Case Report

Acute Congestive Heart Failure in a Young Woman

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ABSTRACT

We describe the case of an adult First Nations woman employed as a daycare worker who presented with clinical symptoms, signs, and imaging suggestive of acute heart failure. In our report, we discuss the likely diagnosis of acute rheumatic fever. Novel teaching points: The incidence of rheumatic fever in Canada is likely underestimated, and it is important to consider the diagnosis of acute rheumatic fever in individuals presenting with acute heart failure. More research is needed in Canada to further identify groups most at risk for developing this disease.

Case

A previously healthy 29-year-old First Nations daycare worker presented to the emergency department with a 5-day history of orthopnea and swelling of the lower limbs. She denied fever, and chest or joint pain. She described a transient rash to her anterior chest.

Two weeks prior to her presentation, she had been diagnosed with Group A Streptococcal pharyngeal infection with Streptococcus pyogenes confirmed by throat culture. She was treated with oral amoxicillin, which she prematurely discontinued after only 6 days, due to gastrointestinal side effects.

Her clinical examination demonstrated oxygen saturations of 89% on room air, and mild increased work of breathing at a rate of 20 respirations per minute. She was afebrile, with an oral temperature of 36.9 °C, and hypertensive, with a blood pressure of 166/117 mm Hg. Her jugular venous pulsation was biphasic and measured at 5 cm above the sternal angle. Cardiac auscultation revealed a soft first heart sound and a grade 3/6 holosystolic murmur with radiation to the apex.

There was also a grade 1/6 mid-diastolic rumble. The second heart sound was normal. Breath sounds were diminished in the lung bases, with crackles and expiratory wheeze present bilaterally. She had bilateral pitting edema to the knees. Although not visualized in the hospital, she described a history of a rash on her chest, which we postulated represented erythema marginatum.

Her initial investigations showed elevated high-sensitivity troponin T at 60.9 ng/L, and an N-terminal-prohormone brain natriuretic peptide of 4191 pg/ml (normal: <300). Her C-reactive protein (CRP) level was also elevated, at 15.5 mg/L. Her white blood cell count was within normal limits. Chest radiograph demonstrated bilateral air space disease and increased cardiac size. Electrocardiogram showed a sinus rhythm with a rate of 93 beats/minute and a normal PR interval. Due to diagnostic uncertainty, a computed tomography scan of the chest was performed in the emergency department, which showed pulmonary edema and no pulmonary embolism.

The patient was admitted to the cardiology service and given loop diuretic until her congestion was relieved. A diagnosis of acute rheumatic valvulitis was made based on the positive Group A Strept throat culture, acute valvulitis, and history of rash. Echocardiography performed the next day showed normal left and right ventricular function. The mitral valve apparatus was normal, but the mitral leaflets were thickened with associated severe mitral regurgitation (Video 1, view video online). The tricuspid valves were also thickened with corresponding mild-to-moderate regurgitation. While in the hospital, she was started on furosemide, amiodipine, and candesartan for treatment of her edema and hypertension. Her congestive symptoms resolved, and she was discharged after 7 days in the hospital. She was started on
penicillin G secondary prophylaxis to be continued until at least age 40 years.

At 2 months postdischarge, her previously abnormal laboratory values had normalized, with a CRP level of 2.4 mg/L and N-terminal-prohormone brain natriuretic peptide of 61.30. Echocardiography several months later (Videos 2 and 3 [view videos online]) showed complete resolution of the previous valve thickening with only mild residual mitral regurgitation.

**Discussion**

Acute rheumatic fever (ARF) is a non-suppurative sequelae of pharyngeal group A streptococcal infection that can affect numerous organ systems. The mechanism by which ARF develops is thought to be related to molecular mimicry, whereby cellular components of the streptococcal cell are structurally similar to those seen at various sites in the human body. When antibodies are formed to the components of the streptococcal cells by the immune system, there is an antigen-mediated response and subsequent destruction of those cell components. This leads to autoimmunity related to cross-reaction of proteins and other bacterial cell components with cardiac, brain, synovial, and other tissues in the human body.

Over the past several decades, ARF has decreased in incidence in Europe, North America, and other developed nations. This decrease is attributed to improvements in hygiene, access to antibiotics, health care, a reduction in household crowding, and other socioeconomic changes. In Canada, ARF is not currently a reportable disease, although available data from 2005 showed the incidence to be 0.1 to 2 cases per 100,000 persons in the general population in Canada, the United States, and Western European nations. For those aged 0-19 years, the Canadian Pediatric Surveillance Program conducted a review between 2004 and 2007 and found an incidence of 2.9 cases per million children per year. There have been sporadic reports of small outbreaks of disease, and in 2015, an incidence of 8.33 per 100,000 was reported in Northern Ontario indigenous communities. This alarmingly high figure mirrors the incidence in rural New Zealand and Australia, where public health measures have already been taken to address social, cultural, and political gaps that undermine evidence-based care. A diagnosis of rheumatic fever in adults is more rare than among youth, in developed nations; however, there does not appear to be any recent data regarding the incidence of ARF in the general adult Canadian or North American population specifically. In fact, the true incidence and prevalence of ARF are likely underestimated by a factor of 10, based on the opinion presented in a recent review.

The revised Jones criteria, as outlined by Gewitz et al., are used in diagnosing ARF. They include evidence of antecedent streptococcal infection, as well as major and minor criteria. Major criteria include evidence of carditis, arthritis, chorea, erythema marginatum, and subcutaneous nodules. Minor criteria include hyperpyrexia, arthralgia without signs of inflammation, acute-phase laboratory values (erythrocyte sedimentation rate and CRP level), and prolonged PR interval on electrocardiogram. The diagnosis requires the presence of either 2 major, or 1 major and 2 minor criteria.

The patient described in this case did not meet criteria sufficiently for diagnosis with definite ARF. She had a positive throat culture for group A streptococci, completed a suboptimal course of antibiotics, and had clinical evidence of carditis, fulfilling only one of the major criteria. The antistreptolysin titer was not performed. Her history of rash could have been erythema marginatum, but it was never visualized in hospital. Her CRP level, although elevated, was lower than that required to fulfill the minor criterion.

The revised Jones criteria do stipulate that “possible” rheumatic fever can be diagnosed if patients are presenting atypically in “high-incidence settings.” These settings include rural, underdeveloped areas and overcrowded households. In contrast, our patient resided in an urban setting, but working at a daycare center may be the factor that put her at increased risk for infection. Table 6.1 in the updated Australian Rheumatic Heart Disease Guidelines further stratify groups at higher risk for developing ARF, including specific indigenous groups, owing to higher incidences in these populations. Furthermore, this table outlines overcrowding and additional considerations that increase risk. Adapting these criteria to the Canadian population through epidemiologic research would inform front-line clinicians seeking a more refined approach to borderline cases, such as the one we encountered. Finally, such criteria might increase awareness of the benefit of complete antibiotic treatment of group A streptococcal infection, which can reduce the risk of developing rheumatic fever by up to 60%.

**Conclusions**

It is pertinent for practitioners to consider the diagnosis of ARF in patients of any age presenting with acute congestive heart failure, in order to provide the best care and avoid unnecessary investigations. Despite the low incidence in Canada and other developed nations, the disease remains an important presence in certain populations. Timely identification of affected patients allows for appropriate and expedient treatment of symptoms, as well as secondary prophylaxis to prevent recurrence. More research is needed to further understand the true incidence of ARF in Canada, as well as to better identify those populations most at risk.
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Supplementary Material
To access the supplementary material accompanying this article, visit CJCOpen at https://www.cjcoopen.ca/ and at https://doi.org/10.1016/j.cjco.2020.10.005.