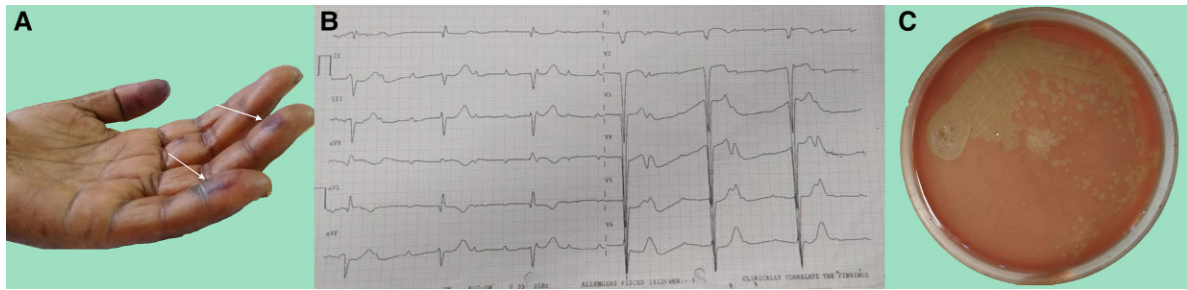


Figure 1 (A) Figure showing the presence of purplish-pink tender, nodules in the finger pads of both hands suggestive of Osler nodes (white arrows). (B) A 12-lead electrocardiogram performed on admission suggestive of complete heart block with infra-Hisian ventricular escape rhythm (ventricular rate: 30 per minute). (C) Figure showing the alpha haemolytic colonies of *Streptococcus viridans* on blood agar.

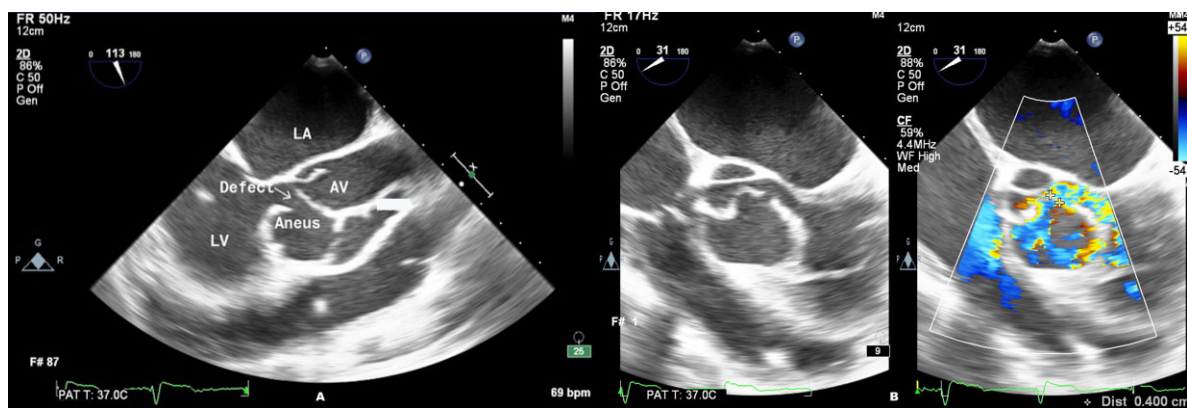


Figure 2 (A) Transoesophageal echocardiogram in the mid-oesophageal aortic valve long axis view showing the aneurysm of non-coronary cusp projecting into left ventricular outflow tract. (B) Transoesophageal echocardiogram in the mid-oesophageal aortic valve short axis view showing the defect at apex of the cuspal aneurysm with regurgitant aortic jet.

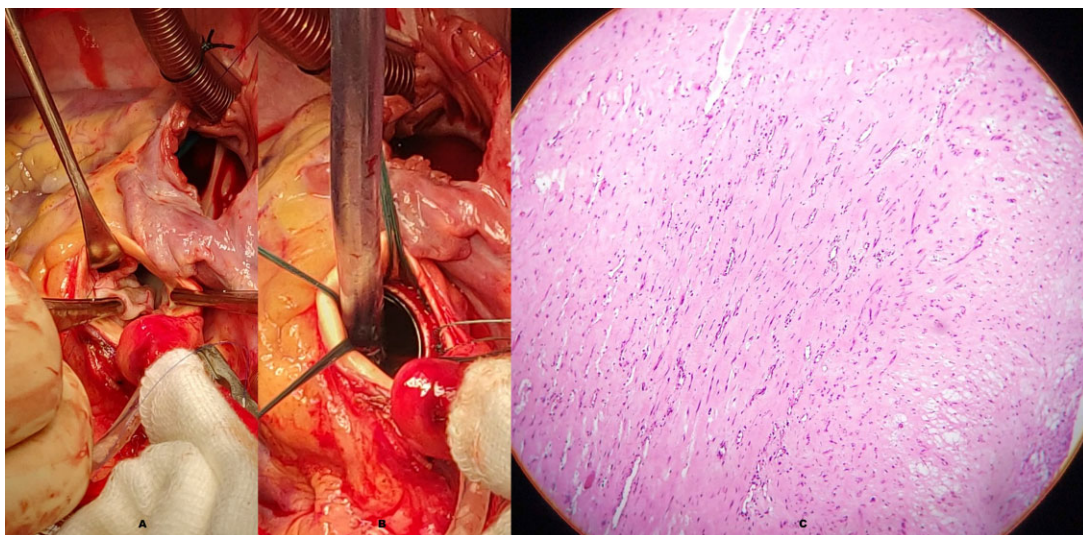


Figure 3 (A) Intra-operative images of the aneurysm of the non-coronary cusp of aorta with a defect (probe being passed through the defect). (B) Intra-operative images of surgical aortic valve replacement with a metallic valve. (C) Haematoxylin and eosin staining of the aortic cuspal tissue ($\times 10$) showing the thick fragments of hyalinized collagenous tissue with foci of myxoid degeneration and varying infiltrates of neutrophils, lymphocytes, and plasma cells.

Table 1 Previous case report/series of perforated aortic cusp aneurysm in tricuspid aortic valve

Case report/ case series	Age	Sex	Aetiology	Blood culture	Perforated cuspal aneurysm	Additional valve involvement	Clinical manifestation	HPE findings	Management
1. Fujiwara et al.	40	Female	IE	Negative	LCC	No	AR	Not described	AVR
2. Aokage et al.	49	Male	IE	Negative	RCC	No	AR	Neutrophils infiltration, necrosis with abscess formation, fibrocollagenous tissue	AVR
3. Kinoshita et al.	33	Male	Unknown	Negative	LCC	No	AR	Significant fibrosis	AVR
4. Plein et al.	63	Male	IE	<i>Streptococcus pneumoniae</i>	LCC	No	AR	Not described	AVR
5. Harada et al.	57	Male	IE	Negative	RCC	Yes	AR + MR	Thinning of RCC with infiltration of inflammatory cells	AVR + MVR
6. de Castro et al.	42	Male	IE with HOCM	<i>Staphylococcus epidermidis</i>	RCC	Yes	AR + MR	Not described	AVR + MVR
7. Asami et al.	67	Female	IE	Negative	LCC	No	AR	Chronic infective endocarditis, thin wall, scar formation	AVR + MAZE
8. Zhang et al.	53	Male	IE	<i>Enterococcus faecalis</i>	NCC	No	Mild to moderate AR	Not described	Antibiotics
9. Singh et al.	26	Male	Probable IE	Negative	RCC with perforation at annulus	No	AR + CHB	RCC showed signs of inflammation.	PPI followed by AVR
10. Naraoka et al.	71	Male	Healed IE	Negative	NCC	No	AR + Angina Pectoris	Inflammatory cell infiltration elastic fibre disappear	AVR + CABG
11. Sugawara et al.	73	Male	Healed IE	Negative	RCC	No	AR + AS	No active inflammatory changes hyalinization	AVR
12. Minamimura et al.	37	Male	IE, annular abscess	<i>Streptococcus viridans</i>	NCC	No	AR	Inflammatory cell infiltration, necrosis with abscess formation, no microorganism	AVR
13. Present case	30	Male	IE	<i>Streptococcus viridans</i>	NCC	No	AR + CHB	Thick fragments of hyalinized collagenous tissue with foci of myxoid degeneration, occasional foci of calcification, and varying infiltrates of lymphocytes/plasma cells without evidence of granulomatous lesions	AVR followed by PPI

IE, infective endocarditis; HPE, histopathological examination; LCC, left coronary cusp; RCC, right coronary cusp; NCC, non-coronary cusp; AR, aortic regurgitation; MR, mitral regurgitation; AS, aortic stenosis; CHB, complete heart block; AVR, aortic valve replacement; MVR, mitral valve replacement; PPI, permanent pacemaker implantation.

Serial blood cultures grew *Streptococcus viridans* species (Figure 1C). A two-dimensional transthoracic echocardiography showed a hyperdynamic enlarged LV with an end diastolic dimension of 64 mm and the presence of a saccular aneurysmal sac arising from the non-coronary cusp (NCC) of aortic valve (AV) and prolapsing into the left ventricular outflow tract (LVOT) without aortic root dilatation (aortic annulus 25 mm, sinus 32 mm, sinotubular junction 30 mm). Additionally, the saccular aneurysm had ruptured at its apex, through which two jets of aortic regurgitation (AR) passed from the aortic root to LVOT (see [Supplementary material online, Movie 1](#)). Transoesophageal echocardiography (TEE), which was performed for better delineation of the AV, showed aneurysmal prolapse of the NCC that resembled a wreath bobbing chaotically into the LVOT during diastole and into the aortic root during systole (Figure 2, see [Supplementary material online, Movies 2 and 3](#)). The jet of severe AR entered the LV through a defect on the NCC. There were no obvious vegetations, root abscess, aneurysmal dilatation of sinus of valsalva, no shunted flow communicating with other cardiac chambers, or any evidence of type A aortic dissection on the TEE. A diagnosis of perforated aortic cuspal aneurysm with CHB subsequent to infective endocarditis was made based on (i) two major and two minor modified Duke criteria being fulfilled [positive blood culture for typical organism, new valvular regurgitation, fever >38°C, and Osler nodes (immunologic phenomena)] and (ii) echocardiographic findings. Patient was administered broad spectrum intravenous antibiotics along with inotropic support followed by transfemoral temporary pacemaker insertion. Following 2 weeks of antibiotic therapy and clinical stabilization, the patient was planned for surgical AV replacement. Intraoperatively, the prolapsed NCC was noted, forming an aneurysmal structure with a rupture hole near its apex (Figure 3A). The dissected LCC was redundant, clean, and transparent without vegetations. The patient underwent successful AV replacement with a metallic valve (Figure 3B). Histopathological examination of the excised AV revealed microscopically, thick fragments of hyalinized collagenous tissue with foci of myxoid degeneration, occasional foci of calcification, and varying infiltrates of lymphocytes/plasma cells without evidence of granulomatous lesions (Figure 3C). Culture of the excised valvular tissue grew *Streptococcus viridians*. Post-operative course was uneventful and failure of return to sinus rhythm even after 6 weeks following surgery led to a dual chamber permanent pacemaker implantation (PPI). The patient is largely asymptomatic in the past 1 year following surgery with good compliance to anticoagulant therapy.

Discussion

Aortic valvular aneurysms are one of the rare complications of infective endocarditis. The mechanism of formation of these aortic valvular aneurysms is still unclear with multiple hypotheses being proposed. The most plausible explanation is that the infective endocarditis of the AV increases the pre-existing valvular tissue injury leading to weakening of the aortic cusp.⁴ Subsequently, due to the diastolic aorto-LV pressure gradient, there occurs the formation of an aneurysmal sac which progressively increases in size and ultimately perforates. Most of the previously reported cases of aortic cuspal aneurysm and perforation present with acute heart failure.⁵ The trigger for acute progressive congestive heart failure is the incarceration

and rupture of the AV aneurysm into the LVOT which explains the haemodynamic deteriorations in our case. The CHB as a presentation of aortic cuspal aneurysm is very rare with just one case reported previously in the literature. Singh *et al.*⁶ documented CHB in a 26-year-old male with severe acute aortic regurgitation and cuspal aneurysm and perforation of the right coronary cusp with the possible aetiology being endocarditis. The CHB complicating bacterial endocarditis has been reported in 4–10% of cases with most of them being associated with AV involvement.⁷ Conduction blocks in these patients could be possibly due to (i) contiguous spread of infection from valve leaflets to the conduction tissue, (ii) inflammatory oedema, and (iii) myocardial abscess.⁷

A review of literature revealed only 25 cases of aortic cuspal aneurysms till date of which perforation was reported in 12 (48%; [Table 1](#)).^{5,6,8–17} Most of these cases were males (83.3%) with an average age of 50.9 years (range: 26–73 years). Infective endocarditis was confirmed or strongly suspected in 11/12 (91.7%) with right coronary cusp being most commonly involved. All the patients had AR (100%) while subaortic stenosis and LV outflow obstruction were reported in one patient each. Diagnosis of aortic cuspal aneurysm is often established on echocardiography with the appearance of a saccular aneurysm beneath the AV towards the LV during diastole phase. This aneurysmal structure disappears or shrinks during systole and appears as a ring-like structure on short axis.⁵ Jets of aortic regurgitation are appreciated on colour Doppler echocardiogram. Treatment is often surgical with replacement of the diseased valve along with appropriate antibiotic therapy for management of infective endocarditis.⁵ Histopathological evaluation of the diseased valve often reveals active inflammatory changes, myxoid degeneration as was seen in our case.⁵ In patients with conduction block and infective endocarditis, the exact timing of PPI remains controversial in the absence of definite guidelines. In the previously reported case by Singh *et al.*,⁶ the authors performed PPI first followed by AV replacement, while in our case we waited for at least 6 weeks following AV replacement. This was done to (i) allow for spontaneous recovery to sinus rhythm with subsidence of infection and inflammatory oedema and (ii) achieve good antimicrobial coverage prior to implantation of PPI.

Lead author biography



Shekhar Kunal is currently an Assistant Professor in ESIC Medical College, Faridabad, India. He completed his medical schooling from Maulana Azad Medical College, New Delhi and subsequently was awarded the Doctorate of Medicine in Cardiology from SMS Medical College, Jaipur. He has special interests in structural heart disease and cardiovascular imaging.

Supplementary material

[Supplementary material](#) is available at *European Heart Journal – Quality of Care and Clinical Outcomes* online.

Consent: A written informed consent was obtained from the patient for submission and publication of this case report including image(s) and associated text in line with COPE guidance.

Conflict of interest: There are no conflicts of interest to declare.

Funding: There was no research funding available for this study.

Data availability: The data underlying this article are available in the article and in its online supplementary material.

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