Case Report

'Scarcity amidst plenty'- A rare combination of enterococcal pulmonary valve endocarditis in a young girl: A case report

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Abstract

Isolated pulmonary valve infective endocarditis (IE) with *Enterococcus faecalis* in a child with an underlying ventricular septal defect (VSD) is rare. A ten year old girl with a VSD with moderate to severe right ventricular mid cavity obstruction presented with fever and loose stools. Examination did not reveal any peripheral stigmata of IE. Five of six blood cultures sent in two sets 12 hours apart, grew *E. faecalis* sensitive to β-lactams. 2D echocardiography revealed a 7×4 mm vegetation on the pulmonary valve. She was treated with intravenous antibiotics for 6 weeks. Inflammatory markers normalized by 6 weeks and the vegetation cleared by the 7th week. Amongst the aetiological agents, isolation of *Enterococcus faecalis* is rare in children and drug resistance is an emerging problem. *Enterococcus* species related native valve IE needs a prolonged course of therapy with penicillin G/ampicillin together with gentamicin for 4-6 weeks. Heart failure and cerebral embolization are the most commonly encountered complications.

***Keywords**: paediatric, infective endocarditis, VSD, Enterococcus faecalis, pulmonary valve*

Introduction

Infective endocarditis (IE) is a well-known entity in children, especially those with a background of underlying structural heart disease.[1] Amongst the aetiological agents, isolation of *E. faecalis* is rare in children.[2] Ventricular septal defects rarely predispose to IE with a rate of 2.4 per 1000 children with the lesion.[3] Right-sided vegetations are rare and isolated pulmonary valve involvement is even rarer with an incidence of 5-10% and 1.5-2% of all IE cases respectively.[4,5] This is attributed to low pressure gradients and lower oxygen saturation of the right heart, different characteristics and vascularization of the endothelium and relatively low prevalence of right sided congenital heart lesions.

We report a paediatric case of IE with isolated pulmonary valve involvement in a background of VSD who had five blood cultures positive for *E. faecalis.* This is the first paediatric

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enterococcal endocarditis recorded in the IE registry of Colombo South Teaching Hospital which is maintained from 2007.

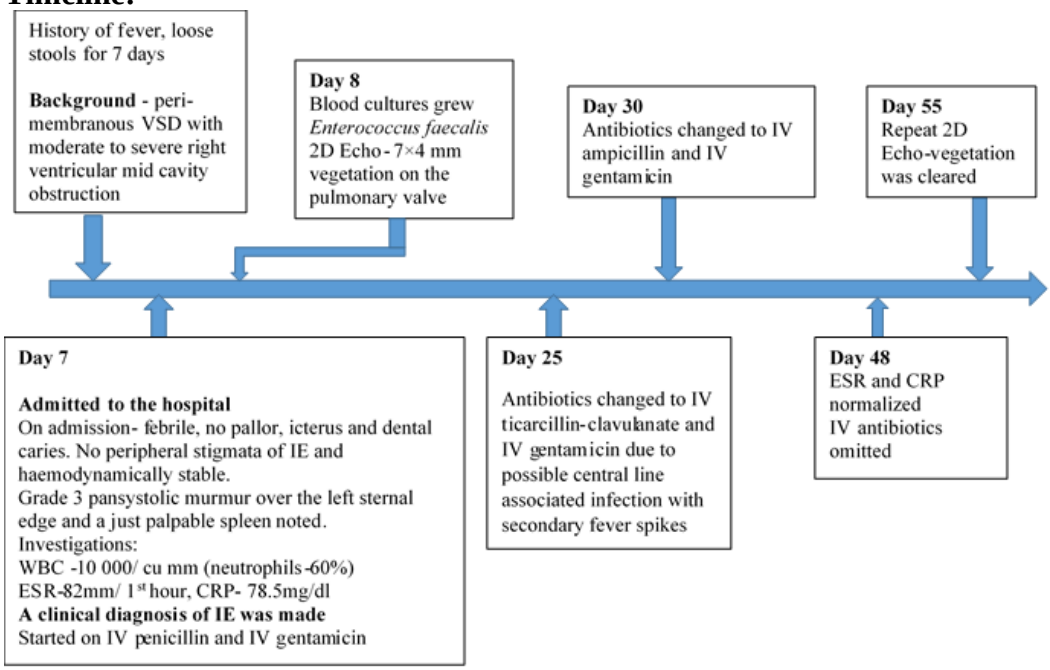
Case report

A ten year old girl presented with a seven day history of fever and loose stools. She was diagnosed with a peri-membranous VSD with moderate to severe right ventricular mid-cavity obstruction since infancy. She was the only child of healthy non-consanguineous parents, with a normal birth and development history. Examination revealed an averagely built febrile child without pallor, icterus or dental caries. There were no peripheral stigmata of IE. She was haemodynamically stable. Precordial examination revealed a grade 3 pansystolic murmur over the left sternal edge. The spleen was just palpable.

On the day of admission, her white cell count was 10,000/cu mm with 60% neutrophils. Platelets were 356,000/cu mm. The erythrocyte sedimentation rate (ESR) was 82mm/1st hour and C-reactive protein (CRP) was 78.5mg/dl. Urine full report did not show red blood cells. Five of six blood cultures taken on day 2 of admission and sent in two sets 12 hours apart grew *E. faecalis* (identification using the BD Phoenix^{TM} system). Antibiotic sensitivity testing was done using both disc diffusion testing and the BD Phoenix^{TM} system. The isolate was sensitive to penicillin, ampicillin (MIC $\leq$ 2 µg/ml), linezolid, teicoplanin, and vancomycin. High level gentamicin resistance could not be checked due to unavailability of discs.

2D echocardiography showed a 7×4 mm vegetation on the pulmonary valve without obstructing the pulmonary flow. The patient was treated with intravenous (IV) penicillin for 19 days followed by IV ticarcillin-clavulanic acid for 5 days to cover a possible central line related infection, and IV ampicillin for 18 days. IV gentamicin was given from day 1 to day 42 in a synergistic dose. The child responded to treatment without any complications. CRP and ESR were normalized by 6 weeks of treatment. The vegetation was cleared by the 7th week.

Timeline:



Discussion

IE is an infection involving the endocardium with an incidence of 1 per 1000 paediatric admissions.[6] This figure has remained static for the past 40 years, even though the aetiological agents have changed.[2] Viridans streptococci, *Staphylococcus aureus* and coagulase negative staphylococci are the main offenders of IE.[2] *Enterococcus faecalis* isolated in our patient is a Gram positive gut commensal species which gives rise to numerous non-cardiac infections and in which drug resistance is an emerging problem.[2] Enterococcal endocarditis usually runs a sub-acute course and affects older males following genitourinary procedures or younger females following obstetric procedures. High risk conditions for IE includes cyanotic congenital heart disease, prosthetic heart valves, systemic to pulmonary shunts and conduits, indwelling central venous catheters and residual post-cardiac defects.[2] Enterococcal endocarditis usually occurs in mitral and aortic valves, yet the pulmonary valve was affected in our patient.[7]

Definitive diagnosis of IE is made by the modified Duke criteria which incorporates clinical, microbiological, pathological, and echocardiographic characteristics.[8] The main presentation of IE is intermittent low grade fever. Hepatosplenomegaly is noted in 15-20% of patients.[2] Cutaneous manifestations include petechiae, Osler's nodes, Janeway lesions, and splinter haemorrhages.[2] However, peripheral manifestations of infective endocarditis due to *Enterococcus* sp. are uncommon (<25% of the cases).[7] IE causes several signs and symptoms due to sterile and infective emboli and immunological phenomena. Embolic strokes lead to focal neurological signs, intracranial haemorrhages and micro abscesses.[2] Embolic phenomena result in osteomyelitis, pneumonia and meningitis, especially in neonates.[2] Immunological reactions lead to vasculitis and glomerulonephritis.[2] Intracardiac complications consist of conduction defects, pericardial disease, myocardial abscesses and valvular insufficiency leading to congestive cardiac failure.[2] Microbiological isolation of the organism requires repeated blood cultures from discrete sites. Although isolation of enterococci in urine is thought to be a useful clue[7], our patient's urine culture was negative.

A prolonged course of 4-6 weeks of parental treatment is needed since the organisms are embedded within the fibrin-platelet matrix in very high concentrations.[9] The duration and choice of treatment is further dependent on the organism and presence of prosthetic surfaces.[9] The recommended practice is to use a bactericidal antibiotic rather than bacteriostatic drugs.[9] Treatment of enterococcal endocarditis is difficult due to relative resistance to IV penicillin/gentamicin commonly used for streptococcal endocarditis.[10, 11] The American Heart Association states that in enterococcal IE of native valves in children, antibiotic treatment should be for 4-6 weeks with penicillin G or ampicillin together with gentamicin.[9] However, more recent studies have shown that relapses were more frequent among survivors treated for 4 weeks than in those treated for 6 weeks.[12] For patients infected with susceptible strains, and who are unable to tolerate β-lactams, vancomycin in combination with gentamicin is recommended.[10]

The commonest complication of enterococcal endocarditis is heart failure, which occurs in around 50% of adult patients, with some requiring valve replacement.[7] Complications with embolization can be seen in 27% to 43% of patients, with the brain as the commonest end organ. Mortality rates can vary from 11% to 35%, and death is usually due to heart failure or embolization.[7] Data from the paediatric population is not widely available. Our patient made a complete recovery without complications or sequelae.

Conflict of Interest statement: None

Ethics statement / Consent for publication: Written consent was taken from the parents of the patient

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Author contributions

Perera TMR – Involved in patient management, literature review, manuscript writing, and provided guidance, critical revision of the article and provided final approval of the submitted version

Ekneligoda AMBWMRNN – Involved in patient management, literature review and manuscript writing

Chandrasiri NS – Involved in patient management, provided guidance, critical revision of the article

Waniganayake YC – Involved in patient management and manuscript writing

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