

Clinical Reasoning: Transient cervical cord swelling in monomelic amyotrophy

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SECTION 1

A 9-year-old previously healthy Filipino-American boy presented to a peripheral hospital with a 2-day history of sudden-onset right arm weakness in the setting of intermittent fevers for 1 week prior to presentation. Over the 2 days prior to presentation, he lost the ability to write, grab any objects, or make a fist with his right hand, as well as the ability to lift his right arm to dress himself. Motor examination of the right arm revealed low tone, 0/5 strength for wrist extension, elbow flexion, and elbow extension, and minimal movement of all right hand interossei and lumbricals. Right trapezius strength was 5/5. Lower extremity strength was 5/5 bilaterally. Sensation to light touch, pinprick,

position sense, and vibration was intact in the affected limb. Cranial nerves 2 through 12 were grossly intact. Deep tendon reflexes were absent in the right upper extremity and normal on the left. He denied arm pain, change in sensation, recent trauma, and bladder or bowel dysfunction. The pediatric neurology team was consulted and he was admitted to the pediatric ward at the peripheral hospital for further evaluation.

Questions for consideration:

1. What are the possible etiologies for this patient's right arm weakness?
2. What would be the next step in management for this patient?

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SECTION 2

The differential diagnosis for sudden-onset right upper extremity weakness includes lesions at various levels of localization: muscle lesions (unlikely), neuromuscular junction abnormalities such as myasthenia gravis (unlikely), plexus lesions such as brachial plexitis, root lesions such as cervical radiculopathy or polyradiculopathy, spinal cord lesions such as cervical cord ischemia, transverse myelitis or monomelic amyotrophy, and brainstem or brain lesions such as intracranial hemorrhage or ischemia. Other intracranial diagnoses such as intra-axial tumor and meningioma are improbable due to the lower motor neuron findings.

MRI of the cervical spine at time of initial presentation showed swelling of the cervical cord at levels C3-C8 with no contrast enhancement (figure, A). Specifically, there was edema of both the central cord and anterior horn cells (figure, B). MRI of the brain and right brachial plexus at this time were within normal limits. EMG and nerve conduction studies at initial presentation were attempted but aborted due to patient intolerance. The patient was treated

with a 3-day high-dose steroid course followed by a 5-day IV immunoglobulin treatment course during this hospitalization. Neither treatment course resulted in any improvement in the motor examination, which was conducted daily by the peripheral hospital's attending pediatric neurologist. The patient was discharged home after 13 days and he started physical and occupational therapy for the affected limb. Repeat MRI of the cervical spine 3 months later showed complete resolution of the previous cervical cord swelling.

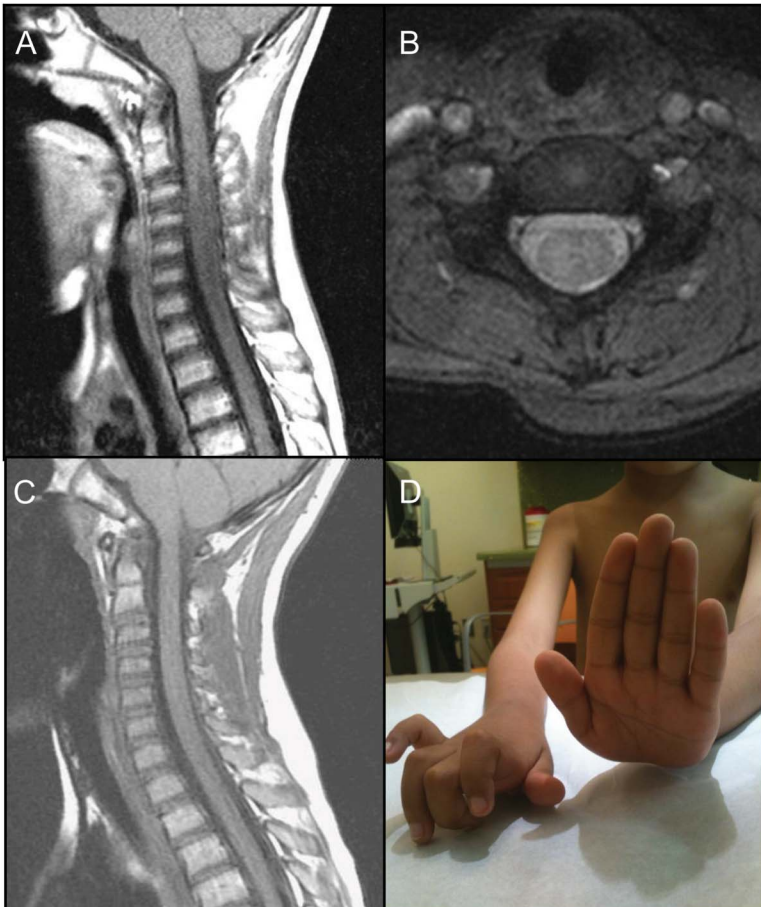
He had a follow-up examination with a pediatric neurologist at the peripheral hospital 5 months post-insult, at which point the motor examination of the right arm was significant for low tone and 0/5 strength for wrist extension, elbow flexion, and elbow extension. He had gained some function of right hand interossei and lumbricals, and he was able to make a fist and grab at that time. He was then referred to our Pediatric Neurology Clinic for a second opinion. The patient's family did not pursue the second opinion until 22 months postinsult when the patient presented to our pediatric neurology clinic.

Questions for consideration:

1. In light of the imaging findings and poor response to both steroid and immunoglobulin treatment, what is your differential diagnosis at this point?
2. What would be your next steps in management?

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Figure Cervical cord swelling and resolution in monomelic amyotrophy



Cervical cord swelling (A), including anterior horn cells (B), with resolution (C), and muscle atrophy and weakness (D).

SECTION 3

At 22 months postinsult, the patient's motor function of the right hand was mildly improved, and the patient was able to make a fist and grasp objects. Marked atrophy and hypotonia was present in all muscles of the right upper extremity and shoulder, with muscle strength graded at 1/5 in the right biceps and triceps and 2/5 in the right hand interossei, lumbricals, and wrist flexors and extensors (figure, D). There were no fasciculations. Lower extremity muscle strength was 5/5 bilaterally. There was no Babinski sign. Deep tendon reflexes were absent in the right upper extremity and 1+ throughout the left upper extremity. Sensation remained intact to light touch, temperature, and vibration throughout all 4 extremities. Repeat MRI of the cervical spine and brachial plexus 22 months postinsult were both within normal limits with no evidence of the previous cervical cord swelling (figure, C). The diagnosis of brachial monomelic amyotrophy (MMA) was made and the patient was re-referred for physical and occupational therapy as well as to a hand surgeon.

DISCUSSION MMA is a rare lower motor neuron disease that presents as a pure motor weakness of a single limb, most commonly an upper extremity. The weakness is classically most severe in the hand with atrophy of the intrinsic hand muscles. This painless disorder most frequently presents in young Asian male patients ages 15 through 25, especially those of Japanese or Indian origin. This disease is rarely seen in the United States and Europe.¹ There is currently no known cause of MMA; however, there have been reports that suggest radiation exposure, trauma, and familial inheritance as possible etiologies.² Proposed pathogenesis includes cord compression resulting in focal cord atrophy, viral infections, autoimmune processes, and changes in the microvasculature of the anterior spinal artery with neck flexion, causing ischemia to the anterior horn cells.^{3,4} Our patient may have developed MMA secondary to an infection, as he had an antecedent febrile illness. He had negative titers for cytomegalovirus, herpes simplex virus, varicella, and hepatitis C; however, he could have contracted another viral illness that he was not tested for, such as West Nile virus. His antinuclear antibody was negative, which decreased clinical suspicion for autoimmune diseases such as systemic lupus erythematosus.

Neurologic examination in patients with MMA may show hyporeflexia isolated to the affected limb. Upper motor neuron signs are usually absent, as was the case for our patient. However, there have been reports of hyperreflexia of the unaffected lower limbs without an accompanying Babinski sign.¹

Diagnosis is based on clinical history as well as neurologic examination, and definitive diagnosis may take several months. At initial presentation, the

differential diagnosis includes brachial plexitis, which presents with transient, severe arm pain followed by muscle weakness that typically resolves with time. MMA differs from brachial plexitis in that the former is painless. Transverse myelitis can occur abruptly, but often has sensory findings as well as upper motor neuron findings in the lower extremities.

Further testing such as EMG in patients with MMA will show reduced amplitudes of muscle action potentials as well as fasciculations consistent with denervation of the extremity. Muscle atrophy without other structural abnormalities may also be present on MRI or CT imaging of the affected limb. Our patient had a unique finding of isolated cervical cord swelling, including both the central cord and anterior horn cells, upon initial onset of symptoms. This cervical cord swelling was completely resolved on subsequent MRI at 3 months and 22 months postinsult; however, the progressive degeneration of the anterior horn cells resulted in persistent right upper extremity muscle weakness and atrophy.

Our patient continues to undergo standard MMA treatment, which includes physical and occupational therapy. There is no known cure for MMA. Disease progression is typically slow and usually occurs over 1–4 years following initial presentation. After this period, the weakness tends to remain stable.⁵

AUTHOR CONTRIBUTIONS

Dr. Lisa DeGregoris contributed to drafting and revising the manuscript. Dr. Murray Engel oversaw the drafting and contributed to revising the manuscript.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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