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Clinical Study

# Mechanisms of upper limb amyotrophy in spinal disorders

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ABSTRACT

Upper limb amyotrophy may occur as an indirect consequence of various spinal disorders, including ventral longitudinal intraspinal fluid collection, Hirayama disease and high cervical cord compression. We present patients who suffer from each of these and review the literature on the three conditions with emphasis on the pathogenesis of amyotrophy. We propose that pathology some distance from the lower cervical spinal cord may affect normal venous drainage, resulting in venous congestion and reduced perfusion pressure which, in turn, could result in anterior horn cell dysfunction in all three disorders.



## 1. Introduction

There are many possible causes for muscle wasting in the upper limbs, but the mechanism of wasting is usually obvious, with signs and symptoms fitting patterns consistent with focal injury to a nerve root, peripheral nerve, or the anterior horn cells.

Occasionally patients present with upper limb amyotrophy with no apparent direct cause of lower motor neuron dysfunction. Some have associated disorders which may cause amyotrophy indirectly. Three such conditions are high cervical spinal cord compression, ventral longitudinal intraspinal fluid collections (VLISFC) and Hirayama disease.

We present case reports exemplifying these three disorders. We hoped that review of these new cases in the context of the literature might shed some light on how amyotrophy develops in these conditions.

## 2. Case reports

### 2.1. Patient 1

Patient 1, a 48-year-old man, had a 17 year history of progressive wasting of the muscles in his upper limbs. Initially, wasting was noted in the intrinsic muscles of the right hand, and several years later the wasting had extended to involve the deltoid, supraspinatus and infraspinatus but largely spared the triceps. Biceps and brachioradialis jerks were absent on the right but other reflexes were normal as was sensation. MRI of the spine showed an extensive ventral intraspinal, extradural arachnoid cyst from C4 to T12. A shunt from cyst to peritoneum was inserted through a low thoracic hemilaminectomy. The fluid in the cyst was under high pressure. No clinical improvement resulted. Seven years on, all right arm muscle groups showed Medical Research Council grade 3 to 4 power with sensation preserved. Occasional fasciculation in the left biceps was also noted, and another 5 years later there was left upper limb muscle weakness. On examination, scattered fasciculations were seen throughout the upper limbs. Power in the right upper limb was grade 2 for deltoid and biceps, grade 1 for supraspinatus and infraspinatus, grade 4 for triceps, and grade 3 to 4− in the intrinsic muscles of the right hand. All of these weak muscles were wasted. Left upper limb power was grade 4 throughout, except in the hand where power was grade 4+. Upper limb reflexes were barely attainable. The lower limb examination revealed slight reflex asymmetry but down-going plantars bilaterally. There was now some patchy sensory loss over the right arm, trunk and leg. The rest of the examination was unremarkable. Upper and lower limb somatosensory evoked potential testing was normal. Serial MRI scans showed stable appearance of an anterior epidural cyst extending from C2–T9, bilateral mild to moderate foraminal stenosis at C5/6 and C6/7, and (in the most recent sets of scans only) increased T2-weighted signal from C3 to C5/6, affecting the anterior grey matter bilaterally (Fig. 1). Recent CT myelography suggested that cerebrospinal fluid (CSF) was filling the cyst between T5 and T9. Surgical exploration, centred at T7, failed to confirm a localised site of CSF leakage. Dural repair material was inserted into the epidural space over four levels, but without any obvious improvement in upper limb function post-operatively.

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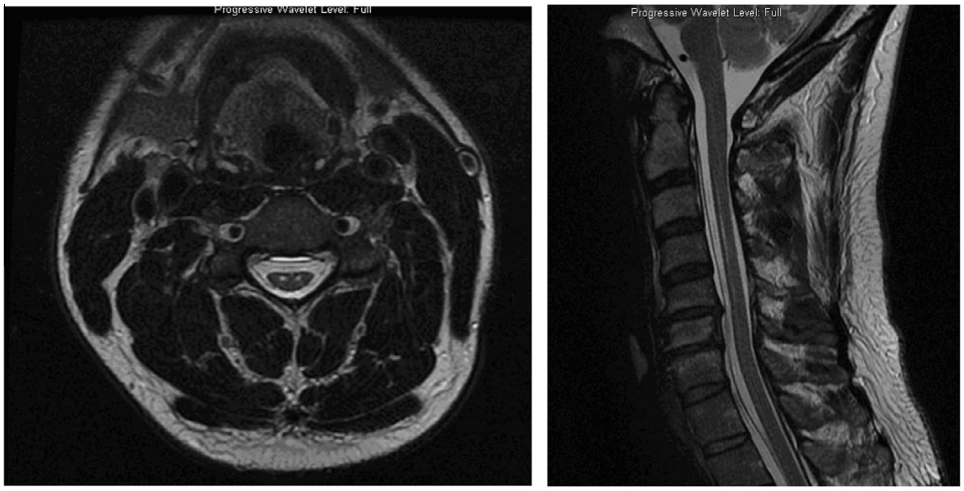


**Fig. 1.** Patient 1. (A) Axial T2-weighted MRI of the cervical spine showing evidence of signal change in the region of the anterior horns which is out of keeping with the minimal compression exerted by the cyst at this level. (B) Sagittal T2-weighted MRI of the cervical spine showing signal change in the cervical cord between levels C3–C7, which is superior to the maximal compression of the cyst which is predominantly in the thoracic spine.

### *2.2. Patient 2*

Patient 2, a right-handed 29-year-old man of Italian descent, had no family history of primary muscle or nerve disorders. He first presented at age 18, with an 18 month history of progressive weakness in the upper limbs. Initially there was weakness in extension and abduction of the right fourth and fifth digits. Subsequently he developed cramping and marked weakness of all intrinsic muscles of the right hand. Twelve months later he noticed weakness in the left hand, again commencing in the fourth and fifth digits. Over the next 6 months, there was weakness in his triceps muscles, associated with cramping and wasting and also a tingling sensation with neck flexion. He described worsening limb weakness with cold weather. Examination revealed wasting of the hands and distal forearm muscles, with well preserved proximal forearm extensors. Finger abductors and extensors were assessed as grade 1 power on the right and grade 3 on the left, and finger flexors were grade 4+ on the right and grade 5 on the left, except for flexor pollicis longus which was weaker bilaterally. The rest of the neurologic examination was normal. Electromyography (EMG) revealed evidence of chronic partial denervation in the muscles supplied by C7–T1. MRI of the cervical spine revealed short segment spinal cord atrophy between mid C4 to mid C6 levels, the right side more affected than the left; further imaging in a flexed posture demonstrated anterior displacement of the spinal cord (Fig. 2). There was bilateral paramedian intramedullary high signal on T2-weighted images consistent with myelomalacia. He had an array of blood tests, which were unremarkable. CSF showed normal protein and glucose levels, and three white cells. Enzyme testing for a variety of inherited disorders, including Tay-Sachs disease and Sandhoff disease, was normal. A diagnosis of Hirayama disease was made. During subsequent follow-up visits over the next 10 years, no further progression or improvement was observed.

### *2.3. Patient 3*

Patient 3, a 72-year-old right-handed man, presented with numbness of the medial three fingers in both hands, and a tendency to drop things. This was not associated with neck pain or radicular pain. Relevant medical history included known C5/6 disc prolapse, a previous mid-thoracic vertebral fracture and chronic lower back pain. On examination, the patient had marked wasting of the intrinsic muscles of the hands bilaterally, with occasional fasciculations. Upper limb neurological examination revealed: grade 4+ power bilaterally for shoulder abduction, adduction and wrist flexion; grade 4 for elbow flexion and extension, wrist extension, and finger flexion and extension; and grade 3 power in the finger abductors and adductors. Neck flexion did not cause further weakness. Reflexes were brisk but normal bilaterally in the upper limbs, and sensation was slightly decreased in the C7 to T1 dermatomes bilaterally, with the right side more affected than the left. Examination findings in the cranial nerves and lower limbs were unremarkable. Nerve conduction studies found electrophysiological evidence of chronic denervation in the intrinsic muscles of the hand bilaterally, with preserved sensory potentials. MRI revealed degenerative cervical canal stenosis maximal at C5/6 and C6/7 with bilateral myelomalacia at C6 extending to the level of the body of C7. This T2-weighted signal hyperintensity was maximal in the grey matter on both sides (Fig. 3). No intraspinal venous abnormality was seen. The patient underwent a C5–C6 and C6–C7 anterior cervical discectomy and fusion procedure, and subsequently improved. The most recent examination revealed symmetrical findings in the upper limbs with normal power at deltoid and biceps and minimally reduced power of triceps, wrist extension and finger flexion. Finger extension at the metacarpophalangeal joints was mildly weak (grade 4+). The interossei were markedly wasted and weak (grade 4−) with fasciculations noted in first dorsal interosseous on both sides. The thenar groups showed moderate wasting and weakness (grade 4). Upper limb reflexes were all brisk normal. There was a subjective alteration of sensation in the hands but no sensory loss to pin-prick, light touch, vibration or joint position sense. Lower limb findings were normal except for an equivocally extensor right plantar reflex.

## 3. Discussion

### *3.1. VLISFC*

Hader and Fairholm in 2000 observed gradually progressive bilateral upper limb amyotrophy in patients with previous nerve root avulsion (two patients) or spinal flexion injury (one patient) in the distant past [1]. Imaging showed large anterior extradural CSF collections extending from the upper cervical to the lower thoracic and lumbar levels in each patient. These authors mentioned a possible parallel with the well-recognized phenomenon