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Clinical Improvement of Monomelic Amyotrophy After Avoidance of Sustained Neck Flexion

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To the Editor:

Management of monomelic amyotrophy (Hirayama disease) is controversial and includes clinical observation, use of a cervical collar, and cervical decompression and fusion.¹ Cessation of symptom progression with persistent arm weakness is the most common outcome.^{1,2} Some have documented regression of symptoms with cervical collar use or after cervical spine surgery.² We describe clinical improvement in a young man with monomelic amyotrophy who was asked to avoid prolonged neck flexion without using a cervical collar.

A 19-year old man presented with 8 months of progressive asymmetric arm weakness and atrophy. There was no preceding trauma. He denied pain or sensory loss. When flexing his cervical spine, he described a “pulling” sensation originating in his lower cervical/upper thoracic region and radiating into his right fifth digit. He was starting college and when studying in bed with his head propped in flexion the hypothenar eminence and fifth digit would twitch or cramp. His medical history was unremarkable. His examination revealed atrophy in the right thenar and hypothenar eminences, fasciculations in the right first dorsal interosseous (FDI), and asymmetric weakness in muscles of the C7/C8/T1 myotomes [right elbow extensors, wrist extensors, finger flexors, finger abductors, and thumb abductors all medical research council (MRC) grade 4] but was particularly prominent in the right finger extensors, most dramatically in digits 3 and 4 (MRC grade 0 in digits 3 and 4; grade 3 in digit 5; and grade 4 in digit 2). The rest of the neurologic examination was normal. Nerve conduction studies were unremarkable. Electromyography demonstrated abnormalities in muscles innervated by C7/C8/T1 anterior horn cells, worse on the right. Fibrillation potentials and positive sharp waves were seen in the right FDI, extensor digitorum communis (EDC), and flexor carpi radialis. In these muscles, there were either no voluntary motor unit potentials (EDC) or reduced recruitment of chronically remodeled units (FDI, flexor carpi radialis). Similar changes were also seen in the right abductor pollicis brevis and triceps, and in the left FDI, abductor pollicis brevis and EDC. Needle examination of the legs and thoracic paraspinals was normal. Cervical spine magnetic resonance imaging

revealed the loss of normal cervical lordosis but no disc herniation or cord signal change. Computed tomography myelogram demonstrated indentation on the right anterior cervical cord at the C5-6 vertebral level in flexion views (Fig. 1). The patient was advised to avoid all activities involving substantial neck flexion and to sleep with his neck in a neutral position. A year later, the patient reported improved strength in the right finger extensors, finger flexors, and wrist extensors (MRC grades 4, 5, and 5, respectively). These changes in strength resulted in significant improvement of his ability to type and perform manual tasks. After 3 years of follow-up, he has remained stable and continues to avoid neck flexion.

The radiographic findings and clinical course argue in favor of a compressive etiology of monomelic amyotrophy in this patient. Conservative management was a moderately successful strategy even with radiologic signs of cervical cord compression.

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FIGURE 1.

Computed tomography myelogram. A, Axial image at C5-6 with neck neutrally aligned. B, Axial image at C5-6 with neck in flexion. Note the flattening and indentation (arrow) on the right anterior cervical cord.