Supine Digital Subtraction Myelography for the Demonstration of a Dorsal Cerebrospinal Fluid Leak in a Patient with Spontaneous Intracranial Hypotension: A Technical Note

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ABSTRACT

A patient with spontaneous intracranial hypotension due to a spinal cerebrospinal fluid (CSF) leak required localization of the leakage site prior to surgical management. Conventional, computed tomography and prone digital subtraction myelography failed to localize the dural tear, which was postulated to be dorsally located. We present here a digital subtraction myelographic approach to accurately localize a dorsal site of CSF leakage by injecting iodinated contrast via a lumbar drain with the patient in the supine position.

CASE REPORT

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A 34 year-old male presented to the emergency department at our institution approximately one week following the sudden onset of headache and acute neck pain. This was accompanied by nausea, vomiting, photophobia, diplopia and bilateral tinnitis. The pain was described as constant and rated as 5/10 (verbal) at the time of presentation and 8/10 (verbal) at its worst. Symptoms were exacerbated by sitting and palliated by both walking and approximately 30 minutes of supine positioning. Coughing and sneezing were also identified as aggravating factors. The temporal course was one of less pain upon arising followed by progressive worsening throughout the day. There was no associated history of loss of conscious, facial numbness or tingling, fever or sweating. The patient denied any history of rhinorrhea or otorrhea. No prior back surgery, trauma, lumbar puncture or similar headaches were reported.

Initial clinical exam revealed a normal, pain free cervical range of active motion with an unremarkable neurological examination. An atraumatic lumbar puncture performed in the Emergency Department revealed mildly elevated protein and normal glucose and red and white blood cell counts. A gram stain and culture documented sterile CSF. Unfortunately, opening CSF pressure was not recorded.

Initial CT head demonstrated no abnormal findings. MR imaging of the head and cervical spine revealed diffuse thickening and enhancement of the pachymeninges with flattening of the anterior pons and low lying cerebellar tonsils (Figure 1). The venous hinge angle was reduced to 70 degrees (Figure 2). A nuclear medicine CSF circulation study revealed no evidence of CSF leakage (Figure 3).

A spinal MR demonstrated normal cord signal with an incidental vertebral hemangioma at T5. Extensive abnormal low T1 and high T2 epidural collections were present within the spinal canal, ventrally from C2 to T12 and dorsally from C7 to T8 and T9 to L2 (Figure 4). Neither a dural tear nor a pseudomeningocele were evident.

Conventional myelography revealed a definite large volume CSF leak based on significant extradural contrast pooling, which was observed once the patient was transferred from a prone to right lateral decubitus postion (Figure 5). A dorsal site of leakage was favored to lie in the vicinity of T12 to L2. Extensive extradural contrast was observed on CT myelography, after the patient had been supine for a short period (Figure 6).

A diagnosis of headache secondary to intracranial hypotension was made and the patient was initially treated with five 20 cc epidural blood patches at vertebral levels L3-4 and L4-5 each resulting in weeks to months of symptomatic improvement. Due to the recurrent nature of the headaches, definitive surgical management was planned and a new approach was devised to accurately document the leakage site prior to surgery.

The patient was taken to the neuroangiography suite on the day of surgery and a 14 gauge, 9 cm Tuohy needle was introduced into the thecal sac at the level of L2-3 under fluoroscopic guidance. An 80 cm lumbar catheter (Medtronic Inc., Minneapolis MN) was advanced to the level of L1 and secured to the patient's skin. The patient was placed supine and biplanar digital subtraction myelography was performed during the real-time injection of 8 cc of Omnipaque 300 over approximately 1 second. Two dorsal sites of leakage were confidently visualized at T9-10 and T11-12. (Figure 7 & 8)

Operative repair was undertaken. Approximately 10 cc of blood-tinged CSF was aspirated from the lumbar drain, mixed with 0.2 cc of sterile fluorescein and then re-injected in to the thecal sac. The lumbar drain was then closed and the spinal canal opened which confirmed the intrathecal placement of the drain. CSF leaks were observed as fluorescein-tinged CSF was exuding through thickened arachnoid deep to a large dural defect extending from T9 to T11. The dural defect was closed with a split fascia graft.

At one month follow-up, the patient had an 80% improvement in headache, with complete resolution of tinnitus. At one year follow-up, the patient's symptoms remained stable.

DISCUSSION

Spontaneous intracranial hypotension (SIH) was initially described in 1938 by Schaltenbrand as a syndrome characterized by orthostatic headache [1]. More recently, diagnostic criteria have been proposed by the International Headache Society (IHS) and by Shievink for headache attributable to spontaneous (or idiopathic) low CSF pressure [2,4]. To fulfill these criteria, the headache must worsen within 15 minutes after sitting or standing and be associated with at least one symptom of neck stiffness, tinnitus, hypacusia, photophobia or nausea. Evidence of low CSF pressure on MRI, imaging evidence of CSF leakage or CSF opening pressure below 60 mm H_2O are also required criteria. To be categorized as spontaneous or idiopathic, there can be no history of dural puncture or other cause of CSF fistula. Such headaches should also resolve within 72 hours of epidural blood patching.

The estimated annual incidence of SIH is 5 per 100,000 with a female to male ratio of 2:1 and a peak incidence in the 3rd to 4th decades of life [3]. Spontaneous CSF leaks are the most common attributable cause [5]. While the exact cause of spontaneous spinal CSF leaks is largely unknown, an underlying structural weakness of the meninges is suspected [6]. A connective tissue disorder may underlie the meningeal weakness, as evidenced by the association between spontaneous intracranial hypotension and several known connective tissue disorders, such as autosomal dominant polycystic kidney disease, Marfan syndrome, neurofibromatosis type 1 and Lehman syndrome [5]. A history of trivial trauma accompanies approximately one third of diagnoses [5]. Extradural pathology, such as herniated discs, osteophytes and ossified posterior longitudinal ligament may also contribute to dural tears [3].

Spontaneous CSF leaks most commonly occur at the cervicothoracic junction or within the thoracic spine [5]. Although no specific data is available regarding the most common orientation of leakage at these levels, our experience suggests that dorsal sites of spontaneous CSF leakage are uncommon.

Diagnosis of SIH is suggested by characteristic findings on head MRI, including subdural fluid collections (hygromas), diffuse pachymeningeal enhancement, engorgement of venous structures, pituitary hyperemia and downward displacement of the brain [7]. Given the spectrum of imaging findings, the differential diagnosis would include meningitis, chronic subdural hematoma and dural venous sinus thrombosis. Meningeal metastases and meningeal sarcoid are less likely due to the smooth nature of pachymeningeal thickening in SIH. In the absence of clinical findings to support intracranial hypotension, idiopathic hypertrophic cranial pachymeningitis would be a consideration.

The etiology of the imaging findings described in SIH is proposed to be secondary to decreased intracranial CSF volume [8,9]. The Monro-Kellie doctrine states that the sum of the volumes of intracranial blood, CSF and brain tissue must remain constant in an intact cranium [10]. Compensation for a loss of intracranial CSF is accomplished by venous engorgement, which results in secondary effects of pituitary gland enlargement, meningeal thickening and enhancement and subdural effusions [11]. The loss of CSF volume also reduces buoyancy forces acting on the brain, resulting in its physical descent when the patient is upright. The descent of the brain can produce traction on pain-sensitive structures such as cranial nerves, likely resulting in the orthostatic nature of the clinical findings [9]. This is also responsible for the characteristic imaging features of a "slumping midbrain" and downward displacement of the cerebellar tonsils [7].

Recently, a decreased "venous hinge" angle (VHA) has been described as an objective sign of SIH [12]. The internal cerebral veins (ICVs) converge with the basal veins of

Rosenthal to form the vein of Galen (VOG). The VHA, measured between the ICV and VOG, is decreased in SIH due to the downward displacement of the ICV while the VOG remains relatively fixed at the tentorial insisura. A VHA of less than 79 degrees was found to be 88% sensitive and 92% specific for SIH.

Spinal MRI may demonstrate extradural fluid collections [13]. Myelography with iodinated intrathecal contrast followed by CT may identify a site of CSF leakage through a spinal dural defect. However, CT myelography lacks sufficient temporal resolution in cases where a high volume leak allows equilibration between the intra- and extra-thecal contrast before the CT is performed. Dynamic CT myelography, with infusion of intrathecal contrast during CT scanning, has been described to address the challenge of localizing high flow CSF leaks. The limitation of this technique is the elimination of real-time fluoroscopic monitoring of contrast during injection [14].

Conventional myelography supports accurate, real-time temporal resolution but is limited in spatial resolution depending upon patient positioning and available equipment. Digital subtraction myelography has been described and has application in the diagnosis of CSF leaks with large volumetric flow rates [16]. In both techniques, the need to inject contrast during the procedure limits patient positioning to prone or lateral decubitus postures due to the maintained presence of the spinal needle.

The clinical and imaging characteristics of intracranial hypotension have been well described and are an increasingly recognized cause of new daily persistent headaches in middle-aged patients [15]. In our case, the patient met all clinical and imaging criteria for diagnosis of a spinal CSF leak. While many cases may resolve spontaneously, primary treatment is by epidural blood patch with surgical correction of dural pathology reserved for refractory cases [3]. As our patient failed to obtain lasting relief from conservative management, surgical planning necessitated accurate documentation of any sites of CSF leakage.

With our case, the presence of a large volume of extradural fluid on MR and ready extravasation of contrast on conventional and CT myelography confirmed a dural CSF leak. While dorsal sites of leakage were suspected, they were not definitively diagnosed using conventional imaging procedures. To identify the levels of involvement and confirm their dorsal location, we postulated that performing myelography in real time with the patient in a supine position would allow the effect of gravity to better demonstrate flow from dural tear(s). Our technique involved the placement of a lumbar drain to inject contrast with the patient in the supine position. Using this method, contrast extravasation was rapidly and accurately identified at two dorsal sites, confirmed at surgery.

TEACHING POINT

Digital subtraction myelography is a useful adjunctive tool to conventional and CT myelography when spinal CSF leaks are present. In the case of a dorsal CSF leak, isolation of leakage sites may be enhanced when imaging is performed in the supine position. Injection of contrast through a lumbar drain instead of a spinal needle allows for supine positioning to detect dorsal CSF leaks.

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FIGURES

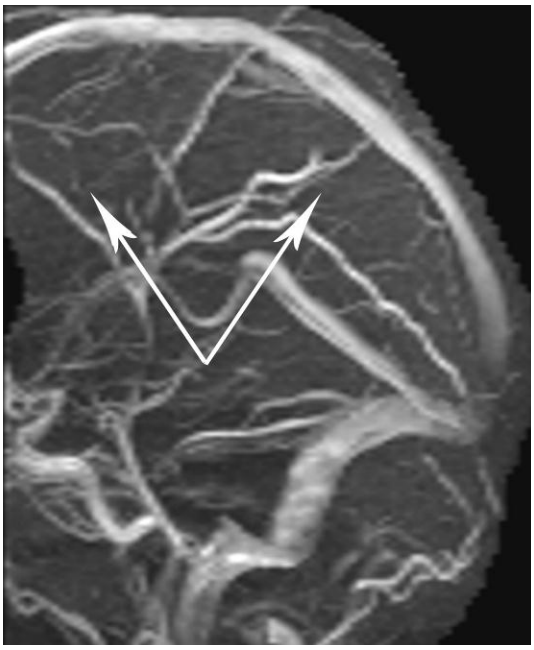


Figure 2 (top): 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. Sagittal midline image from MR venogram (1.5 Tesla magnet; TR, 22 msec; TE, 4.4 msec) demonstrates decreased venous hinge angle of 70 degrees.

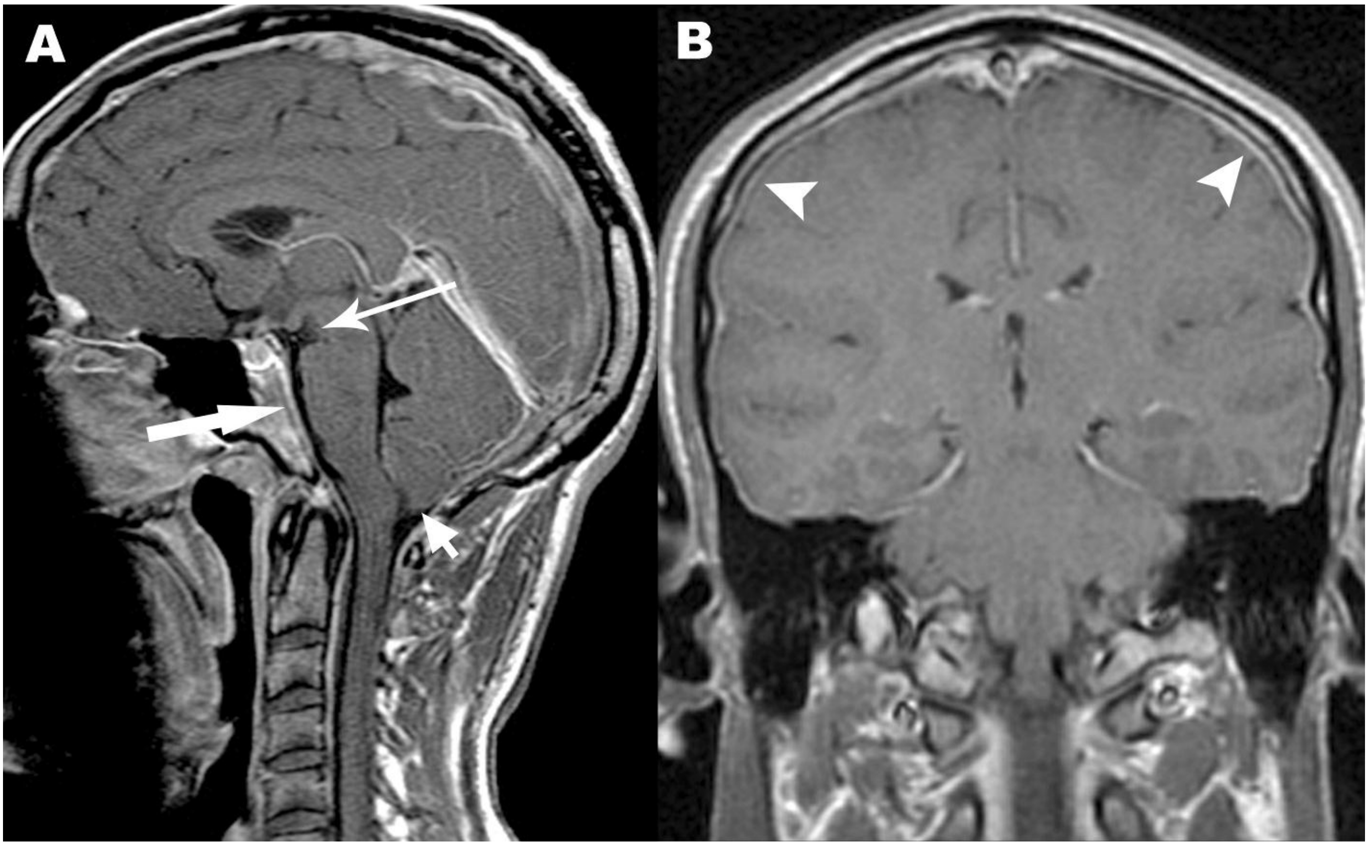


Figure 1: 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. A. Sagittal midline T1-weighted MRI (1.5 Tesla magnet; TR, 517 msec; TE, 9 msec; 7.7 ml Magnevist) contrast enhanced image of the head demonstrates caudal displacement of the cerebellar tonsils (short arrow) and slight flattening of the anterior pons (thick arrow) and decreased mammilopontine distance (long thin arrow). B. Coronal T1-weighted MRI (1.5 Tesla magnet; TR, 517 msec; TE, 9 msec; 7.7 ml Magnevist) contrast enhanced image shows diffuse pachymeningeal enhancement (arrowheads).

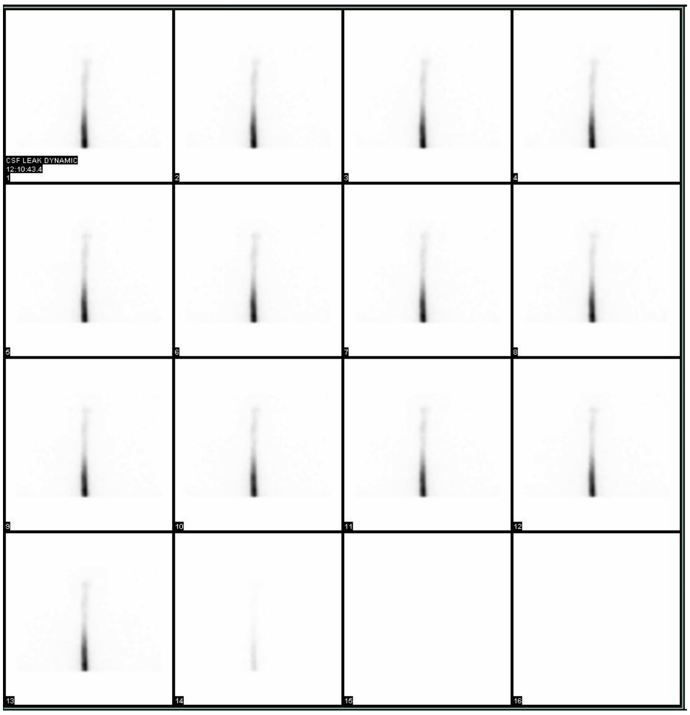


Figure 3 (left): 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. Nuclear Medicine CSF leak study performed with intrathecal injection of 138 MBq of technetium 99m labelled DTPA. Static and dynamic images were obtained and no evidence of CSF leakage is demonstrated.

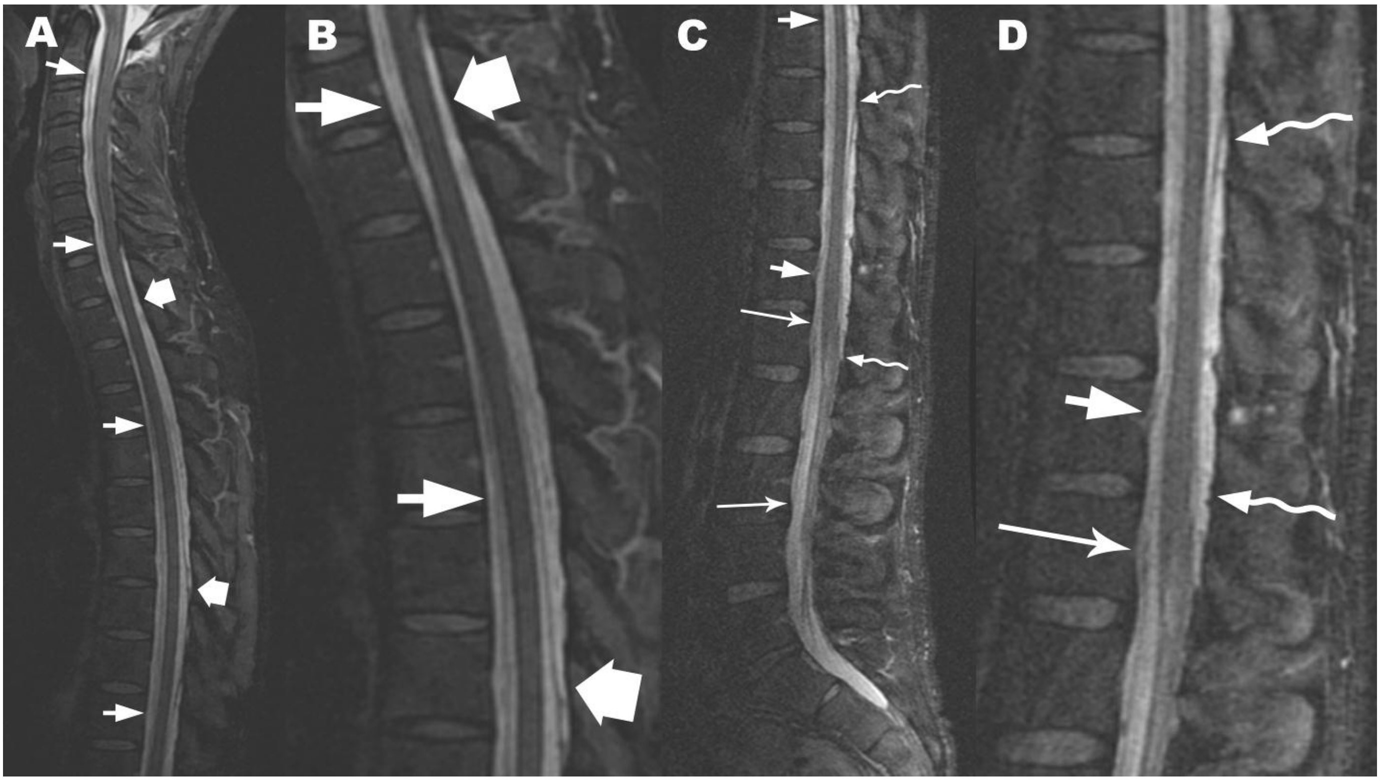


Figure 4: 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. A. Sagittal FIR MRI (1.5 Tesla magnet; TR, 9,730 msec; TE, 35 msec) image of the cervical and upper thoracic spine shows high T2 extra-dural collections ventrally from C2 to T12 (arrowheads) and dorsally from C7 to T8 (thick arrows). B. Magnification of Image A. C. Sagittal FIR MRI (1.5 Tesla magnet; TR, 6,206 msec; TE, 37 msec) image of the lower thoracic and lumbar spine delineates the lower aspect of the collections identified in A) and an additional dorsal collection from T9 to L2. D. Magnification of Image C.

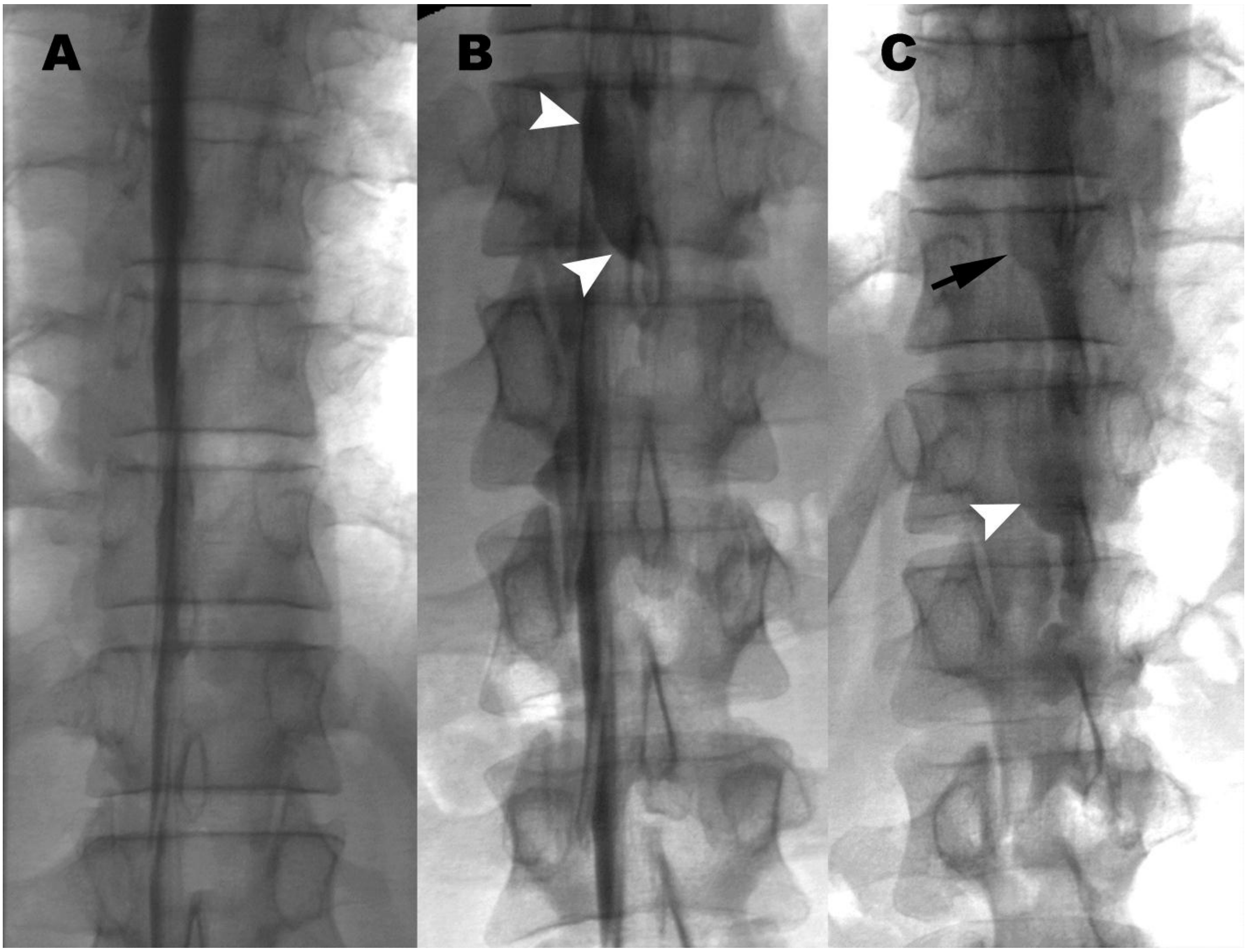


Figure 5: 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. Images from conventional myelogram (10 cc Omnipaque 300) A. With patient early in left lateral decubitus position shows no extradural contrast extravasation. B. With patient later in left lateral decubitus position, there is extradural extravasation of contrast at the level of T12 (white arrowheads). C. After rolling the patient into the right lateral decubitus position, contrast is again seen at T12 (white arrowhead) and a new extradural collection is visualized at T10-T11 (black arrow). Clear visualization of the site of leakage is not achieved.

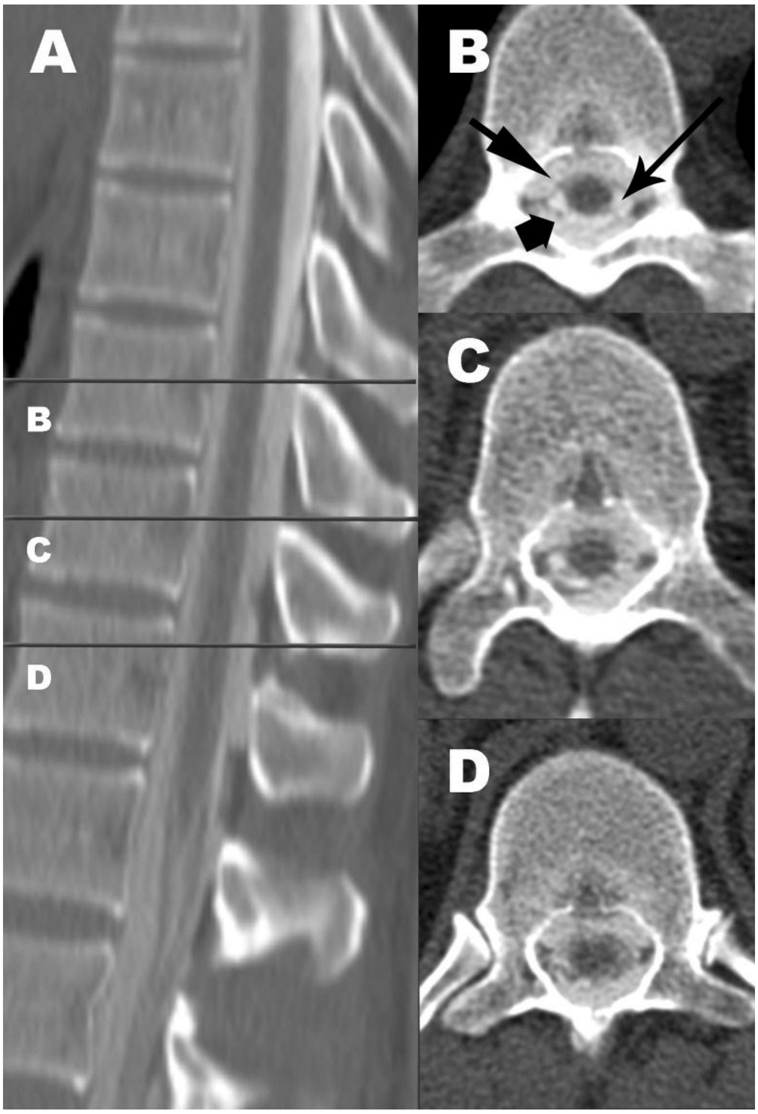
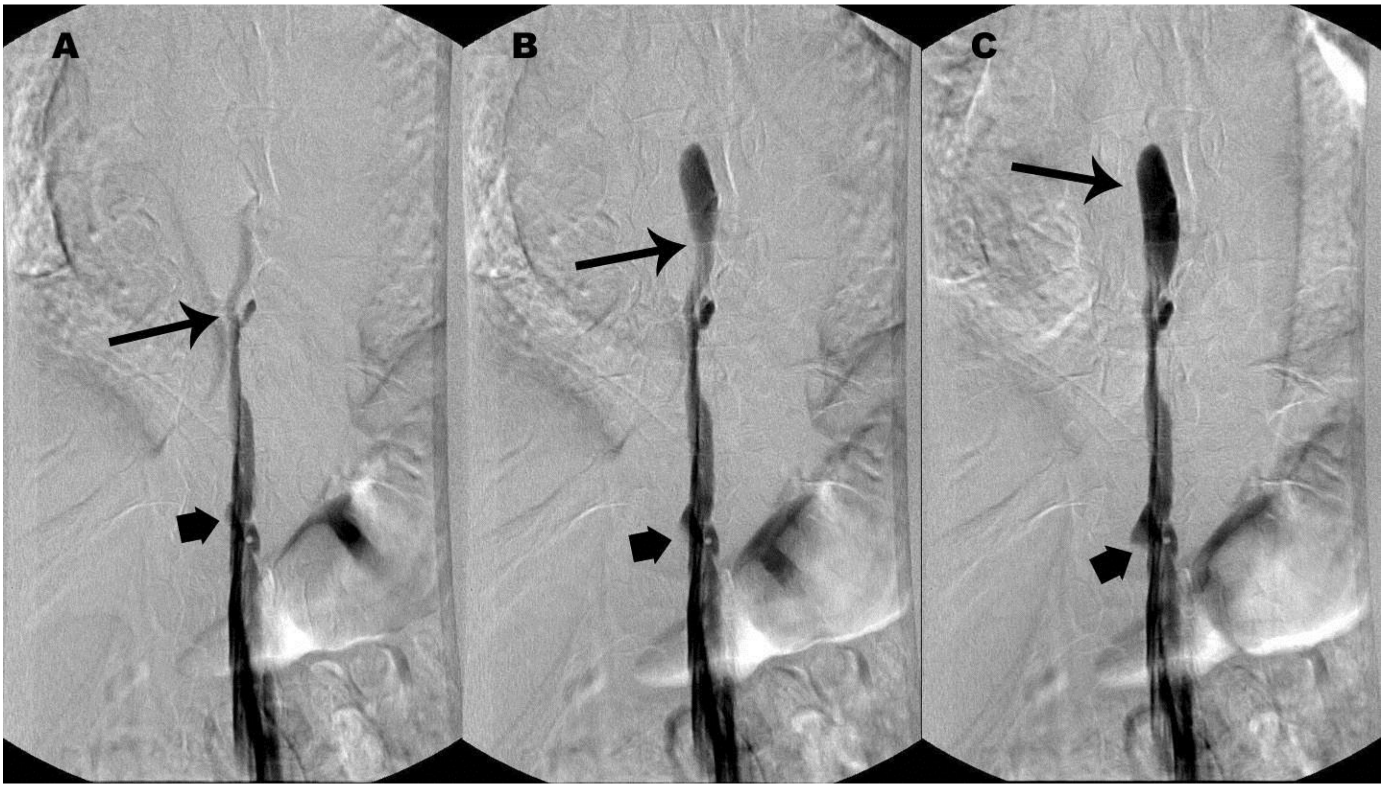


Figure 6 (left): 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. A. Sagittal post-myelography CT (10 cc Omnipaque 300 intrathecal; 64 slice GE Lightspeed scanner; 120 kVp; 200 mA; 5 mm slice thickness) image from T8 to L1 showing extrathecal contrast from T9 to T12 dorsally, confirming the presence of an active CSF leak. B. Axial image at T10 shows ventral (large arrow) and dorsal (fat arrow) extradural collections. Contrast is present within the thecal sac (skinny arrow). C. Axial image at T11 shows further effacement of the thecal sac. D. Axial image at T12 shows almost complete effacement of the thecal sac.

Figure 7 (bottom): 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. A through C. Progressive, frontal projections from a digital subtraction myelogram (8 cc intrathecal Omnipaque 300) performed with the patient in the supine position as contrast was injected through a lumbar drain. Two sites of active contrast extravasation are identified at T9-10 (thin arrow) and T11-12 (thick arrow).



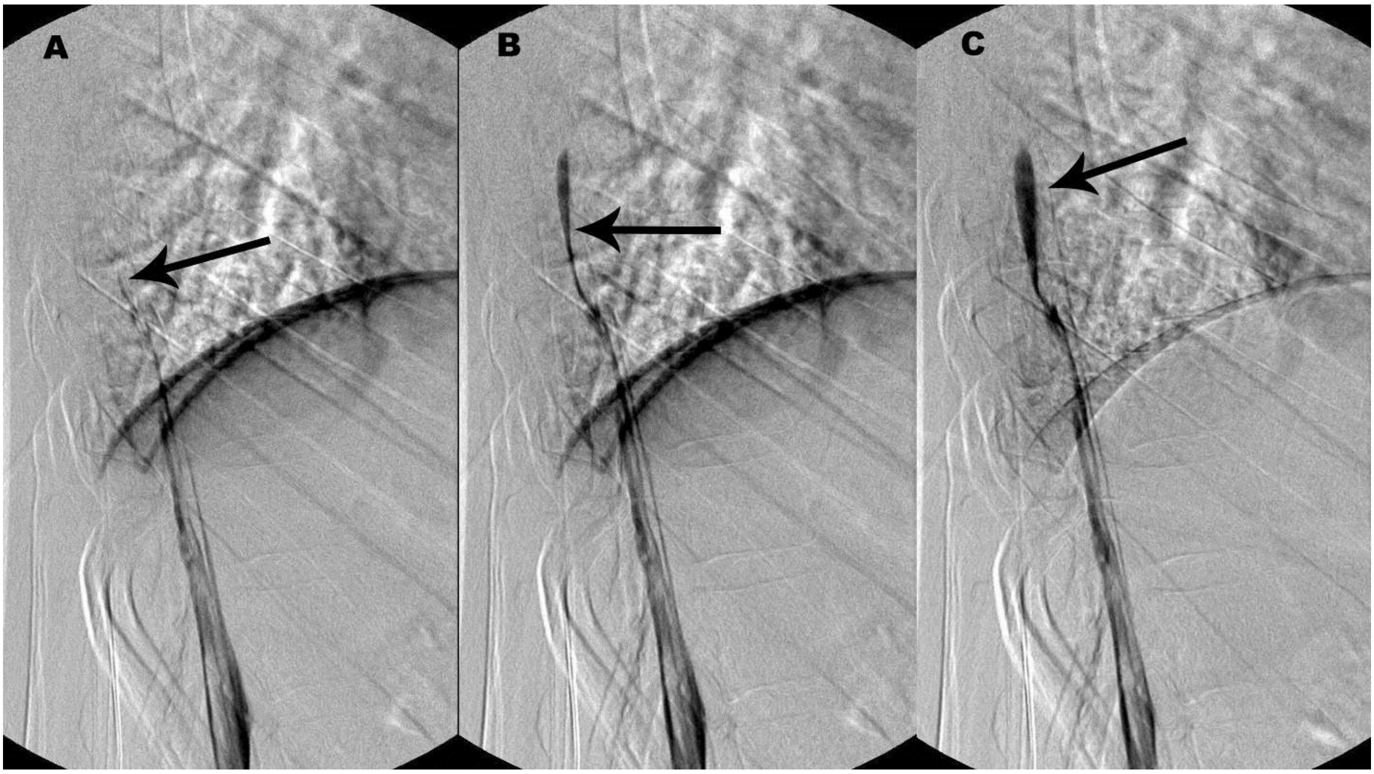


Figure 8: 34 year old male patient with spontaneous intracranial hypotension and dorsal cerebrospinal fluid leak. A through C. Progressive, frontal projections from a digital subtraction myelogram (8 cc intrathecal Omnipaque 300) performed with the patient in the supine position as contrast was injected through a lumbar drain. Active contrast extravasation at T9-10 (arrows) confirms the dorsal location of the leak. The leak at T11-12 is not well visualized in this position.

Etiology	• most common cause of SIH is a spinal CSF leak through a dural tear • decreased CSF production and increased absorption may also be possible causes
Incidence	• estimated annual incidence is 5 per 100,000
Gender Ratio	• female to male ratio of 2:1
Age Predilection	• peak incidence is in the 3rd to 4th decades of life
Risk Factors	• connective tissue disorders, such as autosomal dominant polycystic kidney disease, Marfan syndrome, neurofibromatosis type 1 and Lehman syndrome • history of trivial trauma accompanies approximately one third of diagnoses of SIH • extradural pathology, such as herniated discs, osteophytes and ossified posterior longitudinal ligament may also contribute to dural tears
Treatment	• while spontaneous recovery occasionally occurs, primary treatment is by epidural blood patch • surgery is reserved for refractory cases • dural defects can be repaired by suture or with placement of fibrin glue • fascial grafts may be required for large defects
Prognosis	• no specific data are available • low CSF pressure headache recurs in approximately 10 percent of patients regardless of the treatment methodology used
Findings on Imaging	• MRI – T1: demonstrates sagging midbrain, decreased venous hinge angle, enlarged pituitary due to hyperemia and engorgement of venous structures • MRI – T1 C+: enhancement of thickened dura • MRI – T2: subdural collections (variable signal), thickened dura (usually hyperintense) • CT Head: may show subdural collections • CT Myelogram: may show extradural contrast collection • conventional myelography: may show active extravasation from dural leak

Table 1: Summary table for spontaneous intracranial hypotension

	Spontaneous Intracranial Hypotension	Meningitis	Chronic Subdural Hematoma	Meningeal Metastases
CT	• +/- subdural collections • spinal CT post-myelogram may show extradural contrast extravasation	• m/c commonly normal • +/- mild ventricular enlargement	• subdural collection of varying density	• biconvex dural mass +/- displacement of underlying brain • CT insensitive for carcinomatosis
MRI-T1	• sagging midbrain • decreased venous hinge angle • enlarged pituitary due to hyperemia • venous engorgement	• isointense sulcal exudate	• subdural collection of varying signal intensity, depending upon age of blood products	• hypointense to gray matter
MRI – T1 C+	• enhancement of thickened dura	• enhancement of exudate and pia	• +/- dural enhancement	• enhancement of dural masses • nodular enhancement in carcinomatosis
MRI-T2	• subdural collections • thickened dura (usually hyperintense)	• may see hyperintense exudate in sulci	• subdural collection of varying signal intensity, depending upon age of blood products	• usually hyperintense to gray matter
MRI-DWI	• no specific findings	• diffusion restriction if empyema formation	• variable diffusion restriction	• may show restricted diffusion
Myelography	• may see extravasation of contrast through a spinal dural tear	• if spinal involvement, may see block of CSF flow and irregular contour of thecal sac	• no specific abnormality	• if spinal involvement, • single of multifocal nodular filling defects • expanded cord • thickened nerve roots

Table 2: Differential diagnosis table for spontaneous intracranial hypotension

ABBREVIATIONS

CSF = Cerebrospinal fluid
CT = Computed tomography
DTPA = Diethylene triamine pentaacetic acid
FIR = Fluid attenuated inversion recovery
ICV = Internal cerebral vein
IHS = International Headache Society
MBq = Megabecquerel
MR = Magnetic resonance
MRI = Magnetic resonance imaging
SIH = Spontaneous intracranial hypotension
TE = Echo time
TR = Repetition time
VOG = Vein of Galen

KEYWORDS

Spontaneous intracranial hypotension; digital subtraction myelography; CSF leak