CASE REPORTS

PULMONARY HYPERTENSION IN DERMATOMYOSITIS

BY

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Dermatomyositis is the term used to designate a group of cases characterized predominantly by non-suppurative inflammation and degeneration of muscle, including the myocardium, together with similar involvement of overlying or unrelated parts of the skin and hypoderm. Clinical features include fever, erythematous skin lesions, and muscle fatigability, weakness, pain, and tenderness, which may progress to atrophy and fibrotic contractures. Œdema, particularly over affected muscles and in the orbits, a toxic alopecia, and high levels of creatinuria also commonly occur.

Publications on dermatomyositis have failed to exclude two other confusingly similar conditions: (a) the polymyositic form of polyarteritis nodosa, an acute fulminating or self-limiting condition responding well to cortisone or A.C.T.H., in which cutaneous pleomorphism, œdema, muscle tenderness and leucocyte counts are all excessive, and (b) an acute "dermatomyositis" which is symptomatic of concurrent carcinoma or lymphoblastoma and is potentially reversible with successful treatment of the neoplasia. True dermatomyositis has an insidious—rarely subacute—onset and runs a slow inexorable course with remissions and relapses towards crippling and grotesque contractural deformities with late poikiloderma, diffuse calcinosis, and the Raynaud phenomenon with sclerodactyly. Moreover, it is a systemic disease and through degenerative and proliferative changes in the connective tissue and in the arteriolar walls, cases may occur of primary involvement of serous membranes, the joints, the alimentary tract, the spleen, the liver, and the lymph glands.

Though there are eminent authorities like Dowling (1952) who maintain that dermatomyositis and diffuse systemic sclerosis are one and the same disease, there is a weight of opinion that there are essential differences in the ætiology, clinical picture, and courses of these conditions. Scleroderma, which should be regarded as a symptom and discarded as a synonym, has recently been admirably reviewed by Cullinan and Harper (1953).

Case Report

I.G., aged 34, a railway signalman, was first admitted to hospital in February, 1949. Prior to the summer of 1948 he had been in excellent health and capable of strenuous activity. In October, 1948, after a month or so of vague malaise and lassitude he developed a bright red rash on his brow and cheeks, with transient orbital swelling. By Christmas, 1948, when the rash had all but faded, pain and tenderness developed in the region of the joints of his fingers, wrists, elbows, and knees. Before admission, he became fevered, and severe pain and later weakness successively affected the muscles of his shoulder girdle, arms, buttocks, legs and, finally, the dorso-lumbar region, so that he became quite helpless. At this time an X-ray of the chest showed no abnormality of heart or lungs—cardio-thoracic ratio 0.4 (Fig. 1). A generalized alopecia followed and he began to have intermittent attacks of the Raynaud phenomenon in his fingers. Remissions and relapses of his joint and muscle symptoms followed and when admitted again to hospital in January, 1951, he was found to be grossly emaciated (weight 76 lb. as compared with 150 lb. in 1948) with generalized muscle atrophy and fibrotic contractural deformities of elbows and wrists and rheumatoid appearances of his hands, with radial or ulnar deviation at the metacarpo-phalangeal joints and flexion at the proximal and hyperextension at the distal interphalangeal joints. Slight poikiloderma of the skin of his brow and sternal area was present. Investigations then showed: E.S.R., 95 mm./hour; plasma proteins total, 6 g. per 100 ml. (albumen 2.8, globulin 3.2, ratio 0.87/1); urinary creatine, 350 mg./24 hours; 17 ketosteroids, 5 mg./24 hours; skin biopsy, "collagen degeneration" with scanty perivascular round cell reaction in the corium; muscle biopsy, focal degenerative changes of variable severity, no notable vascular lesion other than scattered lymphorrhages; X-ray, slight cardiac enlargement with prominence of the pulmonary artery and of its main branches, cardiothoracic ratio 0.5; electrocardiogram, marked right ventricular

FIG. 1.—X-ray of chest in June, 1949, showing no abnormality of the heart or lungs.

FIG. 2.—X-ray of chest in June, 1954, showing enlargement of the heart and marked prominence of the pulmonary trunk and its main branches. The peripheral vascular shadows, particularly in the upper lobes, are less evident than usual.

hypertrophy. Short courses of A.C.T.H. were given during early 1951 without any objective improvement. Later in that year his illness was marked by two episodes of acute pericarditis and by the development of generalized subcutaneous calcinosis with osteoporosis and the extrusion of chalky bodies from indolent ulcers over bony prominences. At Christmas, 1951, vigorous neck vein pulsation was first noted. Late in 1952 he had recurrent ascites without ankle oedema and a right pleural effusion, but he slowly became ambulant and left hospital in June, 1953. His physical activities were considerably restricted by his locomotor disabilities which made it difficult for him to appreciate his undoubted cardiac and respiratory insufficiency. He returned for review in May, 1954, and from the muscular and cutaneous aspect appeared to have reached a stage of stability. Faint erythema and thickening of the skin of his brow, sternal area, and the extensor surface of his forearms were present without binding to underlying structures. Generalized fibrous muscle atrophy was evident affecting noticeably the facial muscles and masseters to give a cadaveric facies, and the chest muscles to limit much expansion. His fingers were long, thin, and blue, and still showed rheumatoid deformities. Nodules of calcium were palpable subcutaneously in many places, particularly about the tendons. His cardiac and respiratory systems were investigated in detail at this time.

Respiratory System. Expansion of the chest was severely restricted. There were scattered rales at both lung bases and a pleural rub in the right axilla. Tests of respiratory function were done by Dr. C. D. Needham.

<i>Lung Volumes</i>				<i>Observed</i>	<i>Predicted</i>
Vital capacity	..	..	..	2650 ml.	4200 ml.
Inspiratory capacity	..	..	..	1290 ml.	2700 ml.
Expiratory capacity	..	..	..	1360 ml.	1500 ml.
Resting tidal air	..	..	..	650 ml.	—
Residual volume	..	..	..	2270 ml.	2200 ml.
Total lung capacity	..	..	..	4920 ml.	6200 ml.
Helium mixing	..	..	..	98%	>60%
Functional residual capacity	..	..	..	3630 ml.	3350 ml.

Ventilatory Measurements. (a) Timed vital capacity at 2 sec. was 2550 ml., i.e. 96 per cent of total vital capacity (normal greater than 95 per cent). (b) Maximum breathing capacity was 84 litres a minute (normal average is 124 litres a minute).

There was, therefore, a great diminution in inspiratory capacity but emphysema was excluded because (1) maximum breathing capacity was only moderately diminished, (2) there was no air trapping as shown by (a) above, (3) intrapulmonary mixing was above average, and (4) functional residual capacity was normal.

X-ray screening of the chest showed that the right ventricle was enlarged and the pulmonary artery segment prominent. Pulsation of the heart seemed normal. There was no sign of calcification of the pericardium and no evidence of enlargement of the left atrium. Cardio-thoracic ratio was 56 per cent (Fig. 2). Tomograms confirmed the enlargement of the main pulmonary artery and its primary branches. The peripheral vascular shadows in both upper zones were less than normal. No other abnormality was seen in the lungs.

Cardiovascular System. There was no cyanosis or oedema. Marked venous pulsation was seen 2 cm. above the sternal angle as the patient reclined at 45°. Two waves could be distinguished: a jugular venous pulse tracing, with simultaneous electro- and phono-cardiogram, showed that the earlier of the two waves occurred during atrial systole, indicating hypertrophy of the right atrium, and the latter during the ejection phase of ventricular systole. A positive systolic wave similar to the second wave has been described in tricuspid incompetence (Muller and Shillingford, 1954). The blood pressure was 120/90. The apex beat was felt in the fifth left intercostal space 10 cm. from the mid sternal line and right ventricular pulsation in the third and fourth left interspaces. A systolic murmur (grade III) was loudest at the tricuspid area. In the same area there was a rather clicking sound in diastole. The phonocardiogram shows that this sound occurred 0.18 sec. after the beginning of the second sound, simultaneously with the base of the Y descent of the jugular venous pulse, i.e. later than a third heart sound is usually found. The systolic murmur extended throughout systole and was continuous with a short early diastolic murmur, which followed immediately on a split second sound. Such an early diastolic murmur has been described in tricuspid incompetence (Messer *et al.*, 1950).

There were large P waves in leads II and aVF of the electrocardiogram and tall R waves in leads aVR and V1 (10 mm.), indicating much hypertrophy of the right ventricle. Cardiac catheterization showed that there was hypertension in the pulmonary artery and right ventricle. The mean pressure readings, all expressed in mm. of mercury above a plane 5 cm. behind the manubrium sterni, were (1) pulmonary "capillary" 4 mm. Hg; (2) main pulmonary artery 52 mm. Hg; (3) right ventricle 36 mm. Hg; (4) right atrium 5 mm. Hg, and (5) superior vena cava 5 mm. Hg. The oxygen content of samples of blood from the right heart, estimated by the method of Roughton and Scholander (1943), were (1) pulmonary artery 13.2 vol., (2) right atrium 12.9 vol., and (3) superior vena cava 12.9 vol., each per 100 ml.

A femoral artery blood sample contained 18.5 vol. per cent of oxygen. Hæmoglobin was 14.3 g. per 100 ml. and arterial oxygen saturation 96 per cent. Oxygen consumption was 210 ml. a minute and cardiac output 4 litres a minute.

Total pulmonary resistance, calculated from the formula,

$$\text{Resistance (dynes/sec./cm.}^{-5}\text{)} = \frac{\text{Mean pulmonary artery pressure (mm. Hg)} \times 1332}{\text{Cardiac output (ml./sec.)}}$$

was 1040 dynes/sec./cm.⁻⁵. Pulmonary arteriolar resistance, calculated similarly from

$$\frac{\text{Mean pulmonary artery pressure (mm. Hg)} - \text{pulmonary "capillary" pressure (mm. Hg)} \times 1332}{\text{Cardiac output (ml./sec.)}}$$

was 960 dynes/sec./cm.⁻⁵. Although the validity of these calculations, particularly of the second, is debatable, it seems fair to conclude that the pulmonary vascular resistance was greatly increased and that most of the increase arose in the pulmonary arterial tree.

Several unsuccessful attempts were made to obtain arterial blood during exercise to determine if there was interference with oxygen diffusion. The procedure was found to be impracticable because the patient's severe locomotor disability did not allow him to perform an adequate amount of exercise.

Discussion

There is no doubt that this patient presented the typical history and clinical features of chronic dermatomyositis and that he had at no time presented the characteristic hidebinding of "scleroderma."

The outstanding findings in the cardiovascular system were hypertension in the pulmonary artery and right ventricle, and great hypertrophy of the right heart. The normal pulmonary

"capillary" pressure excluded a lesion in the left side of the heart (such as mitral stenosis or constrictive pericarditis) as the cause of the pulmonary hypertension. There was no sign of a left-to-right intracardiac shunt or other evidence of a congenital defect of the heart; also, cardiac enlargement developed along with the dermatomyositis.

The only evidence of a valvular lesion pointed to tricuspid incompetence, which cannot explain pulmonary hypertension of this degree. Fibrosis of the myocardium leading to congestive cardiac failure has been described in scleroderma (East and Oram, 1947). None of the findings in the present case indicated that the cardiac dysfunction was due to myocardial damage of this kind. Emphysema was excluded by the respiratory function tests.

The only adequate explanation of the pulmonary hypertension and hypertrophy of the right heart appears to be a process of narrowing or occlusion of the smaller branches of the pulmonary arterial tree, the result of degenerative and proliferative changes in the vessel walls. There is some support for this view in the contrast between the enlarged main pulmonary artery branches and the somewhat inconspicuous peripheral vessels seen on the X-ray of the chest. This would then appear to be an example of the "vascular fibrosis" in the lungs described by Spain (1950). Right ventricular hypertrophy associated with such obliterative disease in the pulmonary arteries has been previously demonstrated in pulmonary schistosomiasis (Shaw and Ghareeb, 1938) and in carcinomatous infiltration of the lung vessels (Morgan, 1949).

Wood (1952) mentions polyarteritis nodosa and disseminated lupus erythematosus as causes of block of the pulmonary circulation. In a recent comprehensive review of pulmonary changes in diffuse collagen diseases (Ellman and Cudkowicz, 1954) the authors suggest that the pulmonary arterial changes in this group are due to interference with tissue nutrition by deprivation of the only arterial blood supply available to the supporting structures of the lungs—the bronchial arteries. It seems at least as probable that the pulmonary arteries are affected primarily.

Summary

A case of dermatomyositis with severe pulmonary hypertension and hypertrophy of the right heart is described.

It is concluded that the pulmonary hypertension and hypertrophy of the right heart are the result of degenerative and occlusive changes in the smaller branches of the pulmonary arterial tree.

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