RESIDENT & FELLOW SECTION

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Clinical Reasoning: A 45-year-old woman with immobility and incontinence

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SECTION 1

A 45-year-old woman presented with immobility and incontinence in July 2016. Her symptoms started 6 months prior to presentation, when she had multiple falls (without major injuries) at home because of weakness in the lower extremities. She experienced urinary incontinence 1 month later. An indwelling catheter was placed and clamped every 2–3 hours in the daytime and kept open during the night. Later, she preferred not to walk for fear of falling. The patient started to feel stiffness and tightness in her lower extremities 3 months later. The condition worsened gradually to the extent that she could barely move her lower extremities in bed and reported it affected her sleep recently. Along with motor dysfunctions, she also experienced numbness in her lower extremities, but denied pain. She was able to sit in a wheelchair for 7 hours per day. A pressure ulcer was noticed in the sacral region 4 months later. She denied other discomfort. Her appetite has been good. She had no significant weight changes during the last half year.

Medical history included massive subarachnoid hemorrhage extending into ventricles in 2012 (figure 1). Per medical records and the family, repeat lumbar punctures were performed on a daily basis for nearly 1 month to drain the blood until clear. The patient regained consciousness after 21 days and had left-sided paralysis. With therapies, she had no residual functional deficits in her upper extremity, was able to walk slowly, and carried out some activities of daily living independently when discharged. She was found to have moyamoya disease, which was successfully treated with surgery later in 2012. Follow-ups in 2 years revealed good anastomosis. She was referred to our department for rehabilitation in 2015 due to unsteadiness in walking, which was believed to be caused by her previous brain lesion. Following intensive therapies, she was capable of walking independently with a walker. She had no sensation disturbance and no bowel or bladder malfunction at that time. The patient denied infections of any kind in the CNS, spinal anesthesia, myelography, blood patch, spine surgery, or neurosarcoidosis in her life.

Questions for consideration:

1. Where is the lesion?
2. What physical examinations and assessments are needed?

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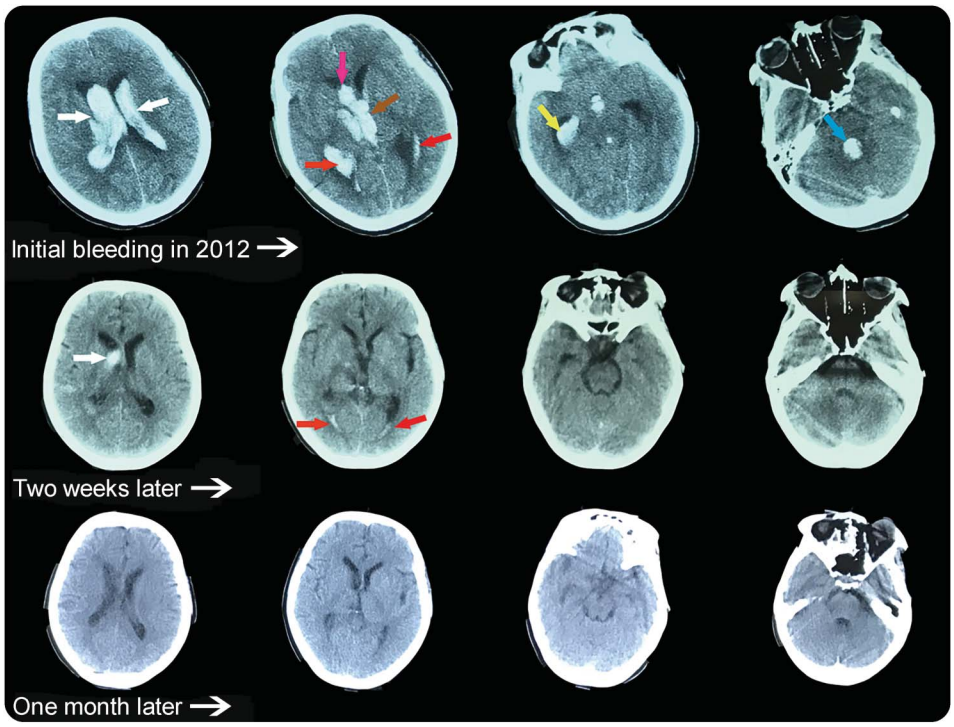
Figure 1 Massive subarachnoid hemorrhage extended into bilateral ventricles (white arrows), occipital/posterior horns (red arrows), right frontal/anterior horn (pink arrow), right temporal/inferior horns (yellow arrow), and the third (brown arrow) and the fourth (blue arrow) ventricles with ventricular expansion in 2012



The blood in the subarachnoid space resolved gradually within 1 month. Repeat lumbar punctures were performed to drain the blood on a daily basis during this period.

SECTION 2

According to the patient's manifestations of paraplegia rather than hemiplegia, it was presumed more likely that the lesion was lying in the spinal cord than the brain. Therefore, initial assessments, including the American Spinal Injury Association (ASIA) classification to evaluate motor and sensory functions of the lower extremities as well as the highest affected level on the trunk, the modified Ashworth scale (MAS) for the severity of spasticity, tendon reflexes, and pathologic signs, should be performed. These are feasible physical examinations and assessments that could be done during the first-time patient encounter at bedside. More sophisticated assessments could be performed later.

On admission, the patient had muscle atrophy in her bilateral lower extremities. She could only voluntarily move her right lower extremity slightly in bed. The muscle tones were rated MAS grade 2–3, indicating severe spasticity. Reflexes were brisk with clonus. Babinski signs were positive bilaterally. The fine and crude sensation were decreased below T6 dermatome bilaterally. Digital rectal examination revealed decreased sensation in the anal region and involuntary contractions of anal sphincters. The pressure ulcer in the sacral region was 4 × 5 cm, down to the deep fascia. Her speech, swallowing, and communication, as well as functions concerning upper extremities, were intact. She was unable to turn over in bed or sit or stand independently. Her life was highly dependent.

Questions for consideration:

1. What are the differential diagnoses for the patient's presentation?
2. What other investigations are needed to help with the diagnosis?

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SECTION 3

Coexistence of spasticity and weakness, brisk reflexes, and positive pathologic signs indicated upper motor neuron lesions. Muscle atrophy may be attributed to disuse. Sensation abnormalities made diseases that purely affect motor pathway, like amyotrophic lateral sclerosis, less likely. Posterior roots involvement would produce a segmental somatosensory deficit accompanied by ataxia and asynergia, while posterior horn involvement mainly affects pain and temperature sensation.[1] However, neither causes spasticity and weakness. The syndrome of combined lesions of the posterior columns and corticospinal tracts, also known as subacute combined degeneration, which is commonly caused by vitamin B_{12} deficiency, may present with spastic paraparesis and unsteadiness in walking, and requires further investigation.[1] Progressive spinal cord transection syndrome caused by tumor or syringomyelia would also produce similar presentations, like spastic paraplegia, sensory deficits, and bowel and bladder dysfunction,[1] which the patient had. CT and MRI were the best way to investigate the lesions in her spine and help with the differential diagnosis. However, intramedullary spinal cord tumor would still be difficult to differentiate conclusively without biopsy.[2]

After admission, a series of imaging studies was performed. Spine MRI with contrast showed syringomyelia from T5 to T12, myelatrophy and degeneration in C7–T4, and tethered spinal cord without enhancement (figure 2). There was mild disc herniation in C3–C7 and small hemangiomas in L1 and S2 vertebra. Excessive fat tissue accumulated in the epidural area compressing thecal sac at L3–L5 level. The thoracic CT showed a dot-like calcification in T6 level. Because of the patient's moyamoya history, magnetic resonance angiography of the spine was performed and showed expansion in the perivertebral veins as well as in the central canal of the spinal cord. Head MRI was consistent with previous lesions. Two attempts at lumbar puncture performed by skilled physicians were unsuccessful to obtain any CSF.

Question for consideration:

1. What is the most likely diagnosis?

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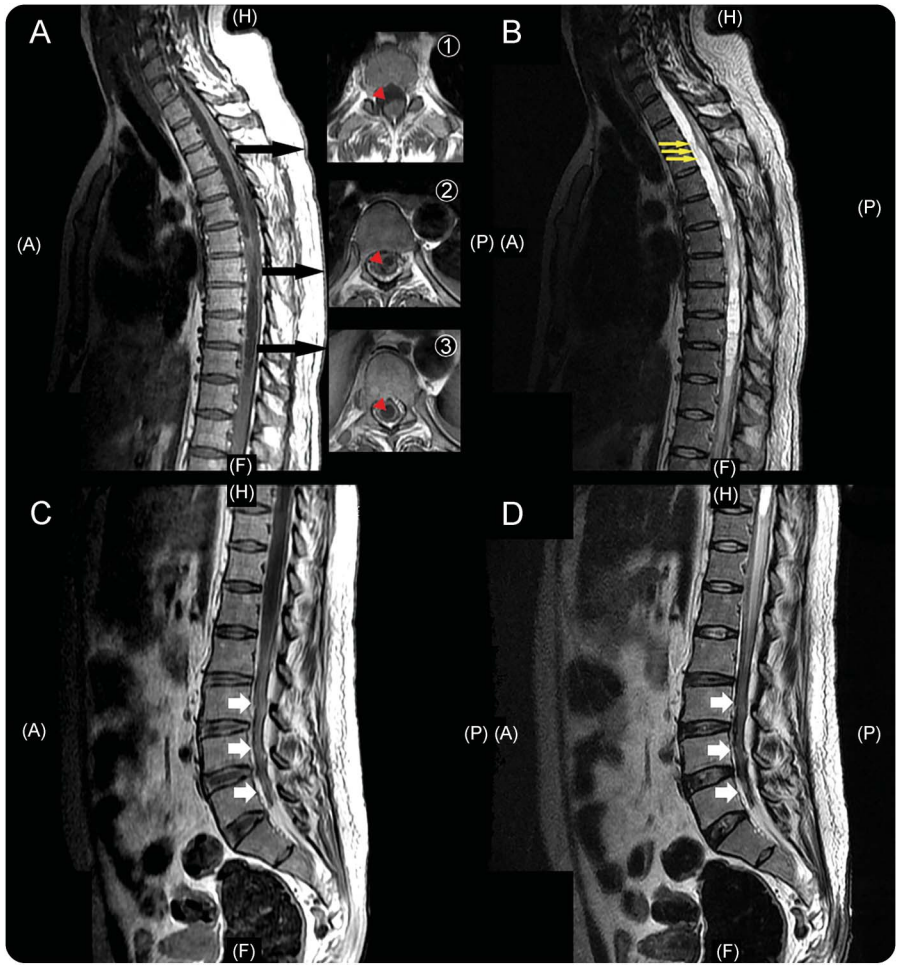
Figure 2 MRI of the spinal cord



The top left (A) and the bottom left (C) are T1-weighted images, while the top right (B) and the bottom right (D) are T2-weighted images. The red triangles in the top left (A) image show the thoracic spinal cord was tethered dorsally (①), distorted with septa (②) and contained a massive syrinx (③). The yellow arrows in the top right (B) image indicate several thread-like low signals, suggesting formation of fibrinoid materials inside the subarachnoid space. The white arrows in the bottom left (C) and right (D) images show the cauda nerves were tethered, clumped, and thickened.

SECTION 4

The diagnosis of adhesive arachnoiditis was rendered based on the patient's signs and symptoms plus imaging manifestations. Massive subarachnoid hemorrhage or occult meningitis resulting from repeat lumbar punctures may be the underlying etiologies of the disease in this case.

After administering baclofen for spasticity, gabapentin for paresthesia, and neuroprotective agents, combined with intensive physiotherapies for 3 months, the patient's spasticity was well-controlled with MAS grade 1 on the right side and grade 1+ on the left side. She was able to lift her right lower extremity above the bed and turn over in bed independently. She was rated as ASIA grade D. Repeat imaging studies did not reveal substantial growth of the lesions in the spinal cord, suggesting little likelihood of tumor. Neurosurgery consult suggested no indication for surgical decompression with current improvements, as surgery itself might aggravate the inflammation process.[3]

DISCUSSION Adhesive arachnoiditis refers to a severe type of inflammation in the arachnoid mater. There have been several case reports regarding adhesive arachnoiditis occurring following subarachnoid hemorrhage.[3,4] Post–subarachnoid hemorrhage arachnoiditis was mostly seen in ruptured aneurysm in the posterior circulation.[3] However, this is one case identified later with moyamoya disease rather than aneurysm. Despite the origin of the blood, it is suggested that the occurrence of spinal subarachnoiditis was related to the volume of blood in the spinal subarachnoid space.[4] Iatrogenic causes, such as repeat epidural blood patch,[5] spinal surgery,[6] and lumbar puncture,[7–9] were also seen in case reports. Interestingly, a 2-session epidural blood patch with only 25–30 mL autologous blood injected into the epidural space caused adhesive arachnoiditis. Regardless of the origin and the volume, it is postulated that factors including hemoglobin from lysed erythrocytes, activated platelets, complements, leukotrienes, endothelins, histamine, bradykinin, proinflammatory cytokines, and free radicals contribute to the inflammatory response at the local level.[3] Therefore, in this case, the blood and the interstitial fluid enriching inflammatory factors leaked into the subarachnoid space, perhaps resulting in the subsequent pathologic changes. The molecular mechanism is unknown. Some cases have developed years after the initial incidence, like this one, mandating meticulous investigations of the patient's history and thorough examination.

The noninfectious inflammation in the arachnoid mater induces irritation to the nerve roots and damages the structure of the spinal cord at different levels. Presentations are highly variable in severity. Pain is the most common symptom and can severely impair a patient's quality of life.[8] Numbness/tingling, muscle cramps, weakness, stiffness, balance difficulties, and loss of mobility were seen in 68%–86% of patients.[8] Other systemic involvements including bladder/bowel dysfunction, increased sweating, and depression were seen in 58%–68% of patients.[8] In most cases, distorted spinal cord with septa, interstitial edema, syrinx formation indicating CSF flow disturbance, syringomyelia, clumping of the nerve roots, and tethered spinal cord have been reported in MRI, as seen in this case.[3,4] Candle dripping appearance can be observed in the myelogram.[7] Calcification, which was unusual in arachnoiditis and reported in one case with iophendylate-induced arachnoiditis, was also discovered in this case.[2]

Currently, no cure has been developed for the disease; surgery may be helpful for decompression and reestablishing CSF flow. From a physiatrist's point of view, signs and symptoms should be well-managed to maximize patients' functional abilities and improve their quality of life.

AUTHOR CONTRIBUTIONS

Dr. Bei Zhang contributed to drafting/revising the manuscript, analysis/interpretation of data, and figure preparation. Dr. Gang Liu contributed to drafting/revising the manuscript and analysis/interpretation of data. Dr. Ying Chen contributed to analysis/interpretation of data and figure preparation. Dr. Shan Tian contributed to analysis/interpretation of data and figure preparation. Dr. Kweku Laast contributed to analysis/interpretation of data and revising the manuscript. Dr. Yulong Bai contributed to the study concept and analysis/interpretation of data. Dr. Yi Wu contributed to the study concept and analysis/interpretation of data.

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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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