

Cardiac involvement in chronic polymyositis

P. G. Lynch

From the Department of Neuropathology, University of Manchester

This is a report of a 30-year-old male patient with a six-year history of chronic polymyositis. There was no family history of muscular or cardiac disease. Three years after the onset of the illness the patient developed congestive cardiac failure with evidence of complete heart block. Treatment with steroids failed to arrest the course of the disease. Necropsy showed abnormalities in the proximal and distal limb muscles, in the sternomastoid muscles, and in the laryngeal muscles. There were extensive fibrotic lesions in the chambers of the heart and in the conducting system.

Severe cardiac involvement is a rare manifestation of chronic polymyositis. Oppenheim (1899) appears to have been the first author to describe cardiac lesions in this condition. Subsequently Sheard (1951) pointed out that polymyositis involving the pericardium occurred in some cases. Radermecker and van Bogaert (1955) described cardiac lesions in 2 cases of acute polymyositis, and Barnard, Rankin, and Robertson (1960) described changes in the heart of a 16-year-old West African girl who died during the acute phase of polymyositis. Garcin *et al.* (1955) stated that clinical evidence of pericarditis or of myocardial involvement was uncommon in polymyositis, though electrocardiographic abnormalities were sometimes found. Walton and Adams (1958) found clinical and electrocardiographic evidence of pericardial involvement in 4 out of 40 cases classified as polymyositis.

The case to be described in this paper illustrates many of the clinical and pathological features of chronic polymyositis.

The biopsy and postmortem histological findings in clinically affected and non-affected voluntary muscle will be described. The results of a detailed examination of the heart including the conducting system will also be presented.

Clinical features

The patient, a 27-year-old warehouseman, was admitted to Manchester Royal Infirmary in April 1965 complaining of weakness of the legs and of slight difficulty in swallowing, of 3 years' duration. The symptoms began in December 1962 when he noticed that his legs seemed heavy after walking a

short distance. He also found difficulty in standing erect and in rising from a recumbent position. The patient had also noticed occasional slight difficulty in swallowing, but had never suffered from double vision nor from drooping of the eyelids. In 1960 he had an attack of unexplained haematuria associated with backache. There was no past history of rheumatic fever, venereal disease, nor of exposure to toxic chemicals. There was no family history of muscular disease.

Physical examination The patient weighed 57 kg. There was slight uniform enlargement of the thyroid gland. **Cardiovascular system:** the pulse showed no abnormalities. Blood pressure 120/65 mm Hg. On auscultation, the first and second sounds at the apex and in the second right interspace were very quiet; in the third left interspace, especially in the expiratory phase, there was a short but loud systolic murmur followed by a clear second sound. No abnormalities were found in the *alimentary* and *respiratory* systems. Examination of the *central nervous system* showed no cranial nerve abnormalities. The ankle-jerks were absent. The plantar responses were flexor. Sensation was normal. Examination of the *musculo-skeletal* system showed no evidence of myotonia or fasciculation. There was slight wasting and weakness of the right and left supraspinatus and infraspinatus muscles. There was weakness of the right and left deltoid muscles and bilateral winging of the scapula. Muscle tone in the upper and lower limbs was slightly diminished on both sides. There was a diffuse wasting of both legs, almost equal above and below the knee. The right and left gluteus maximus muscles were wasted. The hamstring muscles and the ileo-psoas muscles showed much weakness. Skin lesions and muscle tenderness were absent.

Investigations *Haematology:* Hb 15.2 g./100 ml.; leucocytes 9,300/cu. mm. (normal differential); ESR (Wintrobe) 2 mm./hr.

Biochemistry: Glucose tolerance test normal; serum protein bound iodine 5.4 $\mu\text{g./100 ml.}$; radio-iodine test uptake in thyroid at 2 hours - 18 per cent (normal); serum cholesterol 185 mg./100 ml.; blood urea 25 mg./100 ml.; serum Na^+ 139 mEq/l.; serum K^+ 4.8 mEq/l.; serum aldolase 12.2 Bruns units/l. (normal range 3-10 Bruns units/l.); serum creatine phosphokinase 5.3 International units/l. (normal range up to 1 International unit/l.); 24-hour urine creatine 430 mg.; 24-hour urine creatinine 773 mg. (normal range 1-2 g./24 hr.). Volume of 24-hour urine specimen 1330 ml. SGOT 31 units/ml. (normal range 6-40 units/ml.); SGPT 11 units/ml. (normal range 5-35 units/ml.). Serum albumin 4.2 g./100 ml. Serum globulin 3.2 g./100 ml.

Chest x-ray: No abnormalities detected.

Nerve conduction studies: No abnormalities detected.

Electromyography: Most muscles examined showed considerable spontaneous fibrillation potentials and positive sharp waves. Short duration polyphasic volitional action potentials were also noted.

Muscle biopsy (left deltoid muscle): Histological examination showed myopathic changes (Fig. 1).

Treatment The patient was given prednisone and neomercazole.

Progress In November 1965 the patient developed congestive cardiac failure which responded initially to treatment with digoxin and diuretics. In February 1966 he was found to have a pulse rate of 40/min. and an electrocardiogram showed evidence of complete heart block. Treatment was started with frusemide 'lasix' and isoprenaline ('saventrine'). There was an initial favourable response to this regimen, but attacks of heart block recurred later. In June 1966 cardiac catheterization studies gave findings consistent with a constrictive type of cardiomyopathy. Electrocardiography showed right bundle-branch block. A biopsy specimen was taken from the left supraspinatus muscle. Histological examination showed myopathic features, and there was no evidence of inflammation. No antibody against human heart muscle could be found in the patient's serum using the fluorescent antibody technique.

After discharge from hospital the patient's condition deteriorated. An internal cardiac pacemaker was inserted on 17 October 1968, but the patient suddenly collapsed and died on 19 October 1968.

Pathological features

Macroscopical Necropsy was performed 48 hours after death. The body weighed 37 kg., height 180 cm. There was symmetrical wasting of the limb muscles. Skeletal muscle tissue in the affected regions was pale fawn and of a soft consistency. There was uniform enlargement of the

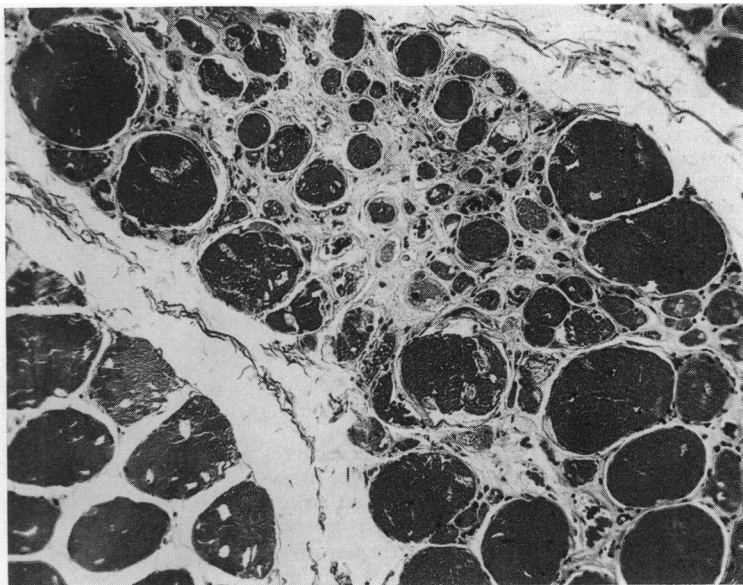
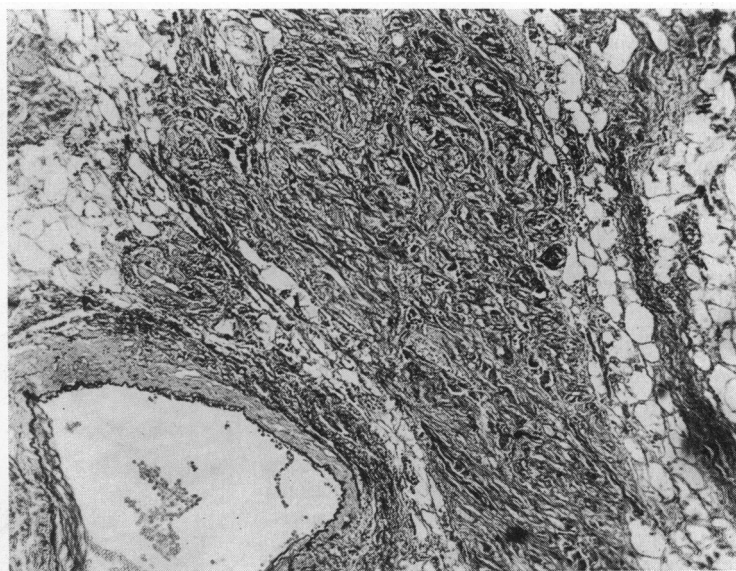


FIG. 1 Deltoid muscle, showing myopathic changes. (Masson's trichrome. $\times 110$.)

FIG. 2 Heart. Sino-atrial node showing fibrosis. The artery to the node can be seen on the left in the lower part of the field. (Anderson's Victoria Blue and Van Gieson. $\times 110$.)



thyroid gland. The lungs showed areas of collapse in their lower lobes and there was atrophy of the cortex of both suprarenal glands. The heart weighed 440 g. There was a fibrinous pericarditis. Cardiac pacemaker leads were stitched in the myocardium of the left ventricle. The average thickness of the left ventricular myocardium was 18.0 mm. (normal being 12 mm.). The cut surfaces of the left ventricle revealed several small foci of greyish-white tissue in the myocardium of the left ventricle. The heart valves, coronary ostia, and coronary arteries showed no abnormalities. The aorta, pulmonary arteries, superior vena cava, inferior vena cava, and pulmonary veins showed no abnormalities. The brain, spinal cord, and peripheral nerves showed no naked-eye abnormalities.

Microscopical Heart: There was extensive replacement fibrosis of cardiac muscle fibres in the right atrium and right ventricle. The left atrium showed mild epicarditis only. The left ventricle showed scattered areas of replacement fibrosis of cardiac muscle fibres. The cardiac valves and coronary arteries showed no abnormalities.

Conducting system: The sino-atrial node arteries were thick-walled but distinct and had good lumens. The entire node showed conspicuous fibrosis and was virtually devoid of specialized myocardium (Fig. 2). Adjacent myocardium of the right atrium showed replacement fibrosis. The AV node showed no abnormalities. The node artery branches were greatly thickened by fibrous tissue. The thickening appeared to affect the tunica media. The bundle of His was prominent and showed some fibrosis towards the bifurcation. The left bundles were seen only proximally near to their origin. More distally the fascicles ran into subendocardial fibrosis of the interventricular septum (Fig. 3), in which scattered Purkinje cells could be identified. The right bundle was readily followed, but in places it was completely fibrous and contained no muscle fibres (Fig. 4). The interatrial and interventricular septa showed moderate replacement fibrosis without cell reaction. The intramuscular vessels appeared normal.

No histological abnormalities were found in the brain, spinal cord, or dorsal root ganglia. The skin showed no abnormalities. The thyroid gland showed the pattern of a diffuse colloid goitre.

Voluntary muscle: Myopathic changes were seen in the sternomastoid, sartorius, peroneus brevis, and posterior cricoarytenoid muscles. No abnormal cellular infiltrates and no blood vessel changes were noted in these muscles.

Discussion

Despite the fact that there was no evidence of inflammation in any of the muscles examined at necropsy, nor in any of the three muscle biopsy specimens taken from the patient during his illness (one of these biopsy specimens was taken at another hospital in 1964), there were many clinical similarities to poly-

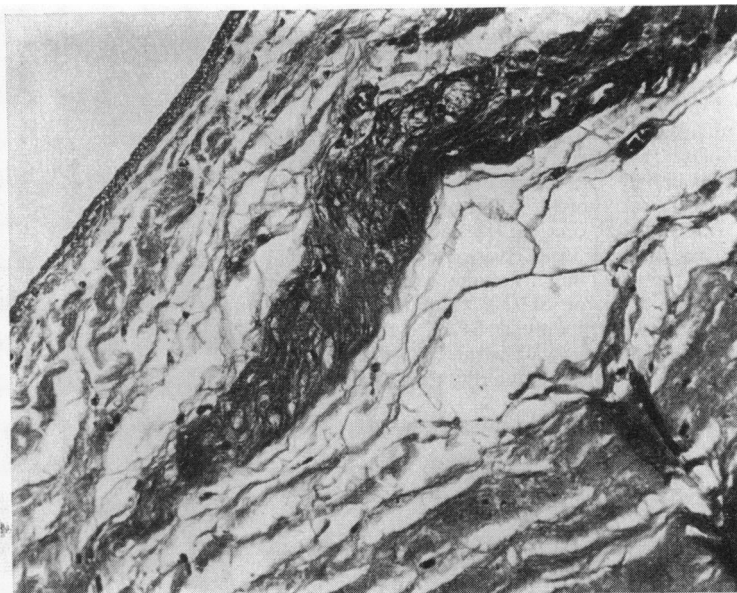
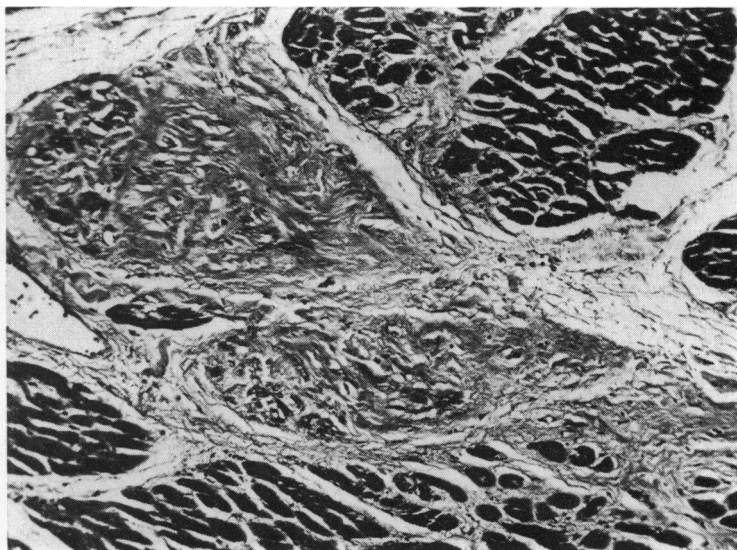


FIG. 3 Heart. Left bundle showing fascicles running into an area of subendocardial fibrosis. (Haematoxylin and eosin. $\times 190$.)

FIG. 4 Heart. Right bundle showing almost total replacement fibrosis of the conducting tissue. (Haematoxylin and eosin. $\times 110$.)



myositis. These features were the rapid progress of the disease, the symmetrical distribution of muscular weakness in the upper and lower limbs, and the presence of dysphagia and dysarthria.

The relation between the changes found in the myocardium and voluntary muscles is obscure. It is tempting to postulate a common aetiological agent affecting both tissues. The absence of lymphocytes in both tissues and the lack of serum antihuman heart antibody seems to exclude a disorder of immune mechanisms. The poor response to steroid therapy is also in keeping with this view.

It is worth remembering the importance of examining the voluntary muscles in all cases of cardiomyopathy. The converse is also true.

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