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Case Report Open Access

Spontaneous Intracranial Hypotension due to Fistula of a Sacral Meningocele

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Abstract

Spontaneous Intracranial Hypotension (SIH) is a syndrome characterized with reduction of amount and pressure of Cerebro Spinal Fluid (CSF) due to leakage. Orthostatic headache is the most significant symptom. Central nervous system pressure below 60 mmH_2O is ordinary.

Primary CSF leakage and dural rupture develops spontaneously or following minor trauma, coughing, sneezing, straining and stretching.

We present a 51-year-old female case with sacral lipoid meningocel. She had chronic headache and constipation for years and we estimated that CSF leakage developed due to strain. The symptoms regressed following the excision of meningosel pouch.

Keywords: Meningocel; CSF leakage; Intracranial hypotension; Headache

Introduction

Spontaneous intracranial hypotension (SIH) develops due to CSF leakage without serious trauma and the most prominent symptom is known to be postural orthostatic headache.

CSF leakage develops primarily due to unknown minor trauma, couhing, sneezing, straining and stretching.

It develops secondarily to Lumbar Punktion (LP), trauma, postoperative over drainage. It accompanies to disease due to genetic factors such as collagen tissue disorders, Marphan Syndrome, Ehlers-Danlos Syndrome, polycystic kidney, arachnodactyly, joint hypermobility and degenerative diseases. CSF leakage mostly develops due to rupture at the exit site of spinal nerve from arachnoid membrane and precipitate the herniation of cerebral and cerebellar compositions [1-8].

Typically, the pain starts in 15 minutes after standing or the existing pain increases. It resolves after 30 minutes of resting. It was first described at 1938 by Schatenbrand as Liquorrhea. Intracranial pressure is generally below 60 mmH_2O. It frequently occurs at the 4th decade and two times more in females. The prevalence is known to be 2/100000. Main symptom is the orthostatic headache. Neck pain, mild neck stiffness, cranial nerve paralysis, dizziness, vertigo, nausea, vomiting, diplopia, phonophobia, tinnitus, instability, numbness of face are the other symptoms that may occur due to intracranial hypotension. Delay in diagnosis may result with subdural hematoma. Parkinson, ataxia, herniation, stupor and coma may also occur. Cases with subdural cerebellar hemorrhage, quadriplegic, galactorrhea are reported.

History, Magnetic Resonance Image (MRI), LP, myelography, radionucleocysternography are important for diagnosis. Conservative or surgical therapy can be applied [3,9-18].

Case Report

A 51-year-old female patient admitted to our clinic with a complaint of pulsatile and orthostatic headache for 3 years. Sometimes nausea and vomiting were accompanying headache. She was examined by several doctors, but no therapy was successful. Cranial MRI findings found to be normal.

Figure 1: Fistulated sacral lipomeningocele.

Her systemic and neurologic examination was normal. We considered orthostatic hypotension due to low CSF pressure and planned to perform lumbar punction (LP). As we opened the lumbar region of the patient to perform the LP, we saw the lipoid meningocel (Figure 1).

The patient never mentioned about this lipoid structure at her back. She had that structure since childhood, and the region was always dry until the beginning of chronic constipation problems for 3 years. After that, when she had to strain due to constipation, the lumbar area was wet. The beginning of chronic, pulsatile and orthostatic headaches was also at the same period of time.

CSF analysis was found to be normal and the pressure was 35 mmH_2O. Lumbosacral MRI revealed the relation between dural sac and fistulae (Figure 2).

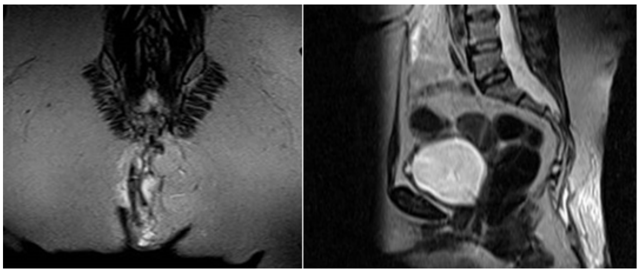

Figure 2: A-B: Lumbar MRI indicating fistulated sacral lipomeningocele, connected to the dural sac.

We indicated the operation of meningocel resection. Sacral meningosel sacculus is removed and closed at the entrance of the spinal canal. There were no neural elements in the sac (Figure 3).

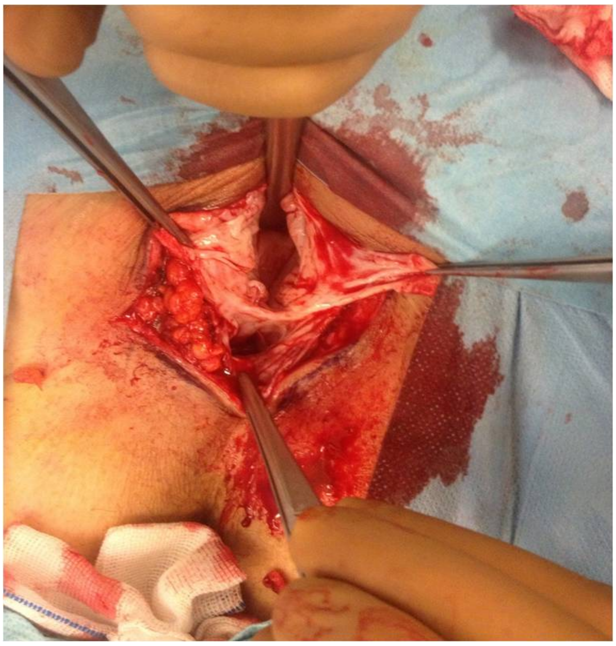

Figure 3: Meningocele sac excision.

After the resection, the headache symptoms are released and there was no CSF leakage. We recommended laxative and discharged the patient from the hospital. At 3 months follow up, headache symptoms were completely gone.

Discussion

Decrease in CSF volume and pressure due to leakage results with inferior displacement of intracranial composition, distention and compression of pain sensitive structures. Traction of subdural veins and arteries supplying the brain and the tentorium causes bleeding and postural headache as seen in our patient. Neck stiffness, interscapular pain, nausea, vomiting, vertigo, horizontal diplopia, photophobia, visual disturbance accompanies headache rarely. Symptoms can be aggravated with coughing, sneezing and jugular compression [3,12,15,19-22]. Besides headache, our case had also nausea and vomiting. Cranial MRI scans were found to be normal. We can assume that, rupture site was away from culdosac, leakage was in small amounts and displacement of cerebral structures were minimal.

It is reported that structural weakness of meninges, plays an important role in intracranial hypotension. Therefore, stretching, sneezing, events provoking Valsalva maneuver, sports, collagen tissue disorders may trigger the start of symptoms. Most of the cases have joint laxity [4-6,14,18,21,23]. Our case had congenital lipoid meningocele and the skin around the sac was quite thick. She did not have a cosmetic complaint due to the meningocele and did not mentioned about it to any of the physicians. However, constipation at the last 3 years obliged her to strain and CSF fistulized from the skin. We did not observe neurological deficits, sphinctary reflexes were found to be normal. In cases with SIH, CSF pressure is lower than 60 mmH_2O. Pressure measurement with LP is essential for definitive diagnosis. To demonstrate the leakage with Cranial MRI and Computed Tomography (CT), myelography and radio isotope cysternography is important for the differention and confirmation of diagnosis. Cranial MRI may reveal signs of meningeal thickening, contrast enhancement, subdural haematoma or hygroma [1,3,13,15,16,20,24]. Cranial MRI of our patient was found to be normal.

Conclusion

Thin sliced spinal CT or MRI reveals meningial diverticules, enlargement of dural veins, site of CSF leakage, as in our patient [13,25]. Leakage from the dural sac to the sacral lipoid meningocele sac is demonstrated in our case.

Most of the cases recover with conservative therapy. Hydrostatic pressure decreases with bed rest and small dural defect with smooth border can heal. Cafein, theophilyn and steroid treatment can be applied. Epidural 15-20 cc autologous blood can be injected to the site of fistula in cases resistant to therapy [3,11,12,15,17,22,26].

Operation must be considered as the last procedure. It can be applied to cases resistant to conservative therapy. Operation area can be covered with foam gel and fibrin products. Our case had non-operated sacral lipoid meningocel, so we decided to perform the surgery, meningocel sac was dissected through the fistula and sutured its entrance to the dural sac. The symptoms of the patient fully recovered after 3 months.

In patients with SIH and orthostatic headache, CSF leakage due to sacral meningocel must be kept in mind, especially in female patients which may be unwilling to show this area.

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