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Differentiating Etiologies of Pain in the Presence of Dorsal Arachnoid Web: A Case Report

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Background: Dorsal arachnoid web (DAW) is a rare spinal pathology involving a thickened band of arachnoid tissue that compresses the thoracic spinal cord, potentially causing myelopathy. Patients with DAW who present with pain or neurological complaints should be evaluated thoroughly to determine the most likely cause of symptoms, particularly in the context of clinical presentations that overlap with other neurological or pain syndromes.

Case Report: A 76-year-old man with a history of inclusion body myositis and prior middle cerebral artery stroke presented with left-sided pain and weakness. Although DAW was noted on imaging, further evaluation indicated that the patient's pain symptoms were due to central pain syndrome secondary to his previous stroke, given the correlation between symptom localization and stroke territory. He was managed successfully with pregabalin and amitriptyline without the need for surgical intervention.

Conclusion: In patients with overlapping neurological and pain syndromes, accurate diagnosis is essential to determining the best choice of treatment.

Key words: Chronic pain, dorsal arachnoid web, diagnostic imaging, central pain syndrome, case report

BACKGROUND

Dorsal arachnoid web (DAW) is a rare spinal pathology characterized by a thickened band of arachnoid tissue compressing the dorsal spinal cord, most often in the thoracic region, causing cord compression and myelopathy. Symptoms vary and may include sensory changes, motor changes, back pain, and hypertonia. The web may obstruct the flow of cerebrospinal fluid, causing syringomyelia at levels adjacent to, above, or below the web. The exact cause is not fully understood, but several factors have been proposed to contribute to the development of this disease, such as congenital abnormalities, trauma, inflammatory conditions, and idiopathic changes. DAW is found more frequently in women (1). Diagnosis is confirmed by the "scalpel sign" on magnetic resonance imaging (MRI) or computed tomography (CT) myelogram, described by Reardon et al (1) as a focal dorsal indentation and anterior displacement of the cord, creating the shape of a scalpel blade. While surgery is often recommended, many patients do not require surgical intervention. In patients presenting primarily with pain, coexisting conditions can complicate the diagnostic process and increase the risk of misdiagnosis. Misidentifying the source of pain can result in ineffective treatments, including unnecessary surgeries. Given the overlapping presentations of various chronic pain conditions, it is essential for providers

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to accurately determine the origin of symptoms to effectively guide treatment. We present the case of an elderly man with pain and neurological deficits who had multiple potential contributors to his symptoms, including a new finding of DAW on imaging. Verbal informed consent was obtained for anonymized patient information to be published in this article. Our institution does not require approval from a board of ethics for reporting individual cases or case series.

CASE REPORT

History

A 76-year-old man who had a history of inclusion body myositis and left hemiparesis secondary to prior stroke presented to the clinic with left facial and hand pain, accompanied by weakness in the left arm and leg. He reported that the pain was sharp, became worse with movement, and fluctuated between a 2/10 and 5/10.

Physical Exam

Physical exam showed that the left upper and lower extremities had a reduced sensation to light touch and muscle strength graded at 4/5. Bilaterally, both the upper and lower extremities, showed absent reflexes and negative Babinski and Hoffman signs.

Imaging

Thoracic CT myelogram revealed dorsal cord flattening centered at T2-3, exhibiting a scalpel sign typical of a dorsal arachnoid web, without discrete cord signal abnormality (1) (Fig. 1).

Diagnosis and Treatment

The patient was referred to neurosurgery for further evaluation, during which it was determined that the weakness in the left lower extremity was most likely related to inclusion body myositis, since there was no clear evidence of myelopathy such as increased lower extremity reflexes or a positive Babinski sign. Therefore, surgical intervention was not recommended. The pain in the patient's face and upper extremity were attributed to central pain syndrome secondary to his prior middle cerebral artery stroke, as the distribution of pain symptoms corresponded with the stroke's anatomical location. He was treated with pregabalin and amitriptyline. At the one-year follow-up, the patient's pain had improved with this medication regimen, and there were no significant changes in sensation or muscle strength in the upper and lower extremities.

DISCUSSION

DAW is a rare condition of unknown etiology and may present with many different symptoms, making early diagnosis challenging. Given the great variety of DAW symptoms as well as the nonspecific nature of many of these symptoms, timely use of advanced imaging, like MRI and CT myelography, is critical for early diagnosis and treatment. The presence of the "scalpel sign" on MRI or CT myelogram confirms the diagnosis of DAW. Reardon et al identified the scalpel sign as a focal dorsal indentation and anterior displacement of the spinal cord, forming the silhouette of a scalpel blade (1). Other findings may include a syrinx, which was found by Nisson et al in 67% of patients with AW (2).

Assessment of symptoms and prompt surgical consultation are necessary to determine the next steps after confirming the presence of the scalpel sign. Appropriate treatment of DAW ranges from medical management to surgical decompression, depending on the severity of symptoms. Back pain and sensory changes are some of the most common initial presentations, but many other symptoms have been reported, including hyperreflexia, gait instability, and urinary incontinence (6)(7). Nisson found that weakness was the most common complaint, reported by 67% of patients, and sensory loss was the second most common, reported by 65% of patients (2). Nisson also found that 35% of patients reported progressive symptoms (2). Ben Ali reported a case of a patient who presented for interscapular pain without neurological deficits and was found to have an MRI that aroused clinical suspicions of DAW, but because his symptoms were minor, a conservative approach was pursued (9). When surgery is required, the most commonly used surgical treatment for DAW is laminectomy, with intradural excision of the anterior web (2). Carr et al found that DAW patients most often underwent surgery for thoracic myelopathy with gait abnormalities and lower extremity paresthesias, but more than half of the patients did not require surgical management at all (10). This range of clinical presentations and management strategies underscores the importance of accurate pain source identification to ensure appropriate, individualized treatment.

Assessing the severity of symptoms caused by DAW can be complicated by the coexistence of other sources of chronic pain, such as in our patient, who had a history of inclusion body myositis and prior stroke. Per some reports, chronic pain conditions are misdiagnosed

40-80% of the time, which may lead to inappropriate or unnecessary interventions (11). Finding the correct diagnosis is challenging, especially because many common chronic pain conditions have overlapping symptoms (12). Knowledge gaps in providers further hinder the appropriate management of pain syndromes (13). Despite these challenges, it is important for clinicians to continue aspiring to better pain management, since undertreated pain can lead to physiological, psychosocial, and economic dysfunction (14). Along with avoiding the undertreatment of pain, we emphasize the importance of avoiding the overtreatment thereof through unnecessary therapies that may not benefit the patient or may have risks which outweigh the potential benefits. Overtreatment can account for up to 30% of health care costs and occurs frequently in chronic pain patients (14,15). Conservative management is often preferred initially over procedural interventions, such as in patients with common conditions, including chronic low back pain, central pain syndrome, and fibromyalgia (15-17). In patients with chronic pain of unknown etiology, providers should minimize opioid use and instead prescribe a stepwise approach appropriate to the individual presentation (e.g., using gabapentin in cases with a neuropathic component [Salduker]). The many considerations of chronic pain management highlight the necessity of precise pain source identification to avoid both under- and overtreatment and thus ensure that patients receive effective care.

In our patient, it was necessary to tease out the cause of pain to determine appropriate treatment and avoid unnecessarily invasive measures. Given the patient's lack of upper motor neuron signs in his lower body, the absence of cord signal abnormality on his MRI scan, and his complaints of pain at sensory levels above the location of the arachnoid web, his arachnoid web did not appear to be causing myelopathy or contributing to his left face and hand pain at the time of evaluation. Because his facial and hand pain aligned with the territory affected by his prior middle cerebral artery stroke, central pain syndrome was highest on the differential and was managed successfully with medical therapy alone. This case underscores the importance of identifying the primary pain generator accurately to guide treatment decisions and prevent unnecessary invasive measures.

CONCLUSION

Central pain syndrome occurs due to a heightened state of activity in the central nervous system, leading to paresthesias, allodynia, hyperalgesia, and other symptoms of pain, even without input from the peripheral nervous system. Post-stroke central pain syndrome generally occurs 3-6 months after a stroke and is associated with psychiatric and functional challenges (18,19). Knowledge of the precipitating factors is limited, but some risk factors include spasticity, reduced upper extremity movements, and sensory deficits (16). After clinicians use a thorough history and physical examination to exclude other causes of pain, central pain syndrome is diagnosed. Central pain syndrome is often treated with antidepressants and anticonvulsants, as it often does not respond to more traditional analgesics like NSAIDs. Using medications such as amitriptyline is not only clinically effective for neuropathic pain conditions like central pain syndrome, the practice is also cost-effective (20). The symptoms of central pain syndrome

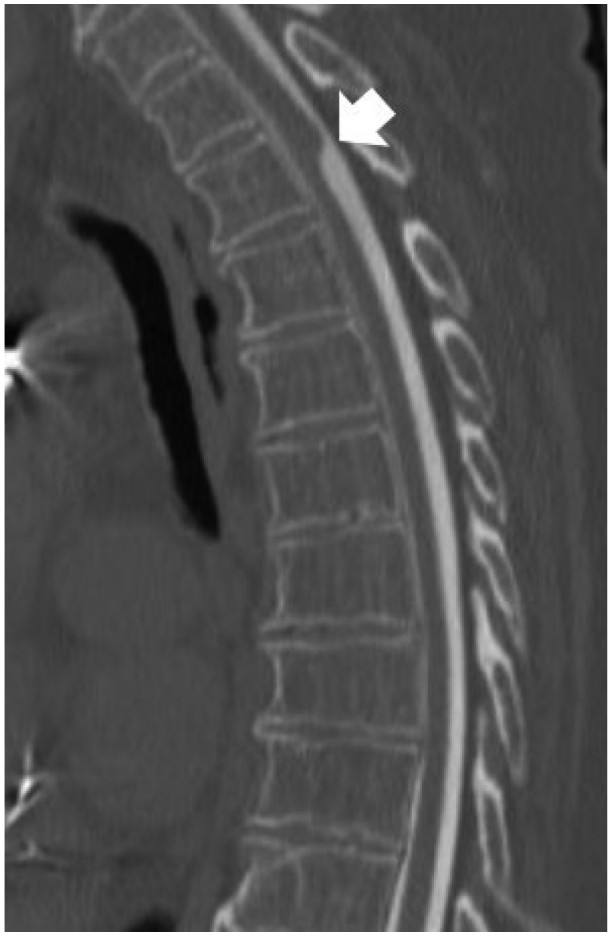

Fig. 1. "Scalpel sign" on CT myelogram.

overlap with those of other pain syndromes, and the condition can be potentially be managed medically at low cost, highlighting the need for careful diagnostic evaluation to correctly identify the pathology and tailor treatment accordingly.

Imaging in our patient confirmed the presence of DAW, and a timely referral to neurosurgery helped rule out the need for surgical treatment. After other potential causes of pain were excluded, the patient was diagnosed with central pain syndrome, which was managed effectively with conservative measures. This approach avoided the risk of procedural complications and led to a cost-effective recovery. Our case highlights the importance of an accurate diagnosis to guide treatment, as etiologies of chronic pain may share overlapping symptoms.

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