Clinical Reasoning: 63-Year-old woman with thoracic cavernous malformation and delayed worsening myelopathy

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Section 1

A 63-year-old woman presented with marked, progressive worsening of lower extremity spasticity and decreased upper extremity strength. In 1980, she developed lower extremity paresthesias and was found to have an intramedullary hemorrhagic lesion at T3. She underwent a laminectomy at T1–T4 and partial resection of a cavernous malformation (CM). Apart from a single, transient worsening of lower extremity sensory symptoms in 1985 (due to CM rebleeding), she did not experience any further exacerbations. At the time of evaluation at our institution, she had noticed worsening leg spasms and difficulty with her gait, as well as decreased muscle bulk, most pronounced in the quadriceps. These symptoms had developed over several months. She also reported worsening strength and endurance in the upper extremities, difficulties with fine motor movements, and dysphagia. She did not report new sensory deficits in her upper extremities or bowel or bladder dysfunction. On examination, she had a spastic gait but was able to walk with a cane. She had mild weakness, Medical Research Council grade 4, in the lower extremities and normal strength in the upper extremities, except for mild weakness, about 4/5 strength, in finger and wrist extension, in the interossei, and proximally. She had 3+ patellar and ankle reflexes, normal reflexes in the upper extremities, and extensor plantar responses bilaterally. Jaw jerk was not assessed. She had marked spasticity in the lower extremities with normal tone in the upper extremities. She had a T5 sensory level to temperature.

Questions for consideration:

1. Based on history and examination, what could explain worsening symptoms?
2. Which details of the patient's history are most relevant?

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Section 2

The patient's prior spine imaging showed postoperative changes, stable appearance of the thoracic CM, and spinal cord tethering at the laminectomy site. Brain MRIs done before the current exacerbation revealed stable findings, with multiple supratentorial and infratentorial CMs. An MRI of the thoracic spine was repeated and did not show any new changes. Cervical spine MRI demonstrated retrolisthesis of C5 on C6 with moderate central canal stenosis. Lack of

changes in CM appearance indicated that the new progressive symptoms were not related to it (explanation of Question 2 from Section 1).

Questions for consideration:

1. What is the most important finding on the recent MRIs?
2. Which additional diagnostic testing can further clarify the underlying etiology for the worsening symptoms?

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Section 3

The differential diagnoses to explain the patient's subacute neurologic deterioration include tethering of the spinal cord at the prior resection site, a microbleed from the known CM, a primary neurologic condition, or cervical spondylotic myelopathy from stenosis at C5–C6 superimposed on an already diseased spinal cord. A comprehensive EMG study of the upper and lower extremities was normal, with no evidence of a generalized neuropathic or myopathic process. Extensive blood tests were normal, including acetylcholine receptor antibodies. A detailed gastroenterology evaluation for dysphagia revealed a disorder of esophageal motility. The degree of spinal stenosis at

C5–C6 was moderate on static MRI and without significant cord compression or T2 signal changes in the spinal cord. A slight increase of the C5–C6 retrolisthesis in extension was noted on dynamic flexion-extension X-rays. Therefore, a flexion-extension MRI of the cervical spine was pursued, which indeed revealed significant worsening of the C5–C6 stenosis in extension (figure). Bone density assessment was consistent with osteoporosis.

Questions for consideration:

1. Would you recommend surgery for this patient?
2. If the above answer is yes, would dysphagia and bone frailty influence the surgical approach?

Figure Comparison among cervical spine MRI examinations

June 2000, sagittal T2* MRI with the neck in neutral position (A); August 2018, sagittal T2* MRI with the neck in neutral position (B), flexion (C), and extension (D).

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Section 4

Surgery was recommended to decompress the C5–C6 level and stabilize the segment. An anterior cervical discectomy and fusion would be the best option. However, an anterior approach was not ideal in the patient’s case because of the risk of worsening her dysphagia from manipulation of the esophagus and distal nerves involved with swallowing mechanism. A C4–C6 posterior cervical laminectomy and fusion was performed with the fixation extended to C4 due to her osteoporosis. After a short period of inpatient physical rehabilitation, she experienced marked improvement of spasticity, strength, and paresthesias with improved gait.

Discussion

Patients with CMs of the spine can present with acute changes, progressive deterioration, or a stepwise neurologic worsening, which can mimic demyelinating disease. In the case described, the lack of new hemorrhage or further extension of the hemosiderin stain related to the thoracic spinal cord lesion, along with the new symptoms in the upper extremities, ruled out the thoracic CM as the sole reason for worsening.

Patients who have undergone surgery for an intramedullary lesion, especially a hemorrhagic CM, are at risk of developing tethering of the spinal cord from adhesions related to scarring between the spinal cord surface and the dura/arachnoid mater. While imaging findings of spinal cord tethering are not that uncommon, few patients become symptomatic. Symptoms can develop acutely in the presence of a triggering event, which causes sudden stretching of the spinal cord with resultant damage as the cord is fixed by the tether rather than floating in the CSF. Some patients with tethering can experience a slow but progressive myelopathy because of repeated microtrauma from normal activities. However, symptomatic tethering manifesting more than 30 years after original surgery without inciting event would be unusual. In such cases, however, symptoms start after a relatively short interval following the original surgery as the tether develops and are not delayed in absence of an inciting event.¹

The possibility of a new primary neurologic condition unrelated to the CM and previous surgery was also considered. This diagnostic possibility was raised because of the long interval from the last clinical event, without documented new hemorrhage and new onset of dysphagia, and loss of fine motor skills and atrophy in the hands. However, comprehensive EMG of the upper and lower extremities and laboratory tests did not show any evidence of neuropathy, myopathy, disorder of the neuromuscular junction, or other cause for myelopathy.

A myelopathy secondary to mechanical compression was at first considered unlikely because of the lack of important compression on the static spine imaging.² However, a closer look of the cervical spine flexion/extension dynamic X-ray imaging suggested worsening of the retrolisthesis at C5–C6 with extension. A dynamic MRI was then obtained. When it was compared to a prior MRI (figure, A), the most recent study showed no change of the CM at T3, but interval progression of the retrolisthesis at C5–C6 in neutral position, although not dramatic (figure, B). Flexion MRI demonstrated mild reduction of the retrolisthesis and canal narrowing at C5–C6 (figure, C). Neck extension resulted in significant worsening of both retrolisthesis and spinal canal narrowing at C5–C6 (figure, D). This observation led us to the hypothesis that the patient’s symptomatology could be caused by dynamic compression with repetitive microtraumas of the cord aggravated by the thoracic tethering. This suggested a dynamic compression in a patient with a tethered cord and decreased functional reserve as the main reason for her deterioration. The fact that her cord was fixed at the level of the prior laminectomy from tethering worsened the effects of repeated microtrauma from the dynamic compression at C5–C6. This hypothesis was confirmed by the postoperative functional recovery.

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Disclosure

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Appendix Authors

Name	Location	Role	Contribution
Elisa Colombo, MD	Mayo Clinic, Rochester, MN	Author	Drafted the manuscript for intellectual content
Robert J. Brown Jr, MD	Mayo Clinic, Rochester, MN	Author	Revised the manuscript for intellectual content
Giuseppe Lanzino, MD	Mayo Clinic, Rochester, MN	Author	Revised the manuscript for intellectual content
Benjamin D. Elder, MD, PhD	Mayo Clinic, Rochester, MN	Author	Interpreted data, revised the manuscript for intellectual content

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