Acute Supratentorial Subdural Hematoma after Craniocervical Junction Arachnolysis in a Patient with Posttraumatic Syringomyelia; Case Report and Literature Review

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Abstract

A 37-year-old woman with chronic progressive myelopathy and syringomyelia and scoliosis presented with difficulty in walking at presentation. Dorsal CVJ CSF collection and adhesive arachnoiditis, compressing cerebellum, brainstem and upper cervical spinal cord with holocord syrinx was detected in neuroimaging beside thoracic scoliosis.

INTRODUCTION

Spinal adhesive arachnoiditis (SAA) is defined as collagen deposition in pia matter of arachnoid space due to inflammation caused by various etiologies (1). Trauma, infection, subarachnoid hemorrhage (SAH) and iatrogenic causes such as surgery or epidural/subdural injections are among the most reported causes in literature (1-3). It has gained various names in the literature like: foramen magnum arachnoiditis (FMA), basal arachnoiditis, hindbrain arachnoiditis, chronic arachnoiditis at the foramen magnum, and arachnoiditis at the craniocervical junction (3-5). SAA is associated with different clinical presentations including progressive myelopathy, quadriparesis, spastic gate, syrinx formation, myelopathy and radicular symptoms (1, 2, 5-8). Although numerous theories have been presented, the most agreed and common basis is cerebrospinal fluid (CSF) accumulation distal to its normal passage blockade site in central canal or spinal cord interstitial space (9-12).

Intracranial hematoma, remote from surgical site, has been reported in literature. These include epidural (EDH), subdural (SDH) or intraparanchymal hematomas following various brain or spine surgeries. Acute supratentorial SDH following posterior fossa surgery or CSF diversion surgeries has rarely been reported before (6, 7, 13-21). Here we present the first case report of acute supratentorial SDH following arachnolysis of delayed posttraumatic CVJ adhesive arachnoiditis and our hypothesis and suggestions based on literature.

CASE PRESENTATION

History and Physical Examination

A 37-year-old single lady from southern Iran was referred to our clinic due to progressive spastic gate and apraxia as well as progressive thoracolumbar scoliosis for the past 6 months prior to presentation. She had no significant past medical, family or drug history except a falling object head trauma 5 years ago. We found abnormal thoracic curvature and lower extremity proximal weakness (3/5). She also had right upper extremity radicular pain and proximal weakness (4/5). Plantar reflexes were upward bilaterally along with

positive Hoffmann's sign. All the deep tendon reflexes (DTR) were exaggerated (3/4). She could walk only with assistance utilizing a walker.

Imaging

Whole spine magnetic resonance imaging (MRI), was conducted revealing a large syrinx extending from C2 down to lower thoracic levels (Fig. 1A), a large S-shape scoliosis with lower curve at L1 with convexity to right and upper curve at T6 with convexity to left (Fig. 1B). In brain MRI, it was revealed that a septate cystic mass in dorsal aspect of CVJ is pushing 4^{th} ventricle and uppermost cervical spinal cord segments, causing signal changes (Fig. 1C). A CVJ junction Cine MRI, was also conducted revealing a septate thin-wall cystic structure (18*12*9 mm) in posterior aspect of CVJ, causing CSF flow compromise, in association with extensive syringomyelia, without any evidence of hydrocephalus. There was normal CSF flow in anterior aspect of CVJ. Delayed post traumatic CVJ adhesive arachnoiditis, compressing fourth ventricle, brainstem and upper cervical spinal cord, causing alterations in CSF circulation and dynamics, was diagnosed and the patient was scheduled for CVJ decompression, durotomy, and arachnolysis in an elective setting.

Surgery

In standard prone position with her head fixed in Mayfield head-holder in slight neck flexion the patients underwent bilateral suboccipital craniotomy. About 3×3cm midline suboccipital craniectomy, including dorsal rim of Foramen Magnum, in addition to C1 partial laminectomy was done. Dura was opened in linear fashion, 5mm from upper edge of craniectomy down to C1 lamina. There was a collection at dorsal aspect of CVJ, covering with numerous thick arachnoid webs, distorting normal anatomy of neurovascular structures in the area, without normal pulsation of arachnoid membranes. Then, microscopic arachnolysis with preservation of tethered arteries and nerves in arachnoid membranes was performed until floor of 4^{th} ventricle was seen. then arachnolysis was continued caudally to reach normal dorsal spinal SAS. After microscopic coagulation of arachnoid webs with preservation of neurovascular structures, direct passage of CSF from 4^{th} ventricle to spinal SAS was seen and confirmed. After being positive about clearance of intraventricular and subarachnoid fluids from hematoma, duroplasty with autologous fascial graft was performed and wound was closed.

Postoperative course

We had no major bleeding or hemodynamic catastrophic events during surgery. A postoperative brain Computed Tomography (CT) scan was performed before transferring to Intensive Care Unit (ICU) that revealed right frontoparietal (FP) acute SDH with nearly 5 mm right to left midline shift (Fig. 3A). The patient transferred to operation room for surgical evacuation of SDH. The patient's physical examination was a GCS score of 7/15 and anisocoria (right: 6mm and left: 3 mm). The patient underwent right frontoparietal craniotomy for SDH evacuation. About 40 cc acute SDH was evacuated and cranioplasty was performed. The postoperative brain CT-scan was acceptable (Fig. 3B). The postoperative MR images of the CVJ revealed that the CSF flow was restored and the CSF blockade was extensively alleviated being associated with decreased size of the cervical syrinx (Fig 3A, B).

Outcome

The patient retained his normal consciousness a day after surgery and pupillary reflexes were bilateral and normal. She developed CSF leakage from CVJ surgical site at the 4^{th} postoperative day, that was managed with insertion of lumbar drainage tube and medications. After 3 weeks of postoperative care, neurological examination improved significantly and she could walk without assistance. Her right upper extremity radicular pain and weakness were also resolved. She was discharged with both surgical wounds healed, having no sign of infection, no CSF leakage or subcutaneous collection at the 23^{rd} postoperative day.

Follow-up

Serial neurologic examination during 6-months postoperative period was acceptable and no new neurological deficits were observed. We obtained serial OPD visit and brain CT scans with two episodes of brain and spine

MRIs which revealed no hydrocephalus or hematoma and partial resolution of syringomyelia. The patient myelopathy and spastic gate improved significantly.

DISCUSSION

CSF Circulation and Pressure

CSF, as the protector of mammalian CNS, is mainly (75-90%) produced by choroid plexus of lateral ventricles and other structures such as brain parenchyma, ependymal cells and interstitial fluid. CSF flows from lateral ventricles to 3rd ventricle though foramina of Monro. Then, passing from cerebral aqueduct, enters 4th ventricle and leaves it from (lateral) foramina of Lushka to cerebral cisterns and SAS; and (medial) foramen of Magendie to spinal SAS. CSF flow is also present between cerebral and spinal SAAs. Its absorption takes place at Superior Sagittal Sinus (SSS) arachnoid granulations and Virchow–Robin space of peripheral SAS (22, 23). Any change in CSF production, absorption or circulation can alter Intracranial Pressure (ICP) and therefore Cerebral perfusion Pressure (CPP) and Cerebral Blood Flow (CBF) autoregulation (22, 24).

Syringomyelia

Syringomyelia has been classified and accepted widely into two subgroups: communicating and non-communicating (25). Normal CSF outflow from 4th ventricle is mainly to SAS and slightly from central canal (22-24). There is also a significant pressure difference between ventricles and central spinal cord canal which causes "suck" CSF from 4th ventricle into the central canal. It is now well known that CSF goes both ways between cranial and spinal paths; defining that in some raised intraabdominal or thoracic pressure instances, CSF flows upward from spinal central canal and SAS to cranial intra- or extraventricular CSF containing spaces (22-25). Communicating syringomyelia is a condition of backflow resistance of CSF from spinal central canal or SAS to cranial regions. Some reasons have been shown to be responsible for these situations, like: trauma, infection, postsurgical and due to Thecal sac injections or manipulations (4, 5, 9). Communicating syringomyelia can be diagnosed in radiological investigations. Upward extension of syrinx to uppermost cervical segments, CSF flow in dynamic imaging and contrast passage to syrinx cavity are among radiologic confirmatory findings of communicating syringomyelia (9, 25).

Adhesive Arachnoiditis

SAA is a condition of forming multiple arachnoid membranes due to various pathologies, mainly caused by inflammation (1, 5). It can lead to neural structures compression due to tethering, mass effect and alteration in CSF circulation and dynamics (2-4). In CVJ, mainly due to TB and sarcoidosis, it has been associated with syrinx and tethering of cerebellum and brainstem in literature which can lead to neurological minor to major deficits (2-5).

Our case and Hypothesis

Here we presented our case of foramen magnum SAA due to previous head trauma. Forming numerous arachnoid membranes in CVJ, it altered CSF circulation in cranial-spinal regions beside local accumulation of CSF in dorsal aspect of FM with compression effect on brainstem and 4th ventricle. It obviously caused resistance in CSF flow and higher-pressure gradient between cranial and spinal regions, documented in CSF dynamic study and syrinx formation. As mentioned in literature, decompression of FM structures and release of arachnoid adhesions, are mainstay of treating such conditions and has shown good clinical and syrinx response (25). We propose that, rapid evacuation of the CSF from the axis, will result in subdural vein sagging and increased venous pressure followed by ruptured of the veins and SDH formation. The mechanism of the SDH in cases who undergo CSF diversion and also CSF drainage is similar. In these patients, prevention of the CSF rapid drainage and replacement of the CSF with saline should be performed to prevent this catastrophic event.

We assumed that CSF dynamics in such situations adapts with a higher-pressure gradient; explaining that CSF flow resistance in CVJ, causes higher cranial and lower spinal CSF pressure. CSF outflow and absorption adaptations, may cause higher cranial CSF pressure without ventriculomegally or other imaging findings of

high ICP as we see in Normal Pressure Hydrocephalus (NPH) or Idiopathic Intracranial Hypertension (IIH) (22, 23). Because of this compensation, brain circulation and autoregulation adapt to higher pressures as well. Thus, such conditions are associated with higher arterial blood pressure to maintain cerebral perfusion pressure and therefore higher venous pressure to keep venous flow normal.

Our suggestion is to consider these compensations in such conditions to prevent acute events occurring after rapid intracranial pressure change due to surgeries. As it is the first case report of this scenario, we suggest reporting every detailed event in such situations and their follow-up with review studies of similar conditions to reach better knowledge of this events.

Conclusion

SAA, caused by different etiologies with main step of inflammation, leads to alterations in CSF circulation and neural structure tethering or compression. In CVJ, beside brainstem and cerebellum compression, spinal syrinx is a common sequence which can be associated with scoliosis and neurologic deficits. Foramen Magnum decompression and arachnolysis with direct observation of CSF circulation from 4^{th} ventricle to spinal subarachnoid apace, is acceptable and widely used treatment so far. With occurrence of post operational acute supratentorial subdural hematoma in our case, we theorized this surgery to be associated with rapid intracranial change, which has been adapted to a higher pressure as a result of chronic CSF outflow resistance. Rapid decrease in CSF volume of the brain leads to superficial vein sagging and subsequent rupture and hematoma formation.

Informed written consent

The patient and their legal guardians provided their written consent for publication of the manuscript and the case for research purposes.

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Figure Legends

Fig. 1 Sagittal T2-weighted MRI of the cervicothoracic spine demonstrating a large holocord syrinx (arrow) extending from C2 to the conus medullaris (A); coronal T2-weighted MRI of thecervicothoracic spine demonstrating a large S-shape scoliosis with lower curve at L1 with convexity to right and upper curve at T6 with convexity to left (B); sagittal T2-wighted MR images of the brain demonstrating severe adhesive arachnoiditis (arrow) and CSF blockade in craniocertebral junction (CVJ) (C).

Fig. 2 Axial brain CT-scan of the patient after CVJ decompression revealing right frontoparietal acute sundural hematoma with 5 mm midline shift (A); postoperative axial brain CT-scan of the patient demonstrating complete evacuation of the right subdural hematoma without midline shift (B).

Fig. 3 Sagittal T2-weighted MRI of the cervicothoracic spine demonstrating suboccipital craniotomy and removal of the C1 arc with free CSF flow in the CVJ (arrow) (A); sagittal T2-wighted MR images of the cervicothoracic juction demonstrating decreased size and diameter of the holocord syrinx when compared to preoperative images (arrow) along with normal CSF flow (B).

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