

Anterior Spinal Extradural Cyst Mimicking Hirayama Disease and Amyotrophic Lateral Sclerosis

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Introduction

Hirayama disease (HD) is a focal motor neuron disorder that occurs in teenagers or young adults secondary to dynamic compression of the cervical spinal cord. Classically it manifests as insidious and progressive muscle wasting and weakness of the upper extremity, thus is often considered in the differential diagnosis of amyotrophic lateral sclerosis (ALS).¹ Spinal extradural cyst accounts for approximately 1% of spinal cord space-occupying lesions.² Here we describe a patient presenting with slowly progressive upper extremity weakness that was initially suggestive of HD or ALS, but was later found to have an anterior extradural cerebrospinal fluid collection secondary to spinal dural defect. We would like to alert readers that anterior spinal extradural cyst should be in the differential diagnosis of slowly progressive cervical motor neuron dysfunction. A brief description of this patient was presented previously.³

Case Presentation

A 47-year-old right-handed male with a past medical history of hypertension presented with progressive painless asymmetrical weakness and atrophy of bilateral upper extremities. Six years prior to presentation, he experienced sudden onset of severe mid-back pain while walking downstairs which forced him to sit down immediately. Back pain persisted for the next 24 hours; along with severe headache and gait imbalance. An evaluation by an emergency medicine physician was non-revealing. Within the next year, he began to notice a gradual worsening weakness in the right upper extremity with difficulty extending and gripping his fingers. Intermittent muscle twitching was observed in his

right hand, right upper arm and left forearm. Over the next several years, atrophy of the right pectoralis, right hand and forearm muscles became evident. There was no paresthesia or pain, bowel or bladder dysfunction, or involvement of bilateral lower extremities.

Neurologic examination at year six following symptomatic onset demonstrated asymmetrical (right more than left) atrophy of the forearm flexors, extensors, abductor pollicis brevis (APB) and first dorsal interosseous (FDI) muscles. Fasciculations were observed in the bilateral triceps, right forearm flexors and bilateral FDI muscles. Muscle strength testing was graded as follows (right/left, Medical Research Council grade): deltoid 5/5; biceps 5/5; triceps 3/4; wrist flexors 3/5; wrist extensors 4/5; FDI 3/4; abductor digiti minimi 2/4; flexor pollicis longus 4/5 and APB 4/4. Muscle strength examination in the lower extremity was normal. Bilateral triceps tendon reflexes were hypoactive while bilateral quadriceps and right Achilles tendon reflexes were hyperactive. Remainder of his neurologic examination was normal.

Electrophysiological findings

Sensory nerve conduction studies of the right upper extremity and the right median compound muscle action potential (CMAP) were normal. The right ulnar CMAP amplitude was reduced. Needle examination revealed fibrillation potentials and long-duration polyphasic motor unit potentials in the following muscles of the right upper extremity: FDI, APB, extensor indicis, pronator teres, triceps, and lower cervical paraspinals. Nerve conduction studies and needle electromyography of the right lower extremity were normal. Electrodiagnostic findings were supportive of a focal motor neuron disorder or cervical polyradiculopathy affecting the C7, C8 and T1 segments or roots. It was felt that the patient may suffer from HD or a slow variant of ALS.

Additional investigation

Cervical spine MRI with and without contrast showed minimal lower cervical cord atrophy and moderate disc herniation at the C5/6 and C6/7 levels without significant cord compression. Flexion and extension MRI showed no anterior shifting of the posterior dura or posterior venous engorgement that would be suggestive of Hirayama disease. Axial T2-weighted image showed the presence of bilateral small hyperintense lesions consistent with a “snake-eyes appearance” in the lower cervical cord (Figure 1). MRI

myelogram revealed the presence of an anterior extradural fluid collection that extended from C5 to L2 (Figure 1). CT myelogram revealed pooling of contrast in the cervical neural foremen, indicating cerebrospinal fluid leakage. Further review of the MRI showed the presence of anterior spinal cord herniation through an anterior dural defect at T2-T3 level (Figure 1).

Clinical course

Patient declined dura repair surgery. He underwent an upper thoracic epidural blood patch treatment. Follow-up exam at one year showed no further progression of his weakness and muscle atrophy. Patient noted he could run quicker, and his stamina also improved. Patient declined follow-up testing. Telephone follow-up for the next 4 year indicated a stable course without further worsening.

Discussion

The occurrence of spinal cysts is associated with surgery, trauma, neural tube defect, and arachnoiditis.⁴ Anterior extradural spinal cysts mimicking HD or ALS has been described previously.⁵⁻⁶ Rahmlow et al described a 34-year old man with a 5-year history of progressive forearm weakness and atrophy. MRI showed a longitudinally extensive anterior spinal extradural cyst extending from C2 to L1. CT myelogram revealed a slow cerebrospinal fluid leak. Patient's strength improved after receiving surgical cyst drainage and fenestration.⁵ Schmalbach et al. described

3 male patients who presented to ALS clinic with weakness and fasciculations in the upper extremities. Clinical and electrophysiological studies in these patients was in accordance with a diagnosis of possible ALS by the revised El Escorial criteria.⁷ In all 3 patients, repeat cervical spine MRI several years into the course showed the presence of anterior spinal cysts. Cyst resection in 1 patient led to clinical improvement.⁶ Authors stressed the importance of repeating cervical spine MRI later in the course for the purpose of identifying cervical extradural cyst that may have been missed with initial imaging.⁶ Like our patient, all 5 patients presented with an asymmetrical upper extremity weakness and atrophy with a symptomatic duration of 5 years or longer.⁵⁻⁶

The etiology of cyst formation in our patient could be related to a minor trauma. Anterior spinal cysts may cause motor neuron dysfunction by compressing anterior spinal arteries leading to compromised microcirculation and eventual anterior horn cell death or compressing ventral roots.⁶ MRI of the cervical spine in our patient showed hyperintense lesions with a snake-eyes appearance in the anterior cervical spinal cord. This appearance was previously described in spinal cord ischemia due to thrombosis or dissection as well as in which HD, further supports a mechanism of chronic ischemia as the cause of anterior horn dysfunction.⁸⁻¹⁰

A surgical repair was offered to our patient but he declined. Instead, a less invasive approach of epidural blood

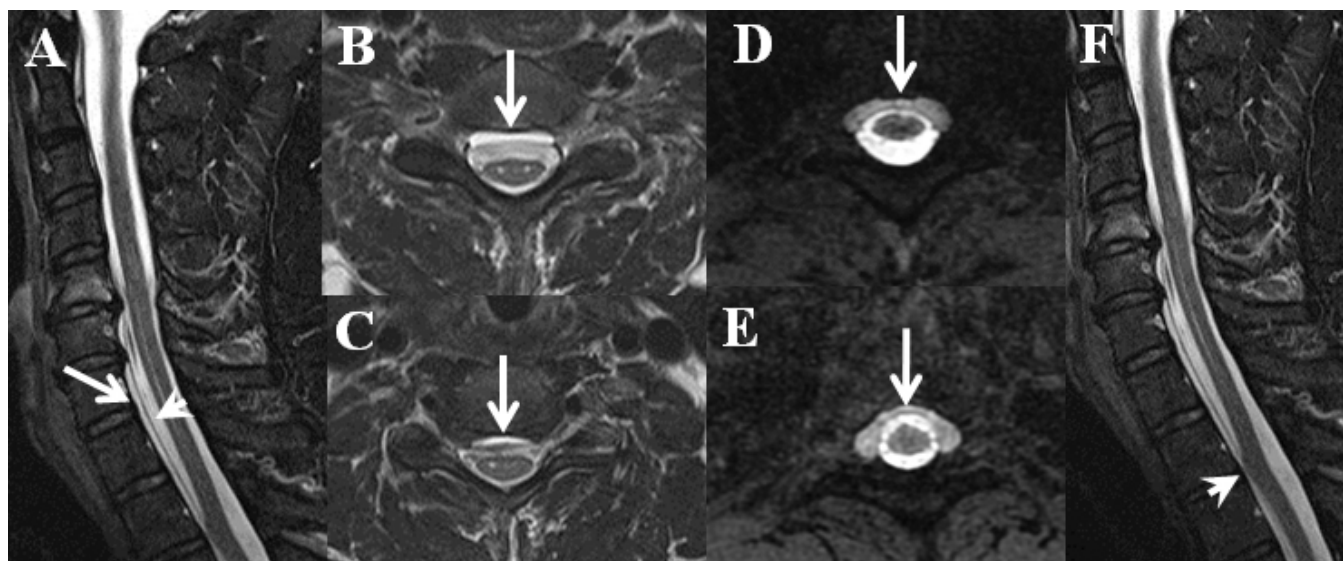


Figure 1: MRI findings. Sagittal cervical spine MRI (A) shows the presence of anterior dura (arrowhead) and a longitudinal anterior extradural spinal cyst (arrow). The extradural cyst is better visualized on the axial T2 weighted MRI sequence (arrows in B and C) and axial MRI myelogram (arrows in D and E). Hyperintense lesions in the anterior cervical cord show "snake-eyes appearance" (B and C). Arrowhead in F indicates the location of spinal cord herniation through an anterior dural defect.

patch was accepted by the patient to repair the dural defect. We were unclear whether such a treatment fixed the dural defect due to a lack of follow-up MRI. However, disease stabilization and improvement occurred in our patient following treatment, suggesting a possible efficacy.

Conclusion

Anterior extradural spinal cyst should be in the differential diagnosis of slowly progressive upper extremity weakness and atrophy of long duration, and its identification via appropriate imaging may lead to possible curative treatment.

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