Calf hypertrophy following paralytic poliomyelitis

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A 41-year-old man developed an acute illness at the age of 9 months during which, following a viral illness with headache, he developed severe weakness and wasting of the limbs of the left side. After several months he began to recover, such that he was able to walk at the age of 2 years and later was able to run, although he was never very good at sports. He had stable function until the age of 18 when he began to notice greater than usual difficulty lifting heavy objects. By the age of 25 he was noticing progressive difficulty walking due to weakness of both legs, and he noticed that the right calf had become larger. The symptoms became more noticeable over the course of the next 10 years and ultimately both upper as well as both lower limbs had become noticeably weaker.

On examination there was wasting of the muscles of upper and lower limbs on the left, and massively hypertrophied gastrocnemius, soleus and tensor fascia latae on the right. The calf circumference on the right exceeded that on the left by 10 cm (figure 1). The right shoulder girdle, triceps, thenar eminence and small muscles of the hand were wasted and there was winging of both scapulae. The right quadriceps was also wasted. The wasted muscles were also weak but the hypertrophied right ankle plantar flexors had normal power. The tendon reflexes were absent in the lower limbs and present in the upper limbs, although the right triceps was reduced. The remainder of the examination was normal.

Figure 1 The patient's legs, showing massive enlargement of the right calf and wasting on the left

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Questions

- 1 What is the nature of the acute illness in infancy?
- 2 What is the nature of the subsequent deterioration?
- 3 What investigations should be performed?
- 4 What is the differential diagnosis of the cause of the progressive calf hypertrophy?

Answers

QUESTION 1

An acute paralytic illness which follows symptoms of a viral infection with or without signs of meningitis is typical of poliomyelitis. Usually caused by one of the three polio viruses, it may also occur following vaccination and following infections with other enteroviruses.¹ Other disorders which would cause a similar syndrome but with upper motor neurone signs would include acute vascular lesions, meningoencephalitis and acute disseminated encephalomyelitis.

QUESTION 2

A progressive functional deterioration many years after paralytic poliomyelitis is well known, although its pathogenesis is not fully understood.² It is a diagnosis of exclusion; a careful search for alternative causes, for example, orthopaedic deformities such as osteoarthritis or worsening scoliosis, superimposed neurological disorders such as entrapment neuropathies or coincidental muscle disease or neuropathy, and general medical causes such as respiratory complications and endocrinopathies.³

QUESTION 3

Investigations revealed normal blood count and erythrocyte sedimentation rate and normal biochemistry apart from a raised creatine kinase at 330 IU/l (normal range 60–120 IU/l), which is commonly seen in cases of ongoing denervation. Electromyography showed evidence of denervation in the right APB and FDI with polyphasic motor units and complex repetitive discharges, no spontaneous activity in the left calf and large polyphasic units in the right calf consistent with chronic partial denervation. Motor and sensory conduction velocities were normal. A lumbar myelogram was normal. Magnetic resonance imaging (MRI) scan of the calves is shown in figure 2.

Figure 2 Axial T₁ weighted MRI scan (TR 588 ms, TE 15 ms) of the calves, showing gross muscle atrophy and replacement by adipose tissue on the left, and hypertrophy of the muscles on the right, with only minor adipose tissue deposition

QUESTION 4

The differential diagnosis of the progressive calf hypertrophy is given in the box.

Causes of calf muscle hypertrophy

Chronic partial denervation

- radiculopathy
- peripheral neuropathy
- hereditary motor and sensory neuropathy
- spinal muscular atrophy
- following paralytic poliomyelitis

Neuromyotonia and myokymia

- Isaac's syndrome
- generalised myokymia
- neurotonia
- continuous muscle fibre activity due to: chronic inflammatory demyelinating polyradiculopathy, Guillain Barre syndrome, myasthenia gravis, thymoma, thyrotoxicosis, thyroiditis

Muscular dystrophies

Myositis

Infiltration

- tumours
- amyloidosis
- cysticercosis

Discussion

Calf enlargement may occur due to muscle infection such as in cysticercosis,⁴ infiltration by tumour or amyloidosis⁵ or by inflammation, as in myositis.⁶ It is well known that it may occur in muscular dystrophies, particularly Duchenne and Becker types. True muscle hypertrophy has been shown to arise in a variety of neuromuscular disorders such as radiculopathy,^{7–10} peripheral neuropathy,^{12–14} spinal muscular atrophy^{15–17} and also in disorders of continuous muscle activity such as myotonia congenita, dystonia and Isaac's syndrome.^{18,19} In these circumstances muscle biopsy has shown that there is hypertrophy and hyperplasia of both group I and group II muscle fibres.^{8,10,20,21} There is both experimental and clinical evidence that group II fibre hypertrophy occurs in response to an increase in muscular work load,²² whilst group I fibre hypertrophy is thought to occur only when there is chronic partial denervation. Hence, following paralytic poliomyelitis in which there is chronic partial denervation with re-innervation, there may be group I fibre hypertrophy of muscles which are partially denervated due to muscle stretch and group II fibre hypertrophy in fibres with normal innervation which are exposed to greater work loads imposed by atrophy of other denervated fibres.

Three previous reports of patients with calf muscle enlargement following paralytic poliomyelitis have been described^{21,23,24}; this was attributed to deposition of adipose tissue without muscle hypertrophy in two cases. In this case we have provided MRI evidence that whilst fat infiltration of the denervated muscle had occurred, hypertrophy of residual muscle had taken place in the stronger leg, and this is likely to have caused the calf enlargement.

It would also appear that degenerative disease of the knee on the weaker side plays an important role, particularly *genu recurvatum*.

Progressive joint deformity has recently been implicated in the pathogenesis of late functional deterioration following previous paralytic poliomyelitis.³ This probably results in a greater than normal stress placed on the gastrocnemius on the other side in order that the patient may continue to walk, with the result that it hypertrophies.

Final diagnosis

Progressive functional deterioration after paralytic poliomyelitis.

Keywords: poliomyelitis; late functional deterioration

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A healthy patient who suddenly developed a foot-drop

Gus Plaut

A 76-year-old man had taken up gardening. A few days after a particularly tiring day he suddenly developed a right-sided foot-drop. There were no other muscle weaknesses. He had not used chemical garden sprays. His medical history included treatment for asthma with inhaled steroids and a successful resection of the colon for carcinoma.

He was suspected to have a metastatic tumour in the upper sacral or lower lumbar region, affecting the sciatic nerve, and was referred to the local oncology clinic. X-Rays and an abdominal computed tomography scan showed no abnormality, apart from minor osteo-arthritis of the vertebral column.

Questions

- What further investigations would you advise?
- What is the likely diagnosis?
- What treatment is indicated and what is the prognosis?

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