Short report

Denervation in wasted hand muscles in a case of primary cerebellar ectopia without syringomyelia

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SUMMARY Wasting of hand muscles and electromyographic changes of denervation in a case of primary cerebellar ectopia *without* evidence of syringomyelia is described. It is suggested that the wasting is due to anterior horn cell damage secondary to grey matter venous obstruction at high cervical cord levels.

Hand muscle wasting has been described in patients having primary cerebellar ectopia *with* syringomyelia.¹ It has also been described with EMG evidence of denervation in cases of high cervical cord compression.²⁻⁵ This communication reports denervation changes in wasted hand muscles in a case of primary cerebellar ectopia with no evidence of syringomyelia.

Case report

A 19-year-old trainee bricklayer presented with a 5 month history of weakness and wasting in the muscles of the left hand. He also noticed tingling in the left fingers and left side of the chest when bending his neck. There were no symptoms referable to either cranial nerves or the other limbs.

On examination, abnormalities were limited to the left upper limb. Weakness (MRC grade 4 to 4+) was found in the thenar, hypothenar, interosseous, wrist extensor and flexor, extensor digitorum communis and triceps muscles. Wasting was also noted in these muscles, particularly in the interossei. There were no objective sensory abnormalities in any of the limbs. Reflexes were preserved and symmetrical with flexor plantar responses. There was no evidence of Horner's syndrome, spinal deformities or bruits, short neck or skin lesions.

Plain radiographs of the cervical spine were normal. Metrizamide myelography demonstrated herniation of the cerebellar tonsils down to C2 level and atrophy of the mid and lower cervical cord (fig). There was no variation in cord size with changes in patient position. Computed tomography

with metrizamide confirmed the cord atrophy and showed no visible contrast within the cord substance on delayed scans (up to 24 hours). The cerebral ventricles were of normal size.

EMG, using a concentric needle electrode, demonstrated fibrillation, fasciculation and large polyphasic motor units of long duration in the left thenar, first dorsal interosseus, wrist flexor and extensor, extensor digitorum communis, biceps and triceps muscles. Similar but less severe changes were also seen in the right biceps and first dorsal interosseous muscles. Median and ulnar motor conduction velocity, F wave latency, finger to wrist sensory action potentials and somatosensory evoked potentials to median nerve stimulation were normal in both upper limbs.

At operation, the posterior margin of the foramen magnum was removed and laminectomy of C1 and C2 was performed. Marked cerebellar ectopia was found, more pronounced on the left where the cerebellar tonsil extended down to C2 level. The cord, down to C3 level, appeared atrophic with no cystic enlargement. It was of normal consistency and was not collapsible or pulsatile. Needling of the cord was therefore judged to be unnecessary. The vertebral and anterior spinal arteries were not examined during surgery. Eighteen months after the operation, the patient reported no clinical deterioration and was back at full employment; preoperative EMG abnormalities were still evident.

Discussion

The hand muscle wasting presented in this case differs from that described previously in cases of primary cerebellar ectopia by the absence of dissociated anaesthesia or loss of reflexes in the upper limbs characteristic of syringomyelia.¹ In a series of 100 cases of syringomyelia,⁶ none presented with muscular wasting alone; all cases of wasting were associated with loss of upper limb sensitivity and reflexes.

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Fig Myelogram (metrizamide 300mg/ml, 10ml, C1-2 puncture). Supine position. The cerebellar tonsils are shown to descend to the level of C2. The sagittal diameter of the cervical cord is reduced at C5 and C6 in the absence of any narrowing of the theca or spinal canal at these levels, suggesting atrophy.

Although the radiological and operative findings described have not demonstrated any evidence of an expanded cavity, they do not exclude a small central cleft in the cord. However, it is difficult to imagine such a lesion causing this extensive and severe wasting and bilateral denervation without interrupting central fibres and producing some loss in upper limb sensibility and reflexes. Indeed a case of cerebellar ectopia which came to necropsy and was shown to have a central cleft had presented with hand numbness rather than muscle wasting.⁷

From the evidence discussed above, it is concluded that syringomyelia is a most unlikely cause for the muscle wasting in this case. High cervical cord compression has long been associated with hand muscle wasting which was attributed to anterior horn cell damage.²⁻⁴ In the case presented here, the clear denervation changes demonstrated in the absence of any radiological evidence of a root lesion and the normal sensory and F wave studies suggest a similar pathology. The findings suggest that these are due to remote effects of cervical cord compression by the prolapsed cerebellar tonsils.

The mechanism leading to anterior horn cell damage is controversial. Ischaemia due to occlusion of the anterior spinal artery by the compressing mass is unlikely because of the asymmetry of clinical signs and the symmetry of anterior spinal artery distribution.⁸ Stagnation ischaemia secondary to obstruction of cervical cord venous drainage has also been suggested.³ This is supported by experimental high cord compression in monkeys which can produce hand muscle weakness.⁹ Venous dilatation, grey matter haemorrhages and anterior horn cell loss were found on both sides of the cervical cord but maximal at C7, C8 level ipsilateral to the compression. The asymmetrical distribution of these pathological changes, which is similar to that of the clinical signs, and their restriction to the grey matter have been attributed to separate venous drainage systems for white and grey matter which are independent on the two sides. However, EMG studies were not performed on the weak muscles to link the pathological and clinical findings.

From the available evidence and the cranial direction of cervical cord venous flow,¹⁰ this study suggests that the muscle wasting presented is due to anterior horn cell damage secondary to obstruction of the venous drainage of grey matter at the level of the cerebellar tonsillar herniation.

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