CASE REPORT

Rheumatic fever with severe carditis: still prevalent in the South West Pacific

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SUMMARY

Rheumatic heart disease (RHD) has a worldwide prevalence of 33 million cases and 270 000 deaths annually, making it the most common acquired heart disease in the world. There is a disparate global burden in developing countries. This case report aims to address the minimal RHD coverage by the international medical community. A Tahitian boy aged 10 years was diagnosed with advanced heart failure secondary to RHD at a local clinic. Previous, subtle symptoms of changes in handwriting and months of fever had gone unrecognised. Following a rapid referral to the nearest tertiary centre in New Zealand, urgent cardiac surgery took place. He returned home facing lifelong anticoagulation. This case highlights the RHD burden in Oceania, the limited access to paediatric cardiac services in countries where the RHD burden is greatest and the need for improved awareness of RHD by healthcare professionals, and the general public, in endemic areas.

CASE PRESENTATION

Patient A, a normally fit Tahitian boy aged 10 years, presented to his community clinic with a 1-week history of fever, worsening breathlessness and nausea leading to loss of appetite in August 2016. On clinical examination, he was sweating, tachycardic and a murmur of grade 3/6 was noted. He also stated he had a sore throat 3 months previously, which was dismissed as trivial due to its intermittent nature. His mother highlighted a rash on the upper aspects of both thighs. He had intermittent hip and knee pain for the 12 months, resulting in limping and reduced mobility—emphasised by the fact he was visiting his neighbour much less than usual. He commented on a 3-month history of a change in handwriting at school, and a greater tendency to drop objects, suggestive of a subtle chorea.¹ There was no significant medical history. The birth history includes a 2-week stay in hospital after birth due to ongoing fevers. The social history includes residing with three other family members in a two-bedroom apartment; however, there are occasions when family members stay for periods of time and the household can exceed nine persons. Overcrowding is a risk factor for developing acute rheumatic fever (ARF), as there is increased risk of transmission of group A streptococcus.¹ Patient A's mother provides the primary household income; she left school at 20 years and works in a manual occupation, emphasising rheumatic heart disease (RHD) as a disease of poverty.

He was referred to a regional adult cardiologist in French Polynesia (FP) 3 days after his presentation to the local clinic. The cardiology team performed ECG that detected sinus tachycardia (128 bpm) and left atrial (LA) and left ventricular (LV) hypertrophy. Transthoracic echocardiography (TTE) revealed severe mitral-aortic disease with predominant mitral incompetence and pulmonary arterial hypertension. There was evidence of LA and LV dilation, with preserved ejection fraction. Blood cultures were negative. The primary differential diagnosis was ARF meeting the Jones criteria² with carditis, raised inflammatory markers, temperature and elevated group A streptococcal titres. Following a diagnosis of ARF and secondary heart failure, he was started on steroids, diuretics and penicillin and placed on bed rest. Given the advanced carditis found on the TTE, a semiurgent referral was made to Starship Children's Hospital, New Zealand, which provides tertiary cardiac care for children in the South Pacific.

Three weeks after presentation to his local clinic, patient A arrived in Auckland. His weight was 54 kg (>95th centile) and height 154 cm (>94th centile). Initial observations indicated lethargy and heavy sweating, with symptoms of sore throat and nausea. There were no signs of finger clubbing or infective endocarditis with a regular radial pulse. The jugular venous pulse was 2 cm elevated. On auscultation, there was a hyperdynamic praecordium with a displaced apex beat to the sixth intercostal space, in the midclavicular line. There was a parasternal heave and a pansystolic murmur, radiating to the axilla. A diastolic rumble was audible at the lower left sternal edge (LLSE), with diastolic murmur over the aortic area. There was reduced air entry at the lung bases. The abdomen examination revealed right upper quadrant tenderness. The liver edge was palpable 2 cm below the right subcostal margin. Initial blood test results showed haemoglobin 89 g/L, ESR 114 mm in 1 hour and CRP 178 mg/L. The antistreptolysin O titre was 885 IU/mL (normal limit 240 IU/mL) and anti-DNAse antibodies were 1200 U/mL (normal limit <680 U/mL). Peripheral blood cultures showed no growth. Transoesophageal echocardiography replicated the findings of the TTE that was performed in FP 2 weeks earlier: the mitral valve showed mixed stenosis and regurgitation. The mitral valve was thickened and the posterior mitral valve leaflet restricted in motion. There was a visible coaptation defect with severe mitral regurgitation. Severe aortic regurgitation was also present. There was dilation of the LA and LV; LVEDD

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6.6 cm with Z score +5.1 and LVEDS 4.4 cm with Z score +4.4. The ejection fraction was preserved. The diagnosis of ARF with carditis seemed secure; a differential diagnosis of infective endocarditis was eliminated by negative blood cultures.

Cardiac surgery took place 2 weeks after arrival at Starship and was indicated due to continued cardiac failure and pulmonary hypertension. The valves were not amenable to valve repair with the mitral valve showing acute and chronic changes. He received a size 25 mm On-X prosthetic mitral valve replacement and a size 21 mm On-X prosthetic aortic valve replacement and underwent a tricuspid valve annuloplasty. There were no peri-operative complications. Postoperative TTE indicated LV modelling with LV dysfunction; this is expected to improve in time. The prosthetic aortic and mitral valves were functioning normally, with a mild prosthetic valve gradient. Postoperative management included optimising pain relief through a patient-controlled analgesia device, drug treatment for congestive heart failure with spironolactone, furosemide and captopril, improving compliance with drug treatments (eg, exchanging paracetamol tablets for syrup) and physiotherapy/play therapy. He was started on anticoagulation with Coumadin, a warfarin substitute used in FP, and continued on intramuscular benzathine penicillin every 28 days as secondary prevention for ARF. He was discharged from hospital after 7 days and followed in outpatients for INR control before returning to FP after 4 weeks.

GLOBAL HEALTH PROBLEM LIST

- ▶ Awareness of signs and symptoms of ARF
- ▶ Missed opportunity to diagnose ARF earlier as hip and knee pain symptoms seemed minor to the family. Symptoms and signs of Sydenham's chorea can be very subtle.
- ▶ Disease burden of RHD in Oceania
- ▶ Access to cardiac surgery in for those in remote regions and lower middle-income countries (LMIC)

GLOBAL HEALTH PROBLEM ANALYSIS

RHD is the most common acquired heart disease in the world, with a disparate global burden affecting children in developing countries.³ In wealthier countries, the burden of RHD tends to be confined to indigenous populations, such as the Aborigine population of Australia and the Maori population of New Zealand.⁴ The 2013 Global Burden of Diseases study estimates a worldwide prevalence of 33 million cases, causing 9 million disability-adjusted life years lost and 270 000 deaths annually.⁵ In 2016, the ARF rate in FP was 42/100 000, though it is unclear how the various cultures in FP, for example, Maori, Cook islanders and Samoans, contribute to this. The global health problems associated with RHD are: the disproportionate burden in LMIC, delay in the recognition of ARF/RHD and limited access to cardiac surgery for ARF/RHD. RHD has minimal reportage from the worldwide medical community; this report is an effort to raise awareness of a disease conferring immense cardiovascular mortality and morbidity in childhood and young adulthood.

What is rheumatic fever?

Rheumatic fever is caused by a group A β haemolytic streptococcal infection, and initially presents as a sore throat. Approximately 2–4 weeks later, symptoms manifest as fever, polyarthralgia, polyarthritis, chorea and erythema marginatum.² Acute rheumatic carditis occurs when there is a cross-reaction between the bacterial carbohydrate cell wall and the valve tissue (antigenic mimicry), causing chronic damage to valve tissue,⁶

which is then termed RHD. Risk factors include poverty, overcrowding, lack of access to medical care and young age.⁷

Disease burden and recognition of ARF in FP and LMIC

FP, population 270 000, is a collection of 118 Pacific islands—one of which is Tahiti, where our patient is from. The WHO estimates RHD incidence in FP school children at 8/1000.⁸ An ongoing rise in incidence has been attributed to improved record keeping.⁹ Despite the ease of prophylaxis, delayed symptom recognition by professionals and patients (as with our case), and poor antibiotic compliance have been linked to the existence of this preventable disease.¹⁰

Another country in Oceania with a high RHD prevalence is Fiji; a recent study of RHD-associated deaths there found that RHD is responsible for a significant premature loss of human life.¹⁰ The 50% RHD mortality under 40 years in Fiji directly affects economic productivity in young adulthood,¹⁰ the age group on which economies thrive. A particularly poignant case by Steer *et al*¹¹ indicates the importance of healthcare professional training to recognise RHD in Fiji. A Fijian girl aged 7 years died from RHD-associated severe valve deformation, after being misdiagnosed 2 years earlier with asthma. The nurse who recognised her symptoms on her last, fatal presentation had attended an RHD workshop the previous day.¹¹ In a similar presentation to patient A, joint pains and irregular movements went undiagnosed, despite ongoing fevers for several preceding months. Irregular movements can manifest as changes in handwriting,¹ which our patient also commented on. His mother was not aware of this, as ARF symptoms can be subtle unless arthritis is severe. This suggests improved community ARF education is needed to enable early presentation to healthcare services; in our case, the patient's mother could have recognised symptoms earlier. Furthermore, healthcare physicians and nurses from Europe working abroad are not aware of, or have not seen ARF cases, hence training may be required to enable symptom recognition as in the Steer *et al* case.

The international RHD burden in LMIC is also worth noting; the Global Rheumatic Heart Disease Registry (the 'REMEDY' study) found the average age of an individual in Africa to be affected by RHD is 28 years, with over 66.7% women.¹² The disparate burden on women's health in the reproductive age illustrates the need for adult and paediatric RHD services and improved obstetric care, especially as RHD is a major cause of maternal death in Africa.¹³

Access to cardiac surgery

The access to cardiac surgery and postoperative care in geographically remote regions (as for our patient) and in LMIC is notoriously difficult. Despite free public clinics, some FP regions on outer islands have only a nurse or a health assistant for primary healthcare. There is no paediatric cardiac surgeon on site in Tahiti, but close communication between the island doctors and colleagues in New Zealand meant patient A's treatment plan was agreed via email correspondence. He was booked on a flight to New Zealand with a medical escort (BP, personal communication). Although patients in FP are fortunate to have a health system that offers transfer to nearby countries, this is not an international standard. In Western countries, there is 1 congenital heart surgeon per 3.5 million inhabitants, whereas in Africa, services vary from 1 per 35 million¹⁴ to 2 paediatric cardiologists serving an entire country.¹⁵ It has been recommended as a 'global priority' that an improved multidisciplinary approach to valve repair requires specialist surgeons,

high-quality echocardiography and maintenance of therapy in the local populations.¹⁶

Reduced access to cardiac care is exacerbated by the post-operative need for anticoagulation. Children with RHD often have damaged valvular tissue, and surgeons make it a priority to repair rather than replace valves,¹⁶ as 40% of patients from LMIC do not adhere to anticoagulation guidance after valve replacement.¹⁷ The REMEDY study¹² demonstrated that if valve repair is not possible, cardiac centres use tissue rather than mechanical valve replacement to reduce postoperative thrombotic risk.¹⁸ It is not clear whether non-concordance is due to lack of understanding by the patient, poor communication by healthcare professionals or lack of access to anticoagulative medications and monitoring facilities.¹⁹ An encouraging study from Japan showed that point-of-care testing improves INR measurements,²⁰ though whether this intervention is applicable to LMIC populations is uncertain.

Historically, global health interventions for paediatric cardiac care (PCC) involved 'surgical tourism' (when a medical team of paediatric cardiologists and cardiac surgeons went to underserved area for short period of time), leaving little legacy. Alternatively, accessing healthcare from high-income countries was an option only available for wealthier families from developing countries, aggravating healthcare inequalities. Experts recommend promotion of partnerships between developed and underserved countries, which may enable LMIC creation of sustainable cardiac facilities.²¹ It has also been demonstrated that improvement in one medical area within an LMIC can result in parallel upgrades in other hospital facilities and local infrastructure.²¹ PCC can be overlooked in countries with a very limited healthcare budget, as treatments for more widespread diseases are prioritised.²²

Finally, a key determinant of whether PCC programmes succeed is funding. In this context, our patient can be said to be unlucky to have been affected by ARF and RHD, but fortunate to have had access to cardiac surgery. There is reasonable access to cardiac surgery for children in the South West Pacific mainly to New Zealand, and recently to Indian cardiac units. Patients from FP can also be treated in France. Some patients in Oceania have access to fly-in units.¹⁹

Indications for cardiac surgery in ARF/RHD

Occasionally, a child with ARF may present in acute pulmonary oedema to ruptured chordae tendinae of the mitral valve, leading to a rapid rise in LA pressure and pulmonary oedema. Often mistaken for acute pneumonia, cardiac surgery in this scenario is lifesaving.²³ The more common scenario in ARF with severe carditis is cardiac compensation and remodelling of the LV size over months to years. There is a well-developed ARF/RHD register and good rates of secondary prophylaxis in FP (BP, personal communication). Cardiac surgery is indicated based on symptoms and LV size relation to body surface area.^{1 24} Our patient also had severe dilation of the LV, which has been associated with reduced LV function due to increased preload and afterload.²⁴ In the case of our patient, who had mitral and aortic regurgitation, valvular replacement was a necessity as the heart valves were too damaged, and the consequences of a failed valve repair in the context of established heart failure were too hazardous. Postoperative care for our patient includes lifelong anticoagulation in a remote region of a well-resourced country. As well as the risk of subsequent thromboembolism, our patient may require reoperation when he reaches adult size if the prosthetic valves are too small.

CONCLUSION

There is international consensus on how to reduce the global health burden of RHD; current guidance focuses on secondary antibiotic prophylaxis,³ primary prevention and primordial prevention by improving living conditions.⁵ Themes reinforced in this case report include the need for improved training of healthcare professionals to detect ARF symptoms earlier, informing the population to attend a clinic at an early stage (sore throat, joint pains), access to echocardiography, surgical valve repair rather than replacement and particularly in the case of the latter, enhanced postoperative care.

Learning points

- ▶ Awareness of the global health problem of rheumatic heart disease and the disproportionate burden on low-income and middle-income countries (LMIC).
- ▶ Barriers to early recognition and diagnosis, for example, lack of knowledge of unusual symptoms of acute rheumatic fever such as irregular movements, as seen in patient A, as well as infrastructure issues such as poor quality of echocardiography and reduced access to cardiac care units for remote populations and in LMIC.
- ▶ International efforts to improve access to cardiac surgery.
- ▶ Indications for cardiac surgery in paediatric cases of RHD.

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