SYSTEMIC SCLERODERMA WITH COMPLETE HEART BLOCK

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INTRODUCTION

Although involvement of the heart and the occurrence of arrhythmias in systemic scleroderma is well documented, [1,2,3,4,5,6] only a few cases of complete heart block in generalised scleroderma have been reported in the literature. [3,7,8,9] We report two patients with generalised scleroderma who presented with symptoms secondary to complete heart block. One patient successfully underwent permanent pacemaker implantation with relief of his symptoms.

CASE 1

A 33 year old Chinese female was first admitted into the University Hospital, Kuala Lumpur in October 1967 with a two year history of symmetrical polyarthritis involving mainly the small joints of the fingers. Clinical examination revealed both knees were tender and swollen, as were most of the proximal interphalangeal joints. No abnormalities of the skin were noted. Pulse rate was 100/min.; BP: 120/80 mmHg and examination of the heart was normal. The electrocardiogram showed normal sinus rhythm. The patient responded satisfactorily to indomethacin and prednisolone.

Eighteen months later, she developed stiffness of the hands. Typical skin changes of scleroderma were noted in the face, dorsum of hands and forearms. No Raynaud's phenomenon was demonstrable. Examination of the cardiovascular system was normal. Rose-Waaler test was negative. Respiratory function test revealed a restrictive ventilatory defect and barium swallow examination revealed a dilated oesophagus with gastro-oesophageal reflux. At fluoroscopy, decreased oesophageal peristalsis were noted. A chest x-ray showed a normal sized heart.

Three months later she was readmitted to the hospital with a four week history of effort dyspnoea and occasional chest pain, not typical of angina pectoris. Her pulse was 104/min, regular and blood pressure was 110/60 mmHg. The heart was enlarged but no evidence of heart failure was noted. A chest radiograph confirmed the cardiac enlargement. An electrocardiogram showed first degree heart block and left bundle branch block with a sinus rate of 104/min (Fig. 1).

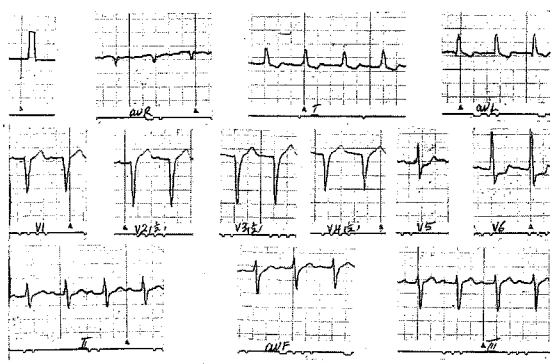

Fig. 1 Case 1: Initial ECG showing first degree heart block with left bundle branch block.

She noted the first of a series of dizzy spells in the ward and an electrocardiogram done revealed complete heart block with frequent ventricular extrasystoles (Fig. 2). The heart rate varied between 40-70 beats per minute during this period. Over the following week she experienced numerous Stokes-Adams attacks. Response to ephedrine was unsatisfactory. Syncopal attacks became less

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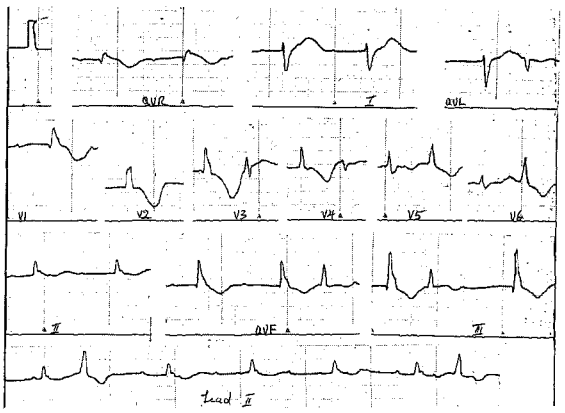

Fig. 2 Case 1: Subsequent ECG showing complete heart block with frequent ventricular extrasystoles.

frequent but she continued to experience frequent dizzy spells.

She was next treated with oral saventrine reaching a maximum dose of 300 mg daily with some positive results. Stokes-Adams attacks were infrequent and the heart failure which she subsequently developed was adequately controlled with diuretics and she was able to go home.

She was readmitted two weeks later with severe bronchopneumonia and exacerbation of her heart failure. Pulse rate was 40-50 beats per minute. She responded poorly to antibiotic and anti-heart failure therapy and frequent syncopal attacks persisted despite oral sympathomimetic drugs. She died on November 11th 1969, eleven weeks following onset of complete heart block. No autopsy was performed.

CASE 2

A 56 year old Chinese male was referred to the University Hospital in June 1979 following confirmation of complete heart block in another hospital.

He was first told of a slow heart rate four years previously when he saw his family doctor for an unrelated complaint. He volunteered a history of attacks of dizziness on exertion but denied any fainting spells. These dizzy spells became so frequent and disabling that he had to resign from his job as a cinema-hall manager. At about the same time he noted gradually worsening tightness of his fingers and progressive difficulty in forming a fist. The skin over his face and dorsum of the fingers became thick and less wrinkled. He did not experience any dysphagia but has been having severe constipation since 1975. There was no history of muscle weakness or Raynaud's phenomenon.

He experienced the first of many syncopal attacks while straining at stool in December 1978 and was admitted into a nearby district hospital. Following confirmation of complete heart block, he was started on saventrine with unsatisfactory results. He continued to have frequent syncopal spells and was house-bound up to the time of referral to the University Hospital

On examination, the diagnosis of scleroderma was evident from the taut shiny skin over the face, dorsum of hands and fingers. His pulse rate was 30 beats per minute, and blood pressure was 110/80 mmHg. Irregular cannon 'a' waves were noted in the neck veins. There was no cardiomegaly and no evidence of heart failure.

Investigations revealed normal renal function (Creatinine 0.9 mg/100 ml), erythrocyte sedimentation rate and no lupus erythematosus cells or antinuclear antibodies were detected. A chest radiograph revealed a normal sized heart and normal pulmonary parenchyma. An electrocardiogram confirmed the presence of complete heart block (Fig. 3). Barium swallow examination was normal.

A skin biopsy revealed the epidermis to be atrophic with loss of the rete ridges. Collagen bundles in the dermis were swollen and sweat glands and hair follicles were infrequent. Mild perivascular lymphocytic infiltration was noted. These changes were consistent with the diagnosis of systemic sclerosis.

A permanent epicardial demand pacemaker was implanted on 22nd June 1979 with satisfactory results. He was free of his giddy spells and gradually

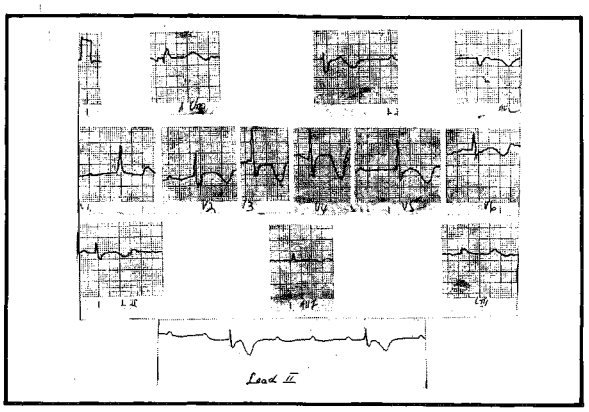

Fig. 3 Case 2: ECG showing complete heart block.

was able to lead a fairly active life. He was followed up in the cardiac clinic two months after implantation and has since continued to be free of giddy spells and syncopal attacks.

DISCUSSION

That patients with scleroderma may have heart disease is well known for over a century. [10] However, it was only in 1943 that Weiss *et al,* [5] in a study of 9 cases, convincingly demonstrated that scleroderma heart disease is a distinct clinical and pathologic entity. Since then however, the nature, prevalence and functional significance of myocardial disease in scleroderma has been hotly debated. Some studies minimized or denied the clinical significance of the myocardial lesions, [3,4,11] Sackner *et al,* [11] in a study of 65 patients with scleroderma concluded that "Scleroderma heart disease" is an uncommon clinico-pathological entity. On the other hand Bulkley *et al,* [12] in a study of 52 autopsied patients concluded that myocardial progressive systemic sclerosis is a distinct entity with relatively frequent occurrence which may lead to arrhythmias, congestive heart failure, angina pectoris with normal coronary arteries and sudden death.

The most common manifestation of scleroderma heart disease is a chronic adhesive pericarditis. [11,13] Heart failure, due to primary myocardial sclerosis or secondary to pulmonary hypertension and fibrosis and systemic hypertension with renal failure is another common manifestation of the disease. Arrhythmias and electrocardiographic conduction disturbances are frequent. [9, 14] While some workers consider that structural abnormalities in the centres of impulse formation and conduction disturbances, [15] others conclude that morphologic abnormalities within the conduction system are difficult to attribute to scleroderma per se. [16]

Complete heart block is not a common manifestation of scleroderma heart disease. Windesheim [14] in a study of ninety patients with acrosclerosis and scleroderma noted none with this abnormality. Escudero [9] found only one case of complete heart block in a study of 60 patients with scleroderma. In common with three of the patients with scleroderma and complete heart block reported in the literature, [1,7,9] death in our first patient was sudden. As with the experience elsewhere, treatment of congestive heart failure in our patient with scleroderma heart disease was unsatisfactory.

Our second patient's prognosis must remain guarded. Although a satisfactory heart rate has been achieved by pacing, the likelihood of continuing myocardial fibrosis will always raise the possibility of myocardial failure in the future.

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