Case reports

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Ventricular tachycardia—an unusual presenting symptom of scleroderma

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A case of localized scleroderma with the unusual presenting feature of ventricular tachycardia has been described. The relevant reports of the electrocardiographic patterns seen in this disease have been reviewed: ventricular tachycardia does not appear to have been previously reported. In this patient there was a good response to intravenous propranolol.

Since the earliest description of cardiac involvement in scleroderma by Heine (1926) and the subsequent emphasis on scleroderma heart disease by Weiss *et al.* (1943), a number of publications concerning the clinical features and pathological changes in this disease have appeared. The electrocardiographic abnormalities with or without clinical evidence of cardiac involvement in scleroderma have been reviewed in detail by Windesheim and Parkin (1958) and Escudero and McDevitt (1958). There is, however, no mention in these reports of the occurrence of ventricular tachycardia in scleroderma. We are now reporting a case of scleroderma which came under observation because of an episode of ventricular tachycardia.

Case report

A 48-year-old man of normal build was admitted to hospital as an emergency with the presenting complaint of palpitation of 2 hours' duration. A similar episode had been experienced by the patient 25 days earlier. His pulse was 190/minute, with blood pressure of 70/40 mmHg. The first heart sound was of variable intensity on cardiac auscultation. There were pigmented and depigmented patches on the face and tightening of the skin on the forehead. The skin on the distal parts of the fingers and toes was also thick and taut. On inquiry the patient admitted to having had Raynaud's phenomenon in the fingers, especially in cold weather. The rest of the clinical examination was essentially normal.

Investigations

Investigations revealed a normal leucocyte count, a hypochromic normocytic anaemia, with normal blood urea, blood sugar, serum sodium, and potassium. The serum bilirubin was 0.6 mg/100 ml; serum cholesterol,

thymol turbidity, cephalin-cholesterol flocculation tests, serum aspartate aminotransferase, alkaline phosphatase, prothrombin time, and index were all normal, but the serum alanine aminotransferase was 42 Karmen units (normal being below 30). Bromsulphalein excretion test showed 13.3 per cent (normal 6%) retention of dye after 45 minutes, but the liver biopsy was inconclusive.

The electrocardiogram showed ventricular tachycardia (Fig. 1). The chest x-ray was normal; no cervical rib was seen. X-rays of the hands revealed tufting of some of the distal phalanges of the hands with extraosseous calcification in left middle finger (Fig. 2). Oesophagus was normal on barium swallow. Skin biopsies from the face and a finger were consistent with the diagnosis of scleroderma (Fig. 3).

The patient's tachycardia did not respond to intravenous procainamide or lignocaine, but sinus rhythm was restored after 1 mg of propranolol intravenously (Fig. 4). There was no further development during a follow-up period of six months. He continued treatment with prednisolone, oral propranolol, and isoxsuprine hydrochloride.

Discussion

Ventricular tachycardia has been mentioned as an unusual feature of such diseases as sarcoidosis (Chamovitz, Culley, and Carlson, 1962) and myxoedema (Hansen, 1961). Scleroderma, a collagen tissue disorder, may remain circumscribed for a long time before visceral involvement becomes apparent. In progressive systemic sclerosis, the cardiac involvement may not be present initially, but becomes evident terminally in the majority of patients. Piper and Helwig (1955) detected cardiac involvement both clinically and on necropsy in 28 out of 31 patients (90%). Sometimes, however, this may be the presenting feature of cardiac

FIG. 1 A 12-lead electrocardiogram showing ventricular tachycardia.

FIG. 2 X-ray of the hands showing tufting of some of the distal phalanges of the hands with extraosseous calcification in the left middle finger.

FIG. 3 Microphotograph of the skin biopsy confirming the diagnosis of scleroderma (H and E $\times 280$).

FIG. 4 A continuous electrocardiographic recording showing lack of response to procainamide and lignocaine, with a response to propranolol.

involvement in scleroderma, as was seen in 3 out of 9 cases described by Weiss *et al.* (1943) and 2 out of 31 cases described by Piper and Helwig (1955). In our case, the presenting feature was ventricular tachycardia while involvement of the integument was minimal and was only noted on careful examination. Raynaud's phenomenon, though it had occurred, was also not conspicuous and only revealed by direct inquiry. The possibility of some early involvement of the liver in this case is suggested by the abnormal bromsulphalein test.

Cardiac involvement in scleroderma is either primary or secondary to pulmonary or renal pathology (Rodnan, Schreiner, and Black, 1957). Though the involvement may be pancardiac in both the atria and ventricles, the major effects are seen in the myocardium. Patchy or transmural fibrosis of the myocardium and minimal coronary sclerosis may be seen. There is, however, no correlation between the degree of coronary sclerosis and the amount of myocardial fibrosis (Piper and Helwig, 1955). The myocardial changes on necropsy are often more than anticipated on clinical grounds.

In view of the pathological changes in the heart in scleroderma, it is easy to understand the various electrocardiographic changes described. Windesheim and Parkin (1958) and Escudero and McDevitt (1958) reviewed these in detail. Such alterations as slurred QRS complexes of low amplitude, notching of the P wave, evidence of ventricular hypertrophy or atrial enlargement, subepicardial or subendocardial ischaemia, bundle-branch block patterns, AV block, intraventricular conduction defects, non-specific ST-T wave changes, ectopic beats, and atrial fibrillation have been described. Gupta and Singh (1963) also observed the occurrence of par-

oxysmal atrial tachycardia in this disease. We wish to add ventricular tachycardia to the list of rhythm disturbances which may occur in scleroderma. The pathogenesis of the ventricular tachycardia is considered to be due to myocardial fibrosis and ischaemic changes.

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