

Complicated Subacute Bacterial Endocarditis in a Patient with Ventricular Septal Defect

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حالة معقدة لالتهاب جرثومي تحت الحاد للشغاف المبطن للقلب لطفلة لديها فتحة قلبية بالحجاب الحاجز البطني

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الملخص: تعد الإصابة بالتهاب الشغاف المبطن للقلب ظاهره غير مألوفة، لكنها خطيرة وذات مضاعفات متعددة قد تهدد حياة المريض. وعلى الرغم من التقدم في العلاج، إلا أن هناك ارتفاع مستمر في معدلات الاعتلال والوفيات. في هذه الدراسة نقدم حالة فتاة تبلغ من العمر ثمانية سنوات كانت تعاني من أعراض ارتفاع درجة حرارة الجسم لمدة شهرين ونصف و قد تم تشخيصها بوجود عيب خلقي في القلب في مرحلة الطفولة الأولى، إن ثبت أن لديها فتحة صغيرة في الحجاب البطني. وقد تم تشخيص الحالة باعتبارها التهاب بالغشاء المبطن للقلب سببه جرثومة ستربتوكوكاس ميوتانس. وقد حدث لديها انتقال للتعفن تسبب في انسداد رئوي. ومع ذلك فقد تم شفاؤها باستخدام مضادات الميكروبات المناسبة. وعليه، ينبغي على الأطباء التحسب بدرجة عالية لعدوى التهاب الشغاف باعتباره سببا محتملا للحمى التي تمكث لفترات طويلة وبخاصة مع وجود أمراض القلب الخلقية. ومع التغييرات الجديدة في تعليمات إعطاء الأدوية، لم تعد أمراض القلب غير الإزرقاقية، التي لم تتم معالجتها بعمليات جراحية من ضمن الحالات التي توصف لها هذه الأدوية الوقائية وهذا بالطبع يدفع باتجاه مزيد من الدراسات المستفيضة لمعرفة نسبة حدوث مثل هذه الحالات قبل الاستنتاج للمدى الطويل.

مفتاح الكلمات: التهاب الشغاف الجرثومي تحت الحاد؛ ستربتوكوكاس ميوتانس؛ انسداد رئوي متعفن؛ حمى عقدية؛ تقرير حاله؛ عمان.

ABSTRACT: Infective endocarditis (IE) is an uncommon but life-threatening infection. Despite advances in management, it still causes high morbidity and mortality. We report the case of an 8-year-old girl who presented with a prolonged fever of 2.5 months duration and a history of a small perimembranous ventricular septal defect. She was diagnosed with subacute bacterial endocarditis secondary to *Streptococcus mutans*. The patient developed a septic pulmonary embolism; however, with the use of appropriate antimicrobial therapy, she made an uneventful recovery. Clinicians should have a high index of suspicion for IE as the possible cause of a prolonged fever, especially in the presence of congenital heart disease (CHD). Currently, IE prophylaxis is not indicated for unrepaired acyanotic CHD. Nevertheless, with the new changes in the guidelines, more prospective studies are needed to investigate the incidence of IE in such lesions, before long-term conclusions can be drawn.

Keywords: Endocarditis, Subacute Bacterial; *Streptococcus mutans*; Pulmonary Embolism, Septic; Erythema Nodosum; Case Report; Oman.

INFECTIVE ENDOCARDITIS (IE) IS A relatively a rare condition in children but it causes significant morbidity and mortality. Repaired and unrepaired congenital heart diseases (CHD) are associated with a high lifetime risk of IE; patients with ventricular septal defect (VSD) have the highest risk.¹ A VSD can present in an acute or subacute phase due to many organisms like the *Streptococcus viridans* group (*S. mutans*, *S. mitis*), and the *Enterococcus* and *Staphylococcus* species.² IE results from complex interactions between the microbial pathogen in the bloodstream, the matrix molecules and platelets at the sites of the endocardial cell damage. For many years antibiotic prophylaxis

has been advocated but the guidelines have recently changed. Early diagnosis and prompt therapy can prevent the drastic complications which can ensue from IE.

Case Report

An 8-year-old girl presented to the Emergency Department of Sultan Qaboos University Hospital, Oman, with a 2.5 month history of an intermittent low-grade fever associated with a decrease in appetite and weight loss. There was a history of an itchy erythematous skin rash on the lower limbs which had initially lasted for a few days. There was

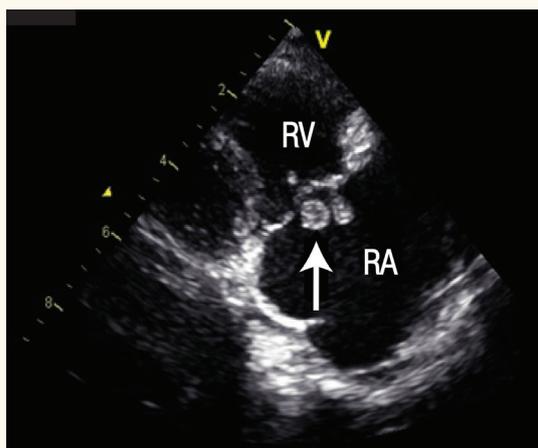


Figure 1: Echocardiographic modified right ventricular inflow view showing the vegetations (arrow) attached to the tricuspid valve.

RA = right atrium; RV = right ventricle.

no history of cough, chest pain, urinary or bowel symptoms; neither was there a history of a recent dental extraction or of joint pain or swelling. She had visited the local hospital and health centres a few times and had been prescribed some oral antibiotics and topical cream which had only resulted in minimal improvement. During the neonatal period, she had been diagnosed with a small perimembranous VSD (pmVSD) and was under follow-up for this condition. Three days prior to presentation, she had developed a high-grade fever of 40 °C; this was associated with a mild cough but with no shortness of breath or chest pain.

Clinical examination revealed a girl of slight build, afebrile, and with a heart rate of 146/min, respiratory rate 20/min, blood pressure 93/66, weight 16.3 Kg (below 3rd percentile), height 163 cm (below 3rd percentile) with normal oxygen saturation.



Figure 2: Purplish macular skin rash at the dorsum of the left foot indicating *erythema nodosum*.

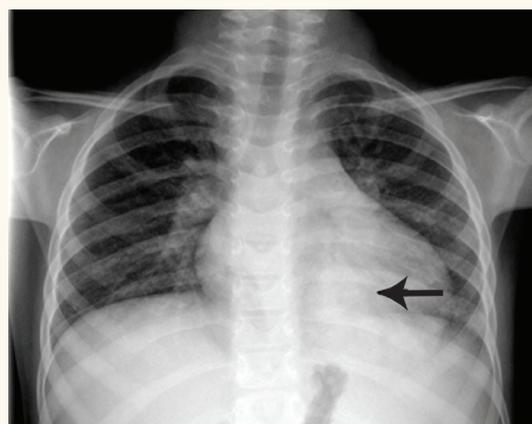


Figure 3: Posterior-anterior chest X-ray showing a large dense consolidation (arrow) in the left lower zone consistent with a pulmonary embolism.

There was no significant lymphadenopathy. An oral assessment revealed multiple dental caries. The cardiac examination revealed a hyperdynamic *precordium* with thrill and a grade 4/6 pansystolic murmur heard best in the left lower sternal border. An abdominal examination showed non-tender hepatomegaly (the liver span was 12 cm) and splenomegaly (8 cm) below the costal margin. The laboratory investigations revealed a picture of hypochromic microcytic anaemia with haemoglobin of 7.2 g/dL, a high red blood cell distribution width of 27, a white cell count of $6.5 \times 10^9/L$ and platelets of $302 \times 10^9/L$. The C-reactive protein level was 32 mg/L and the erythrocyte sedimentation rate 20 mm/hr. The liver and kidney function tests were normal.

An echocardiography showed a 4 mm pmVSD with a left-to-right shunt with a peak gradient of 85

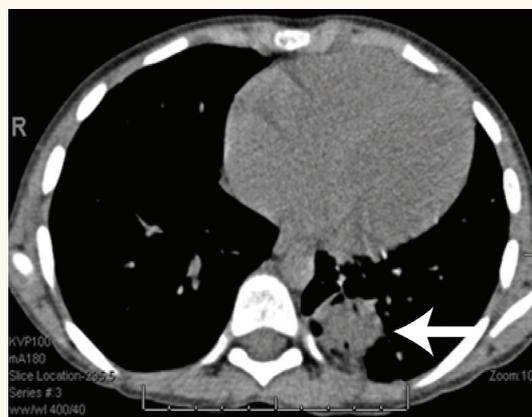


Figure 4: Computed tomography scan of the chest (axial view) showing the large wedge-shaped pulmonary embolism involving the basal segment of the left lower lobe (arrow).

mmHg. There were two vegetations attached to the tricuspid valve with moderate regurgitation and no stenosis. The large one measured 8 x 6 mm and the smaller one 6 x 4 mm [Figure 1]. The patient's left atrium and left ventricle were not dilated and there was no pericardial effusion. The ventricular systolic function was normal. A diagnosis of subacute bacterial endocarditis (SBE) was made based on the clinical picture and the echocardiographic findings.

After taking three blood cultures, the patient was started on intravenous antibiotics: ampicillin 50 mg/Kg six hourly, vancomycin 15 mg/Kg eight hourly and gentamicin 2.5 mg/Kg eight hourly. All blood cultures grew *S. mutans*.

On the eighth day of admission, the patient developed a painful small macular lesion on the dorsum of her left foot measuring about 2 x 1 cm; this was consistent with *erythema nodosum* [Figure 2]. The patient remained clinically stable until day 13 of admission when she developed a sudden lower left chest pain associated with a shortness of breath and vomiting and a fever of 38.6 °C. Her vital signs were normal. Both the chest X-ray [Figure 3] and the computed tomography (CT) scan [Figure 4] showed a wedge-shaped consolidation with an air bronchogram on the basal segment of the left lower lobe consistent with a pulmonary embolism. A repeat echocardiography showed the disappearance of the larger tricuspid valve vegetation with the small one still attached. The patient's cardiac function remained normal with no evidence of pulmonary hypertension. There was no evidence of other embolic phenomena or complications. Both the neurological status and urine analysis were normal. The patient was observed initially in the intensive care unit and remained haemodynamically stable. The patient was continued on ampicillin and gentamicin and repeat blood cultures were negative. Intravenous antibiotics were continued for total of six weeks after which she was discharged in a good condition.

On follow-up, the patient remained afebrile with an improvement in her appetite, the regression of her hepatosplenomegaly and the normalisation of the inflammatory markers. A follow-up echocardiography done after the discharge showed a mild tricuspid valve regurgitation with resolution of the vegetation. She was referred for dental review and VSD repair.

Discussion

We here reported a young girl known to have a small pmVSD who developed serious SBE secondary to *S. mutans*; this is a commensal of the oral cavity and considered a primary cause of dental caries. The organism can survive in the bloodstream and has been reported as a cause of IE with significant morbidity and mortality.³⁻⁵ IE has serious complications in both paediatric and adult patients with CHD with an increasing incidence relative to the decrease in rheumatic fever. Despite improvements in early diagnosis, management and therapy, it can still lead to high rates of morbidity and mortality; these vary from 7–25%.⁶⁻⁷ Hence, IE is an ongoing, life-threatening complication that may affect the long-term outcome of CHD.

The classic presentation of IE includes fever, anaemia, positive blood cultures and heart murmur, all of which were present in our patient; however, there was a delay in the diagnosis despite the presence of the clinical and laboratory findings which were suggestive of SBE. This case highlights the importance of keeping a high index of suspicion for IE as it could present in a subacute phase.

The diagnosis of IE is based on the modified Duke criteria and, once diagnosed, the therapy includes appropriate antimicrobial therapy for 2–6 weeks.⁸ Our patient had two major criteria and several minor criteria for the diagnosis of IE. The presence of IE in CHD can lead to major complications in and outside the heart. Congestive heart failure occurs in up to 40% of cases and is the leading cause of haemodynamic compromise; this could be due to many factors including the destruction of valves, myocarditis or arrhythmias.⁹ Extracardiac complications are also frequent—in up to 43% of cases—and are caused either by embolic events or immune phenomena.^{9,10}

In this patient, the reappearance of fever and the sudden respiratory symptoms were due to a septic pulmonary embolism. The diagnosis of septic pulmonary embolism was based on the clinical presentation, the chest X-ray and the CT findings, in addition to the echocardiographic findings of the dislodgement of the vegetation. One differential diagnosis is splenic rupture; however, this patient was haemodynamically stable. Other differential diagnoses include embolic episodes to the spleen, bowel or other related vascular structures. This

patient's abdominal Doppler ultrasound only showed hepatosplenomegaly with no evidence of embolic episodes. The hepatomegaly was most likely due to the immunological response secondary to the IE as she was not in heart failure. Because she was haemodynamically stable and the blood cultures were negative within a few days of starting the antibiotics, with normal cardiac function, no surgical intervention was warranted.

Given the prognosis, morbidity and the high cost of IE management, antibiotic prophylaxis has long been recommended in order to minimise the incidence of IE and, in fact, the rate has decreased dramatically. Based on expert opinions, case control studies and daily practice, the American Heart Association (AHA) guidelines, last revised in 2007, have led to a significant reduction and limitation of cardiac diseases and procedures in which IE prophylaxis is indicated.¹¹ Currently, IE prophylaxis is not indicated for unrepaired acyanotic CHDs such as VSD, bicuspid aortic valve and patent *ductus arteriosus*. Nevertheless, given the new changes in the AHA guidelines, more prospective studies are needed, especially in developing countries, to monitor carefully the incidence of IE in such lesions before long-term conclusions can be reached.

Conclusion

We have reported a case of SBE secondary to *S. mutans* in an 8-year-old girl who had a small pmVSD complicated by a septic pulmonary embolism. She made an uneventful recovery with the appropriate therapy. IE still carries high morbidity and mortality despite advances in management. Physicians should have a high index of suspicion for IE as early detection and prompt therapy are mandatory to optimise patient outcomes, especially in the presence of CHD. Long-term data should be collected concerning the efficacy of antibiotic prophylaxis against IE, especially in acyanotic CHD, before long-term conclusions on patient management can be reached.

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