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POSTDURAL PUNCTURE HEADACHE IN A PATIENT WITH THORACIC ARACHNOID WEB WITH KNOWN JOINT HYPERMOBILITY: CASE REPORT

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Background: Patients with connective tissue disorders (CTDs), such as Ehlers-Danlos syndrome, can present unique challenges in the management of spinal pathology and procedural complications due to underlying tissue fragility and susceptibility to dural ruptures. Thus, there is a need for less invasive diagnostic and therapeutic care in this population.

Case Report: We present a case of a 48-year-old woman with joint hypermobility who developed acute-on-chronic back pain and radicular symptoms. Imaging revealed an L5-S1 radiculopathy and a thoracic arachnoid cyst. During inpatient rehabilitation, she experienced a postdural puncture headache (PDPH) following a thoracic spine myelogram refractory to conservative measures and the gold standard, epidural blood patches (EBPs), necessitating the need for management strategies tailored to her complex clinical profile.

Discussion: Managing PDPH in patients with CTDs requires careful consideration of alternative therapeutic options and potential complications associated with standard treatments like EBPs.

Conclusions: This case highlights the need for individualized approaches to minimize procedural risks while optimizing patient outcomes in this population.

Key words: Joint hypermobility syndrome, Ehlers-Danlos syndrome, thoracic arachnoid cyst, postdural puncture headache, epidural blood patch

BACKGROUND

This case report presents a rare and complex scenario involving a 48-year-old female patient with a history of joint hypermobility syndrome. Initially presenting with acute-on-chronic low back pain accompanied by new right lower extremity weakness and radicular symptoms, she also reported band-like midthoracic pain. Diagnostic investigations revealed an L5-S1 radiculopathy and the unexpected finding of a thoracic arachnoid cyst or web. Following an acute care hospitalization, the patient was referred to inpatient rehabilitation (IPR) for management of her functional deficits. During her rehabilitation, while undergoing a diagnostic thoracic spine myelogram (TSG), she experienced a significant complication in the form of a postdural puncture headache (PDPH), a positional bilateral headache associated with nausea and dizziness worse in the upright compared with supine position caused by cerebrospinal fluid (CSF) leakage through the dural puncture.

Evidence and Considerations Related to Connective Tissue Disorders and PDPH

Recent literature (1,2) has shown associations between patients with connective tissue disorders (CTDs)

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and higher rates of spinal procedural complications due to the presence of underlying fragile meningeal diverticula (abnormal outpouchings of the spinal arachnoid, nerve root sheath, or dural sac), and dural rents. The anatomy of the epidural space and its surrounding structures plays a critical role in understanding the mechanisms behind PDPH. To access the epidural space, a needle must traverse several anatomical layers, including the skin, subcutaneous tissue, ligaments, and finally, the ligamentum flavum—a bilaterally segmented structure that may not always merge at the midline, increasing the risk of dural puncture. The dura mater, a robust connective tissue layer, can be punctured or damaged during procedures involving spinal anesthesia or epidural interventions, leading to CSF leaks (3).

Generalized CTDs are thought to play a significant role in spontaneous spinal CSF leaks, with some studies (4) reporting characteristics of such disorders present in about two-thirds of affected patients. Prior biomolecular research has shown that these disorders are heterogeneous, potentially impacting components of the dural extracellular matrix. The presence of joint hypermobility in patients has been known to complicate surgical wound closure through attenuation of the dorsal muscular fascia further predisposing patients to CSF leaks postsurgically (4). It has been estimated that approximately one-fifth of patients exhibit subtle skeletal features of Marfan syndrome without genetic mutations in fibrillin-1, suggesting microfibril defects in the extracellular matrix. Less frequently, well-defined CTDs like Marfan syndrome, Ehlers-Danlos syndrome (EDS) type II, and autosomal dominant polycystic kidney disease are implicated (4).

This case underscores the critical considerations in managing epidural procedure complications in patients with CTDs, highlighting the challenges and potential pitfalls in their care.

METHODS

This case report involves a retrospective analysis of clinical data obtained from a patient admitted to the IPR unit at the University of Pittsburgh. As the case report is devoid of patient-identifiable information, it is exempt from institutional review board review requirements as per University of Pittsburgh policy. The report complies with all relevant patient privacy regulations, and consent was obtained from the patient to use de-identified information for educational purposes. Diagnostic imaging, including thoracic and lumbar spine magnetic resonance imaging (MRI), as well as electromyography (EMG) findings, were utilized to assess the extent and nature of the patient's spinal pathology. The occurrence and management of the PDPH following a TSG were documented, including subsequent interventions such as a sphenopalatine ganglion block and an epidural blood patch (EBP).

CASE PRESENTATION

The patient's clinical course was initially marked by a 2-month history of worsening radicular pain precipitated by a nontraumatic incident while sitting on the floor. Thoracic spine MRI revealed a nonenhancing T2 hyperintensity suggestive of a dorsal CSF space abnormality spanning T4-T9, indicative of an arachnoid cyst or web (Fig. 1). Concurrent lumbar spine imaging demonstrated mild bilateral foraminal encroachment without significant stenosis. EMG findings confirmed the presence of L5-S1 radiculopathy.

During a diagnostic TSG at IPR to further evaluate the patient's arachnoid web, the patient experienced a PDPH, a known complication of epidural procedures. Initial conservative measures, including bed rest, hydration, and caffeine intake, were attempted for 24 hours and provided only transient relief. A multimodal approach with medications, including scheduled Tylenol and gabapentin, was also attempted in addition to preexisting as-needed opiates for another 24 hours after the patient failed conservative measures.

Since the patient failed these measures, subsequently a sphenopalatine ganglion block via a transnasal approach was attempted, offering mild symptomatic improvement for a brief period and reduced Numeric Rating Scale scores from an 8 down to a 4 for nearly 48-72 hours postblock (Fig. 2). However, due to persistent symptoms and escalating discomfort, an EBP was performed 9 days postprocedure. While the EBP provided mild relief, its overall tolerability was poor, and the patient did not experience relief within the expected one-week time frame after EBP administration, noting back pain, neck pain, radicular symptoms, and worsening headache after administration. This presentation raised concerns about potential complications, such as infection, arachnoiditis in a patient with preexisting concern for arachnoid pathology, repeat dural puncture after EBP, or worsening radiculopathy.

DISCUSSION

The management of PDPH in patients with CTDs presents unique challenges due to underlying tissue

fragility and potential complications associated with neuraxial interventions. CTDs, such as EDS, are known to predispose individuals to spontaneous dural ruptures and subsequent headaches (1) due to the formation of fragile meningeal diverticula or simple dural rents (2), which may exacerbate with invasive procedures, including myelography or epidural placement. Moreover, the presence of spinal arachnoid cysts or webs further complicates the scenario, as these conditions have been sporadically linked to CTDs and can contribute to diagnostic and therapeutic dilemmas (5).

EBPs, considered standard therapy for PDPH, are generally well-tolerated in the general population (3) but we theorize that these procedures can pose heightened risks in patients with CTDs. Concerns include the potential for infection, exacerbation of meningeal irritation, or complications related to inadvertent dural penetration (6). Current literature lacks sufficient evidence regarding the optimal management strategies for PDPH specifically tailored to patients with CTDs, highlighting the need for further research in this area.

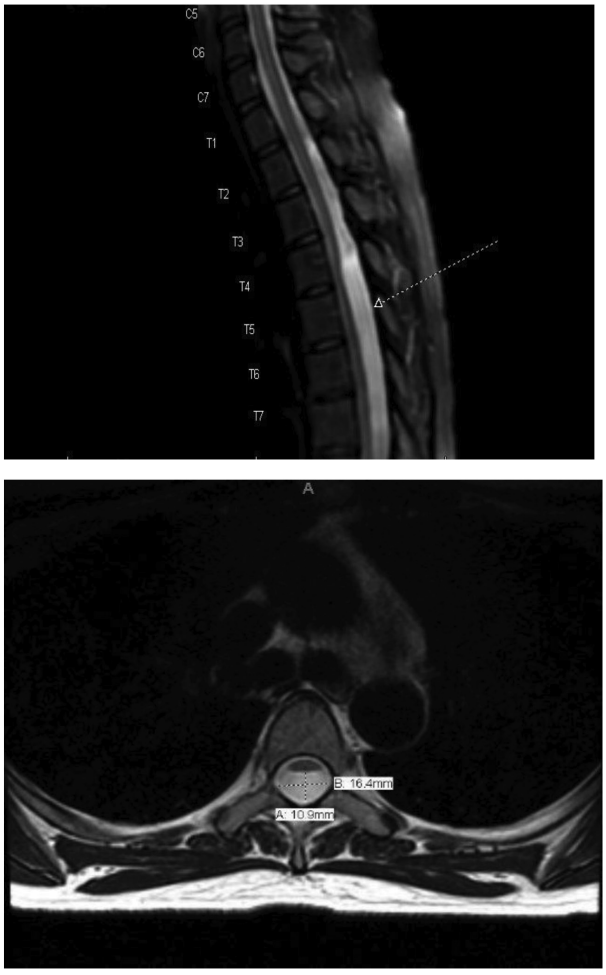

Fig. 1. MRI of the thoracic spine depicting a T2 hyperintensity posterior to the cord around the arachnoid space between T4-T9 representing a likely arachnoid web with some associated cord flattening (sagittal view on the top, axial view at the bottom). MRI, magnetic resonance imaging.

European anesthesia guidelines suggest caution in performing neuraxial procedures in patients with EDS, recommending alternative approaches when feasible (1). A recent study (7) did show an increased incidence of PDPH in patients with joint hypermobility with Beighton scores above 4. The current school of thought is that in cases of CTDs and hypermobile joint disorders, weakness in the dural tissues and issues with the elasticity of structures around the region contribute to the development of intracranial hypotension leading to an increased emergency of a postspinal headache (8,9). Additionally, there is some evidence that there is an association between increased duration of hospital stays and a heightened need for prolonged medical treatment for patients who have joint hypermobility with PDPH (7).

Furthermore, while current data on the prevalence of PDPH in CTDs is limited, an association has been noted based on several prior small, prospective stud-

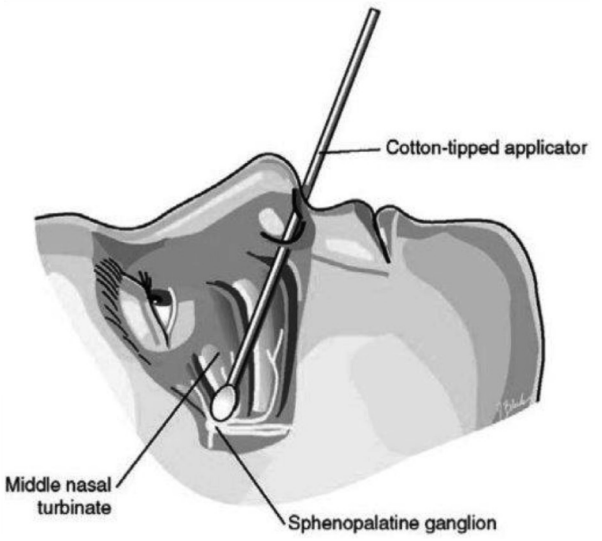

Fig. 2. Depicting a sphenopalatine ganglion block via the transnasal approach. Derived from: sphenopalatine ganglion block for PDPH after invasive CSF pressure monitoring. Case report by Felipe Chiodini Machado et al and is licensed under CC BY 4.0 (https://creativecommons.org/licenses/by/4.0/). PDPH, postdural puncture headache; CSF, cerebrospinal fluid.

ies. In a recent prospective study by Sonawane et al (10), involving 50 patients who underwent neuraxial blockade, most patients had common features suggestive of some type of underlying CTD, including ligamentous laxity (96%), high-arched palate (96%), the blue sclera (45%), joint hyperextensibility (82%), and ejection clicks (64%).

Another prospective study by Schievink et al (2) in 2004 displayed CTD features in 38% of patients analyzed with diagnosed spontaneous intracranial hypotension, including features of Marfan syndrome, EDS, and joint hypermobility. In addition to the recent study by Yilmaz et al (8) that showed an association between higher Beighton scores and PDPH, there have been several case studies and small, prospective studies (11-15) displaying connections between patients with PDPH and CTDs, such as Marfan syndrome and Sjogren's syndrome.

Minimally invasive interventions, such as greater occipital nerve blocks and sphenopalatine ganglion blocks, may offer viable alternatives for managing refractory PDPH in this patient population, potentially mitigating risks associated with more invasive procedures like EBPs. It should be noted, however, that EBPs are generally well-tolerated procedures with few side effects (16). A recent, comprehensive review (3) on EBPs summarized adverse outcomes associated with EBPs. Rare complications include repeat dural puncture (< 1% of cases), epidural hematoma, pneumothorax, cerebral venous sinus thrombosis, and seizures. Commonly reported issues include transient neck and back pain (can be seen in up to 80% of cases), bradycardia, radicular pain, and exacerbation of PDPH, seen in > 1% of cases. Serious but infrequent complications include arachnoiditis, cauda equina syndrome, organ dysfunction, infection, and meningitis, which have been described in case studies in the past (3).

Worsening arachnoiditis linked to EBPs has been reported in some case studies over the past decade (16). Given our patient's medical history, the relationship between arachnoid cysts, webs, and arachnoiditis with EBPs remains poorly understood, highlighting the need for further research. Additionally, there is speculation that EBPs may exacerbate PDPH by creating additional dural rents, a risk particularly relevant in patients with CTDs (16). This could lead to serious complications, such as intrathecal or subdural hematomas, and we theorize that this may necessitate closer diagnostic monitoring in patients with CTDs. While no definitive connection has been established between EBP complications and CTDs, comparing failure rates in patients with and without CTDs and investigating structural factors contributing to failures could provide valuable insights for improving clinical practice.

Additionally, utilizing a multimodal approach involving medications, including nonsteroidal anti-inflammatory drugs and acetaminophen, may help reduce the need for EBP consideration. Other medications that have growing evidence to show efficacy in the management of PDPH, in particular, based on more recent studies (17) include gabapentin, theophylline, and hydrocortisone. These medications should also be explored as standard of practice for this patient population as long as there are no medical contraindications. Moving forward, this case emphasizes the importance of individualized treatment approaches that prioritize patient safety and symptom relief while minimizing procedural risks in the context of CTDs.

CONCLUSIONS

This case report underscores the intricate balance required in managing spinal pathology and procedural complications in patients with underlying CTDs. By highlighting the challenges encountered and the lessons learned from this case, we contribute to the growing body of knowledge aimed at optimizing the care of individuals with joint hypermobility, a complex musculoskeletal condition that can have neurological and functional implications.

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