Diagnosed late cardiac sarcoidosis in a patient with atrioventricular block

Introduction

Cardiac sarcoidosis may present as advanced atrioventricular block. In this case report the patient was first diagnosed with advanced atrioventricular block, with no clinical finding specific to systemic sarcoidosis noted at that time. Six years later, the patient was diagnosed with pulmonary and cardiac sarcoidosis. Prednisolone was administered daily. One year after medication was started, the patient's heart function and other clinical symptoms had improved. This case highlights the difficulty in detecting cardiac involvement in the early stages of sarcoidosis.

Discussion

Sarcoidosis is a multi-organ granulomatous disease. Cardiac involvement can lead to congestive heart failure and life-threatening cardiac arrhythmias (Longcope and Freiman, 1952). Yoshida et al (1997) reported that right ventricular endomyocardial biopsy proved cardiac sarcoidosis in 11.2% of patients (n=89) undergoing permanent pacemaker implantation for advanced atrioventricular block. The extent of scarring and fibrous tissue in the left ventricle, especially the atrioventricular node, bundle of His, and right and left bundle branch, was observed and these pathological changes obtained from autopsy validate the clinical course of development of atrioventricular block in this patient (Valantine et al, 1987).

This patient was not diagnosed with cardiac sarcoidosis despite advanced atrioventricular block of unknown origin. Although it was difficult to diagnose cardiac sarcoidosis at first, at the very least cardiomyopathy that can cause atrioventricular block should be suspected. Unfortunately, when the patient was diagnosed with atrioventricular block, the authors were unable to find any evidence of cardiac sarcoidosis. Therefore, an accurate diagnosis of systemic sarcoidosis was delayed.

Histopathology of cardiac sarcoidosis was not detected, but cardiac sarcoidosis is not often diagnosed based on the histological findings of myocardial granulomas (Dubrey and Falk, 2010). According to guidelines (Hiraga et al, 1993), the patient can be diagnosed as having cardiac sarcoidosis despite no evidence of histopathology. Studies have suggested that 18F-fluorodeoxyglucose positron emission tomography can detect the early stage of cardiac sarcoidosis with higher sensitivity despite there being a

Figure 1. Electrocardiography shows advanced atrioventricular block.

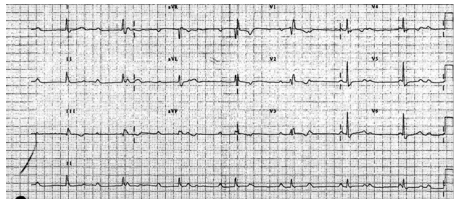
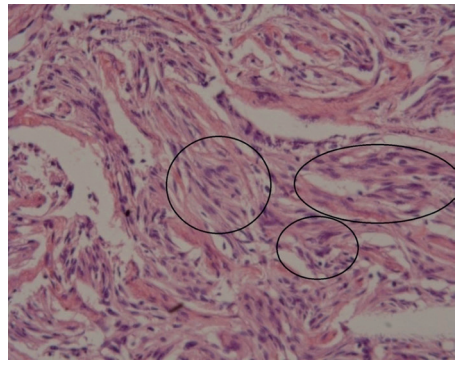


Figure 2. Histopathology of pulmonary sarcoidosis, demonstrating the presence of ill-defined non-caseating granulomas (circled).



CASE REPORT

A 57-year old Asian woman who had dyspnoea and dizziness was admitted to hospital. The patient was diagnosed with heart failure (ejection fraction 42%) and advanced atrioventricular block on electrocardiography (*Figure 1*). The aetiology of the advanced atrioventricular block remained unknown as further examinations such as coronary angiography were performed later. Because of persistent advanced atrioventricular block and other symptoms, the patient received a pacemaker electively. After the procedure, regular follow-ups were conducted with no symptoms or changes in cardiac function detected on serial physical examinations and echocardiography.

Approximately 6 years later, the patient complained of dyspnoea and dry cough. Computed tomography showed right hilar enlargement and the patient underwent bronchoscopy. Detection of granuloma in tissue (*Figure 2*) lead to the final diagnosis of pulmonary sarcoidosis. To differentiate cardiac sarcoidosis from other cardiomyopathies such as cardiac amyloidosis, the patient underwent echocardiography, fluorodeoxyglucose positron emission tomography (FDG-PET) and heart biopsy.

Cardiac function was decreased in comparison to the previous echocardiography (ejection fraction 27%). FDG-PET demonstrated increased uptake in the left ventricular myocardium, especially at the apex and anteroseptal wall (*Figure 3*). Even though there was a negative finding on heart biopsy, the authors believed this was cardiac sarcoidosis because of the presence of advanced atrioventricular block and the FDG-PET findings.

With the diagnosis of pulmonary and cardiac sarcoidosis, prednisolone was administered daily. One year after prednisolone administration, echocardiography revealed an increase in ejection fraction to 39%. The patient's clinical condition is good and her cardiac function is regularly checked as an outpatient.

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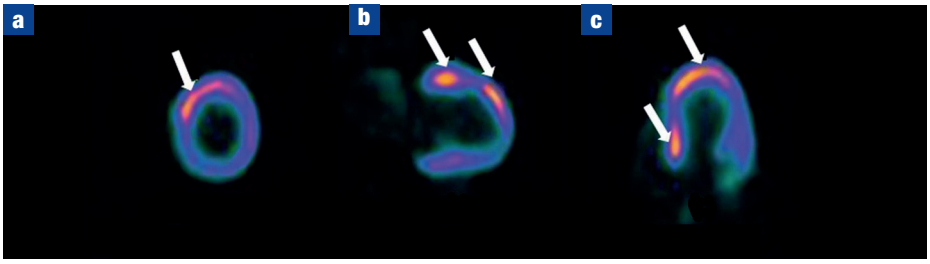
Figure 3. The fluorodeoxyglucose metabolic images demonstrate hypermetabolism in the anteroseptal and apex wall (arrows). **a.** Short axis. **b.** Vertical long axis **c.** Horizontal long axis. A previous cardiac catheterization (not shown) revealed normal coronary arteries.



higher frequency of false positive results (Ishimaru et al, 2005). This modality was used to demonstrate cardiac involvement in the current case. **BJHM**

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LEARNING POINTS

- Cardiac involvement can be difficult to detect in the early stages of sarcoidosis.
- If clinicians detect patients who have unknown origin atrioventricular block, they should actively assess for cardiomyopathy such as cardiac sarcoidosis.
- If cardiac sarcoidosis is suspected without cardiac biopsy, fluorodeoxyglucose positron emission tomography may be a good method of diagnosis.

Spontaneous surgical emphysema: an unusual complication of paediatric asthma

Subcutaneous surgical emphysema is extremely rare in children. It is usually detected in the neck or precordial area; dyspnoea and distended neck veins may be present (Saadoon and Janahi, 2015). Focused history should include predisposing factors, e.g. asthma, repeated vomiting, vigorous coughing or unrecognized trauma (Ameh et al, 2006; Saadoon and Janahi, 2015). Rare causes, e.g. spontaneous tracheal rupture, should be considered (Karthikeyan and Pulimoottil, 2015). Measurement of peak expiratory flow rate is contraindicated (Saadoon and Janahi, 2015).

A 5-year-old asthmatic boy presented with an exacerbation with dyspnoea and violent coughing for 1 week. He was receiving inhaled beclomethasone. Clinical examination revealed tachypnoea, normal breath sounds and no respiratory distress. His neck appeared swollen with crepitus felt over his neck and chest wall. Chest X-ray (*Figure 1*) revealed widespread subcutaneous surgical emphysema bilaterally, no pneumothorax with minimal abnormal shadowing in both infrahilar regions. He was clinically well and treated conservatively with oral azithromycin, and discharged home 48 hours later. Clinical review after 7 days revealed complete resolution of symptoms and subcutaneous surgical emphysema. **BJHM**

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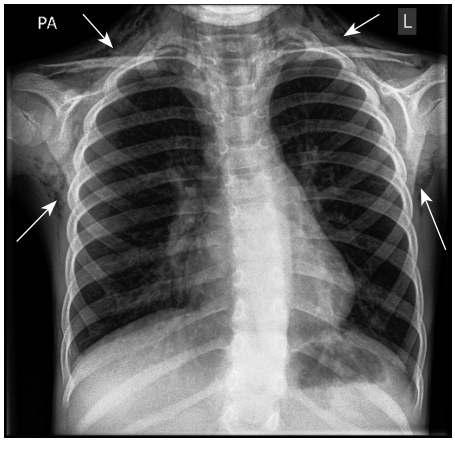
Figure 1. Chest X-ray showed widespread surgical emphysema (arrows).



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