Case reports

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Progressive systemic sclerosis with complete heart block

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A case of progressive systemic sclerosis is reported. The patient presented with chest pain and an abnormal chest radiograph. Later developments included cardiac involvement with complete heart block. The heart block was successfully managed with an implanted pacemaker.

Cardiac involvement was first reported in systemic sclerosis by Matsui (1924) and became widely known after the work of Weiss *et al.* (1943). The complication of complete heart block was described by East and Oram (1947), but it was not until 1970 that a case treated with an implanted pacemaker was reported by Barr *et al.* A second case successfully managed by pacing is described.

Case report

The patient, a farmer's wife aged 42, presented in February 1968 with a variable chest pain which had begun during an influenza-like illness in December 1967. She failed to respond to several courses of antibiotics and her chest radiographs throughout 1968 showed varying degrees of basal shadowing and a high right hemidiaphragm; her ESR was 41 mm/h (Westergren). At the time of referral she had several stigmata of systemic sclerosis including Raynaud's phenomenon, sclerodactyly, a small mouth, with difficulty biting on an apple, and facial telangiectasia. The chest expansion was 1 in (2.5 cm) at the level of the nipples and there were bilateral basal crackling rales. No cardiac abnormality could be detected.

The diagnosis of systemic sclerosis was supported by the radiographic appearances of asymmetrical basal fibrosis without pleural involvement and with normally rounded diaphragms. Pulmonary function studies showed a restrictive pattern with alveolar-capillary block without evidence of obstructive airways disease. The electrocardiogram showed sinus tachycardia with ascendant notching of the P waves, low voltage QRS complexes, and normal PR, QRS, and QT intervals. Treatment was started with prednisolone in reducing doses, decreasing to 10 mg/day one year later. Out-patient pulmonary function tests showed relentless de-

terioration, with a vital capacity of 2100 ml falling to 1200 ml four months later. The forced expiratory volume in one second remained normal. The DCO was 9.43 ml/min/mmHg.

Severe congestive cardiac failure with recurrent syncope and a pulse rate of 38/min necessitated her urgent admission to another hospital in January 1974. She had had no chest pain since the start of steroid therapy in 1968. An electrocardiogram showed complete heart block with deep Q waves in leads II, III, aVF and V2-5, with left axis deviation and QRS duration 0.12 s. An endocardial electrode was inserted transvenously via the right cephalic vein and a Devices Demand Pacemaker, Type No. 3821 RC, was implanted in the anterior abdominal wall by means of a right transverse paraumbilical incision; the threshold was 2 volts. The cardiac failure was effectively controlled by the pacemaker, frusemide 80 mg, spironolactone 25 mg four times a day, effervescent K 24 mEq thrice daily, and digoxin 0.25 mg daily. Since her discharge the exercise tolerance has improved, though she remains very dyspnoeic on exertion. Her blood pressure is 140/70 mmHg.

Discussion

This case illustrates many of the problems of progressive systemic sclerosis, including a rare cardiac complication. The patient's vague and non-specific chest pain, the presenting symptom, is a common finding in this condition (Weiss *et al.*, 1943; Goldman, Young, and Knox, 1954). No satisfactory explanation has been proposed for its origin (Oram and Stokes, 1961) and there are many possible causes (Weiss *et al.*, 1943; Goldman *et al.*, 1954; Banerjee and Mukherjee, 1966). In this case there was no dermal involvement of the chest wall and the pain

disappeared when steroid therapy was started. The basal rates are typical, as are the radiographic appearances (Oram and Stokes, 1961; Bauer, 1955; Hayman and Hunt, 1952). There may be no correlation between them, the disturbance of pulmonary function, the degree of pulmonary hypertension, or the post-mortem changes, which are almost always present (Goldman *et al.*, 1954; Oram and Stokes, 1961; Hayman and Hunt, 1952; Leinwand, Duryee, and Richter, 1954).

Weiss *et al.* (1943), the first to document the pathological changes in the heart, found patchy myocardial fibrosis unrelated to vascular changes and consisting of delicate collagen fibres with numerous fibroblasts. Some but not all of the isolated myocardial fibres within the fibrous lesions were very vascular. Others have confirmed this observation (Piper and Helwig, 1955), and James (1974) has reported a small necropsy series and has drawn attention to the small blood vessels. The small arteries may show mural and intimal fibrosis, endothelial proliferation, medial hyperplasia, and fibrinoid necrosis in the absence of lesions in the larger arteries. Focal myocardial fibrosis was also present and was considered to be the result of fibroplasia of the supporting connective tissue, in particular that of the conducting system. A case with multiple aneurisms of the right coronary artery has been described (Chaithiraphan *et al.*, 1973). Others have added valvular lesions (Leinwand *et al.*, 1954) and verrucous endocarditis (Spühler and Morandi, 1949).

The conducting system in a case of complete heart block has been described by Lev *et al.* (1966), who found mainly fibrous replacement of the sub-endocardial third of the myocardium with a dearth of Purkinje fibres; those present showed degenerative changes which Lev *et al.* related to the widespread abnormality and to fibrous replacement of the bursa of the AV node. Conduction defects are predictably common (Escudero and McDevitt, 1958). They may be transient (possibly the result of partial resolution of the mucinous oedema observed by Sackner *et al.* (1964)) or more usually progressive. Oram and Stokes (1961) found a 51 per cent incidence of all conduction defects. Their series included 49 per cent with arrhythmias and 34.7 per cent with changes that were interpreted as ischaemic. The electrocardiogram may simulate myocardial infarction (Windesheim and Parkin, 1958; Beigelman, Goldner, and Bayles, 1953). The incidence of first degree heart block varies from 15 to 20 per cent in the larger series (Oram and Stokes, 1961; Bianchi *et al.*, 1966; Sackner *et al.*, 1964). Second degree heart block is less frequent, a two per cent incidence is quoted by Sackner *et al.* Third degree

heart block is rare: several cases have been observed to progress to complete heart block. East and Oram (1947) recorded a case progressing from first degree to third degree heart block over a period of 13 months. Lev *et al.* (1966) have reported a detailed histological study in a patient who progressed from right bundle branch block to complete heart block in two years and suffered from intermittent 2:1 block for one year. The case described by Barr *et al.* (1970) deteriorated in four years to episodic Mobitz Type II Block and then to established block with Stokes-Adams attacks.

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