Myotonic dystrophy complicated by peripheral vascular disease

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Summary
A case of myotonic dystrophy complicated by peripheral vascular disease is described. No obvious explanation was found to account for this. A review of the literature revealed a few similar cases. The possibility that disordered peripheral circulation may be a rare feature of myotonic dystrophy is proposed.

Introduction
Myotonic dystrophy was first described in this country by Batten and Gibb in 1909. In 1911, Griffith drew attention to the associated cardiological complications. Since then, many abnormalities outside the neuromotor system such as multiple endocrinopathies have been described in this condition. The following case was complicated by peripheral vascular disease.

Case report
A 40-year-old man was admitted to a surgical unit with a 3-week history of increasing pain in the right foot. In the past year he had experienced pain in both calves on walking half a mile, and in addition, had been falling frequently, his legs ‘giving way’. He had had bilateral cataract extractions one year previously. There was no history of diabetes mellitus or medication. He was an unmarried, unemployed labourer, a non-smoker but a drinker of at least two pints of beer thrice a week. There was no relevant family history available.

On examination, he had the characteristic features of myotonic dystrophy (Fig. 1), including sternomastoid, facial and distal limb weakness and wasting, plus percussion myotonia. There was also hypogonadism and gynaecomastia. The pulse was irregular, the blood pressure 125/90 mmHg. Peripheral pulses were absent below the femorals bilaterally. The right foot was the cooler and there was ischaemic gangrene affecting the great toe (Fig. 2).

Investigations
Full blood count, urea and electrolytes, glucose...
profile, fasting lipids, creatine kinase and chest X-ray were normal. Erythrocyte sedimentation rate (ESR) 17 mm/hr. Liver function tests revealed raised serum aspartate and alanine aminotransferase, alkaline phosphatase and gamma glutamyl transferase concentrations. Serum IgG 6.9 g/l (normal 7–19). Serum luteinizing hormone and follicle stimulating hormone were elevated and testosterone levels were subnormal.

Discussion

Sporadic cases of disordered peripheral circulation in myotonic dystrophy have been published. Waring, Ravin and Walker (1940) in a review of 13 cases found 10 complaining of cold hands and feet with cyanosis of varying degrees; this is however a common finding in peripheral nerve and muscular disease. Leinwand (1948) reported a 48-year-old man, a smoker, who complained of right calf claudication and was without pulses below the right femoral. Another case was presented by Mahoudeau et al. (1956) of a 37-year-old male who had no right radial or dorsalis pedis pulse. Variable loss of other pulses occurred and thrombo-embolism secondary to arrhythmias was one hypothesis to explain this. More recently, 'small vessel disease' in the shape of Raynaud's phenomenon has been noted (Agrawal et al., 1975).

It is probably justifiable to propose tentatively, that abnormalities in the peripheral circulation are a rare complication of myotonic dystrophy.

Acknowledgment

The authors wish to thank Mrs F. Bickerton for her secretarial work.

References


