

Austrian Syndrome with Quadruple Valve Infective Endocarditis – A Case Report and Literature Review of Quadruple Valve Infective Endocarditis

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BACKGROUND:

The triad of pneumonia, meningitis and endocarditis secondary to *Streptococcus pneumoniae* was first described by Heschl in 1862 followed by Osler in 1881. In 1956, Austrian described a case series of 8 patients with pneumococcal endocarditis and rupture of the aortic valve. This triad became known as the Austrian syndrome. We present what we believe to be the first reported case of Austrian syndrome with quadruple heart valve involvement, and review the literature detailing cases of quadruple valve infective endocarditis.

CASE AND METHODS:

- A 54-year-old Malay male teetotaler, presented to our hospital with a two-week history of fever, cough and headache.
- He had no past medical history and denied intravenous drug abuse.
- Clinical examination revealed an early diastolic cardiac murmur with basal crackles in the left lung.
- Chest radiograph and computed tomography scan revealed left lower lobe bronchopneumonia (Figure 1).
- He was commenced on intravenous antibiotics.
- Repeated blood cultures were non-yielding.
- A transthoracic echocardiography showed vegetations involving both the mitral and aortic valves, with aortic regurgitation.
- On the fourth day of hospitalization, he developed right upper limb weakness and two episodes of generalized tonic-clonic seizures.
- Magnetic resonance imaging of the brain revealed leptomeningeal enhancement, as well as cortical infarcts in the left temporal and parietal lobes (Figure 2).
- A lumbar puncture performed was biochemically consistent with meningitis (Table 1). Bacterial gram stain and culture were negative.
- Urinary *Streptococcus pneumoniae* antigen was negative but cerebrospinal fluid *Streptococcus pneumoniae* antigen was positive.
- A diagnosis of Austrian syndrome with septic brain emboli was made.
- Antimicrobial therapy was switched to meningeal doses of vancomycin and ceftriaxone.
- A transesophageal echocardiography (Figure 3) showed quadruple-valve endocarditis, complicated by aortic valvular perforation and regurgitation, as well as the presence of a small ventricular septal defect.
- He was planned for surgery but unfortunately developed multi-organ failure and passed away.

A computerized PubMed search for reports of quadruple valve infective endocarditis was conducted for the literature review. References from each of the articles obtained by these searches were also reviewed for relevant case reports. Only reports available in English were included.

We present our case along with the other case reports in Table 2.

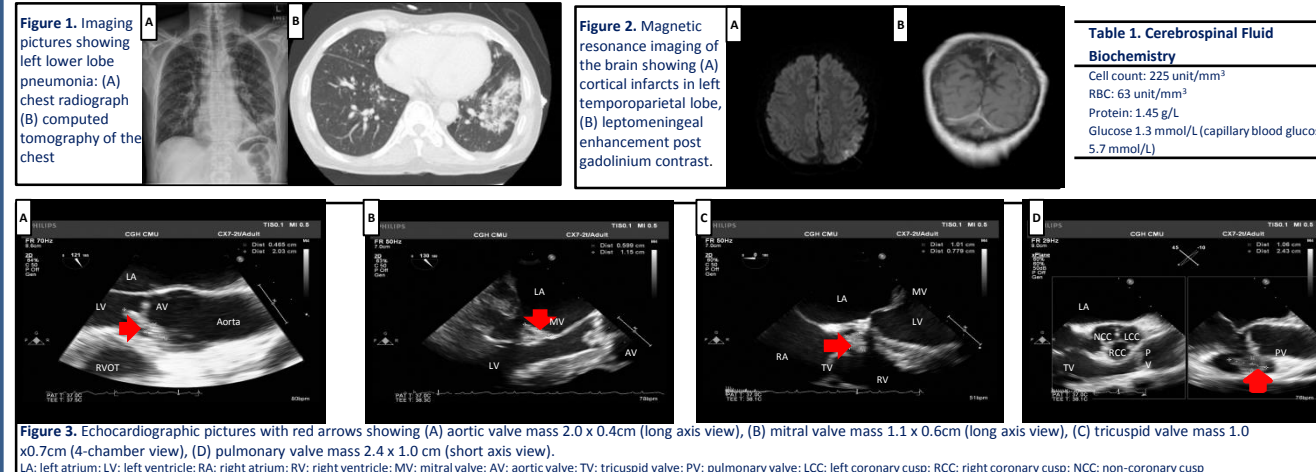


Figure 1. Imaging pictures showing left lower lobe pneumonia: (A) chest radiograph (B) computed tomography of the chest
Figure 2. Magnetic resonance imaging of the brain showing (A) cortical infarcts in left temporoparietal lobe, (B) leptomeningeal enhancement post gadolinium contrast.
Figure 3. Echocardiographic pictures with red arrows showing (A) aortic valve mass 2.0 x 0.4cm (long axis view), (B) mitral valve mass 1.1 x 0.6cm (long axis view), (C) tricuspid valve mass 1.0 x 0.7cm (4-chamber view), (D) pulmonary valve mass 2.4 x 1.0 cm (short axis view).
 LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle; MV: mitral valve; AV: aortic valve; TV: tricuspid valve; PV: pulmonary valve; LCC: left coronary cusp; RCC: right coronary cusp; NCC: non-coronary cusp

Table 1. Cerebrospinal Fluid Biochemistry

Cell count: 225 unit/mm ³
RBC: 63 unit/mm ³
Protein: 1.45 g/L
Glucose 1.3 mmol/L (capillary blood glucose 5.7 mmol/L)

RESULTS:

A total of 19 cases were available, including our patient. The mean age of presentation was 48.3 years, with a range from 7 to 82 years. There were more men (84.2%) than women (15.8%). Four patients had a history of intravenous drug abuse, another four had underlying congenital heart disease and one had both. Two patients (10.5%) had two microorganisms isolated. *Staphylococcus aureus* and *Streptococcus viridans* (three cases, 15.8% each) were the most commonly implicated microorganism. Heart failure was the commonest complication, afflicting eight patients (42.1%). Eight patients (42.1%) underwent surgery. Overall, ten patients died, giving a case fatality rate of 52.6%. Cardiac surgery was of borderline significance in predicting survival (p = 0.054). Endocarditis was diagnosed only at post-mortem in two cases (10.5%).

CONCLUSION:

We report the first case of quadruple valve infective endocarditis secondary to *Streptococcus pneumoniae*, occurring in a patient with Austrian syndrome and newly diagnosed ventricular septal defect. Risk factors for quadruple valve infective endocarditis include congenital heart disease and intravenous drug abuse. Clinicians need to remain vigilant in the presence of these risk factors.

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Table 2. Literature Review of Quadruple Valve Infective Endocarditis

Case	Age/Sex	Underlying Conditions	Risk Factors		Microbiological Diagnosis	Echocardiography		Complications	Key Antibiotics	Surgery	Outcome	Ref.
			Valvular Heart Disease	IVDU		Diagnostic TTE	Diagnostic TEE					
1	54 Male	VSD	Yes	No	<i>S. pneumoniae</i>	No	Yes	Embolism Multi-organ failure (including heart failure) Valve perforation	VAN + CEF	No	Death	NA
2	41 Male	Hepatitis C	No	Yes	Group G <i>Streptococcus</i>	NM	NM	Shock	CP + GEN	Yes	Survived	1
3	39 Female	Nil	No	Yes	Methicillin sensitive <i>S. aureus</i>	No	Yes	Heart failure Valve perforation	CLO + GEN	Yes	Survived	2
4	58 Male	VSD	Yes	No	NM	NM	NM	Renal failure	NM	Yes	Survived	3
5	64 Male	DM, CKD, HTN	No	No	<i>E. faecalis</i>	No	Yes	Renal failure	AMP + GEN	NM	Death	4
6	76 Male	CKD, COPD, Factor XIII deficiency	No	No	<i>E. faecalis</i>	No	Yes	Heart failure	AMP + GEN + IMI	Yes	Survived	5
7	47 Male	VSD	Yes	No	<i>S. pyogenes</i>	NM	NM	NM	NM	Yes	Survived	6*
8	34 Male	Hepatitis B, VSD	Yes	No	Alpha-hemolytic <i>Streptococcus</i>	NM	NM	Multi-organ failure (including heart failure)	CP + GEN + CLI	Yes	Death	7*
9	7 Male	Nil	No	No	Methicillin resistant <i>S. aureus</i>	Yes	Not done	Shock	VAN then TIG	No	Survived	8
10	46 Male	Mental retardation	No	No	<i>S. viridans</i>	Yes	Not done	Heart failure	CP	No	Survived	9
11	57 Male	Nil	No	No	<i>S. gallolyticus</i>	No	Yes	Heart failure	CEF + GEN then AMP-SUL + GEN then AMO-CLA	Yes	Survived	10
12	82 Male	Colon cancer	No	No	Methicillin resistant <i>S. aureus</i>	No	Yes	Embolism	NM	No	Death	11
13	31 Male	Alcohol abuse	No	Yes	<i>Corynebacterium</i>	Not done	Not done	Multi-organ failure (including heart failure) Heart failure	VAN + GEN + CP + TMP-SMX + ERY	No	Death*	12
14	56 Female	DM, CKD, HTN	No	No	<i>P. aeruginosa</i> <i>S. marcescens</i> <i>S. mutans</i>	Not done	Not done	Shock	NM	No	Death*	13
15	48 Male	Alcoholic liver cirrhosis	No	No	<i>S. gallolyticus</i>	No	Yes	Heart failure	CP + GEN	No	Death	14
16	58 Male	Alcohol abuse	No	No	<i>S. gallolyticus</i>	No	Not done	Heart failure Cardiac tamponade	NM	Yes	Death	15
17	24 Male	Nil	No	Yes	<i>P. aeruginosa</i>	NM	NM	NM	NM	No	Death*	16
18	78 Female	Nil	No	No	<i>S. viridans</i>	NM	NM	Heart failure Pericarditis	CP + GEN	No	Death*	17
19	18 Male	Tetralogy of Fallot	Yes	Yes	<i>S. viridans</i>	Yes	Not done	Shock Myocardial infarction	VAN	No	Death	18

VSD: ventricular septal defect; DM: diabetes mellitus; CKD: chronic kidney disease; HTN: hypertension; COPD: chronic obstructive pulmonary disease; IVDU: intravenous drug user; TTE: transthoracic echocardiography; TEE: transesophageal echocardiography; NM: not mentioned; VAN: vancomycin; CEF: ceftriaxone; CP: crystalline penicillin; GEN: gentamicin; CLO: cloxacillin; AMP: ampicillin; IMI: imipenem; CLI: clindamycin; TIG: tigecycline; AMP-SUL: ampicillin sulbactam; AMO-CLA: amoxicillin clavulanic acid; TMP-SMX: trimethoprim sulfamethoxazole; ERY: erythromycin
 *Autopsy performed; *Abstract only