Bilateral Calf Hypertrophy Following Post-traumatic Cauda Equina Lesion

Muneera Al Bunyan, MRCP(UK); Peter Hudgson, FRCP

We report a 45-year-old male, epileptic, who sustained trauma to his back following a seizure episode. This was complicated by cauda equina lesion with partially recovering paraparesis. Progressive asymmetrical gross enlargement of the patient’s legs, particularly calf muscles, was observed at subsequent follow-up. The differential diagnosis and pathogenesis of muscle hypertrophy in such a case is discussed.

Keywords: Bilateral calf hypertrophy. Cauda equina lesion. Muscle hypertrophy.


Calf hypertrophy is a well-recognized feature of several widely differing neuromuscular disorders, notably Xp21 myopathies, spinal muscular atrophies, other forms of chronic partial denervation and even heredofamilial ataxia, in addition, a familial syndrome in which generalized and often gross muscle hypertrophy (hypertrophia muscularum vera) occurs in the absence of any other evidence of either primary muscle cell degeneration or denervation. There is considerable doubt as to the pathogenesis of calf enlargement in such cases and, certainly, genuine hypertrophy of individual muscle fibres does not appear to be responsible in all cases although there is clear-cut histopathological evidence of this phenomenon in some instances. In this paper, we report a 45-year-old male with a traumatic cauda equina lesion affecting bladder and sexual functions for over 21 years. Although he recovered partially in terms of motor and sensory functions, he gradually developed gross asymmetrical hypertrophy of calf and other lower limb muscles. The likely pathogenesis of this man’s muscle hypertrophy will be discussed and the relevant literature reviewed.

Case Report

The patient was a 45-year-old Saudi male, a known epileptic whose seizures were well controlled at the time of presentation by phenytoin (400 mg) in a single dose at night. Twenty-one years earlier, he had had a seizure in which he fell and injured his back, sustaining fractures of his dorso-lumbar spine and had developed what was described as ‘incomplete paraplegia’. He recovered satisfactorily in terms of sensation in his lower limbs but he remained incontinent of urine and impotent. In addition, gradual enlargement of both calves was noted at successive hospital visits, the left side being consistently larger than the right.

Neurological examination at his most recent review showed gross hypertrophy of calf and thigh muscles (Fig. 1a, b) involving the left side more than the right. However, his lower limbs were weak with proximal muscle power grade 4 at hip extendors and grade 3 plus at the knee, planter flexion of left foot was grade zero on the Medical Research Council (MRC) scale. The knee jerks were hypo-active and his ankle jerks were absent with flexor planter response. Sensory testing demonstrated diminished appreciation of pin-prick and light touch in the S2–S3 dermatomes and the anal sphincter was lax. No abnormalities were found in the upper limbs and cranial nerve territory. He could walk unaided, albeit with some difficulty.

From the Division of Neurology (38), King Khalid University Hospital, PO Box 7805, Riyadh 11472, Saudi Arabia M. AL BUNYAN
From the Newcastle General Hospital, Newcastle, UK P. HUDGSON
Received August, 1993. Accepted for publication in final form October, 1993.
The initial diagnosis on the basis of the above history and findings was post-traumatic quada equina lesion. Becker Xp21 muscular dystrophy was also considered. His serum creatinine kinase (CK) was 368 IU (upper limit 210 IU). The electromyographic findings study showed a few fibrillations and positive sharp waves with frequent fasciculation potentials, giant motor unit potentials and partial recruitments which were compatible with chronic partial denervation. Muscle biopsy performed twice for left gastrocnemius muscle failed to demonstrate any significant histological or histochemical abnormalities apart from variation in fibre diameter and increased perimysial and endomysial connective tissue but no group atrophy or fibre type grouping was seen in histochemical sections (Fig. 2a, b). Investigations also included plain X-ray for lumbosacral spines which showed post-traumatic degenerative changes in the form of wedge deformity of L1 with large osteophytic outgrowth at antero-lateral margin of L1 crossing to D12/L1 and L1/L2 disc spaces.

Computerized tomographic scan (CT) for the legs showed gross enlargement of gastrocnemius muscle and increased amount of fatty tissue posteriorly and medially in the left leg (Fig. 1b). Ascending urethrogram and cystoscopy were also performed to assess bladder function. Thickening and trabeculation of the bladder was revealed with distension at the junction of the bulbous and penile-uretha.
Figure 2a. H/E stain for muscle biopsy from right gastrocnemius not showing significant morphological changes. X108.

Figure 2b. ATPASE stain at pH 4.6. No histochemical changes of fibre-type grouping. X108.

Discussion

Calf muscle hypertrophy is a well-recognized finding in a number of chronic neuromuscular disorders at both macroscopic and microscopic levels as described above. The pathogenesis of this phenomenon is uncertain and what constitutes genuine hypertrophy on the one hand and 'pseudo-hypertrophy' on the other is not well understood. Swash & Schwartz in their monograph suggest that pseudo-hypertrophy is marked by pathological 'firmness' of the muscle belly, a tendency to 'ridging' on contraction and, paradoxically, significant weakness. The clinical features are presumed to be associated with muscle fibrosis. This is in line with the concepts of 'pseudo-hypertrophy' as enunciated by Duchenne (1868) and ERb (1884) in their accounts of early-onset X-linked recessive myopathy.

However late-onset X-linked recessive myopathy may be associated with pathologically documented true hypertrophy, antedating the appearance of degenerative abnormalities. In addition, individual hypertrophied fibres (hyaline or otherwise) are seen as a matter of routine in biopsy sections from chronic myopathies of all kinds.

On the other side of the coin, it is clear that both micro- and macrohypertrophy are regular features
of a variety of chronic denervating processes. Certainly microhypertrophy is well-recognized histologically in well-compensated denervating disorders such as spinal muscular atrophy and the presence of ‘giant’ (reinnervated) motor units constitutes an electrophysiological sine qua non for making the diagnosis of compensated partial denervation. Further to this, there are a number of single case reports describing unilateral macrohypertrophy of the calf musculature in peripheral denervation, notably in S1 radicular lesions. We believe that our patient falls into the latter category although, as far as we are aware, his is the first reported case with bilateral calf hypertrophy. It seems clear that he sustained incomplete lesions of the conus, the cauda equina or both as a consequence of his injury. However, we find it hard to understand why this man’s calves, have become so huge (visibly bigger than the previous examples illustrated in the literature), particularly in the absence of any histological/histochemical evidence of denervation and/or reinnervation in biopsies from his gastrocnemius. It is perhaps surprising that the many experimental models of denervation/reinnervation described during the last two decades have not shed more light on the genesis of this phenomenon.

Acknowledgement

We are grateful for the services of Mr Habib Atta for his technical assistance and processing the slides.

References