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Spontaneous Intracranial Hypotension

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ABSTRACT

Introduction: Out of all the painful conditions, headache is one of most common cause of patients coming to hospital. Usually few patients present with orthostatic headache, which signifies low cerebrospinal fluid pressure. **Case description:** A 38-year-old female presented with intense headache in upright position, relieved on lying down in supine position. She was treated with analgesics and epidural blood patch. **Discussion:** Spontaneous intracranial hypotension is one of the treatable causes of headache. It can be detected by magnetic resonance imaging, which can reduce the morbidity and improve quality of life. **Conclusion:** As there is increase in availability of MRI, spontaneous intracranial hypotension can be detected easily.

Keywords: Orthostatic headache, subdural hygroma, epidural blood patch

Spontaneous intracranial hypotension is one of the rare causes of headache. The most characteristic feature is the relative change in intensity of headache with change in posture. The intensity of headache is more in the upright and standing position, relieved in the recumbent position (orthostatic headache). The headache occurs due to lowering of intracranial pressure caused by leakage of cerebrospinal fluid (CSF).

CASE HISTORY

A 38-year-old lady presented with headache for 2 weeks, which was severe in intensity, in bitemporal and occipital region, persisting throughout the day, aggravated in sitting and standing position, relieved by supine position. The headache was associated with pain in upper part of neck and vomiting. She had past history of hypertension with regular treatment with antihypertensives. She had no history of previous lumbar puncture, spinal surgery, congenital disease, connective tissue disorders and type 2 diabetes mellitus. No significant family history obtained.

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On neurological examination, she was conscious, oriented to time, place and person; cranial nerves, motor system, sensory system, cerebellar system, autonomic nervous system were normal. No meningeal signs were present. She was initially treated with analgesics, antidepressants, antihypertensives and intravenous fluid. But her headache was not getting relieved with the medications. Neurosurgery and ear, nose and throat opinion were normal. Routine blood investigations were done (blood and radiological). Her blood parameters were hemoglobin - 11.3 g/dL, total leukocyte count - 8,400/mm^3, random blood sugar - 108 mg/dL, serum creatinine - 1 mg/dL. Magnetic resonance imaging (MRI) of brain revealed thin subdural hygroma in both the frontal convexity, prominent venous sinus and draining cortical veins, especially right transverse sinus. Left lateral ventricle narrowed at the frontal horn (slit-like frontal horn) (Figs. 1 and 2).

These findings were suggestive of intracranial hypotension. She was treated with oral analgesics tablet paracetamol and diclofenac, tablet propranolol, capsule omeprazole and intravenous fluids, but there was not significant improvement. A follow-up MRI of brain revealed increase in volume of subdural collection in right frontoparietal convexity with maximum thickness of 16 mL and left frontoparietal, occipital convexity with maximum thickness of 3 mL (Fig. 3). The lesion showed areas of blooming. Since MRI of brain did not reveal any site of leakage, MRI of whole spine with CSF sequence was done, which revealed hyperintense area at the level T7 (thoracic) vertebra suggestive of CSF leak (Fig. 4). After localization of site of CSF leak, epidural

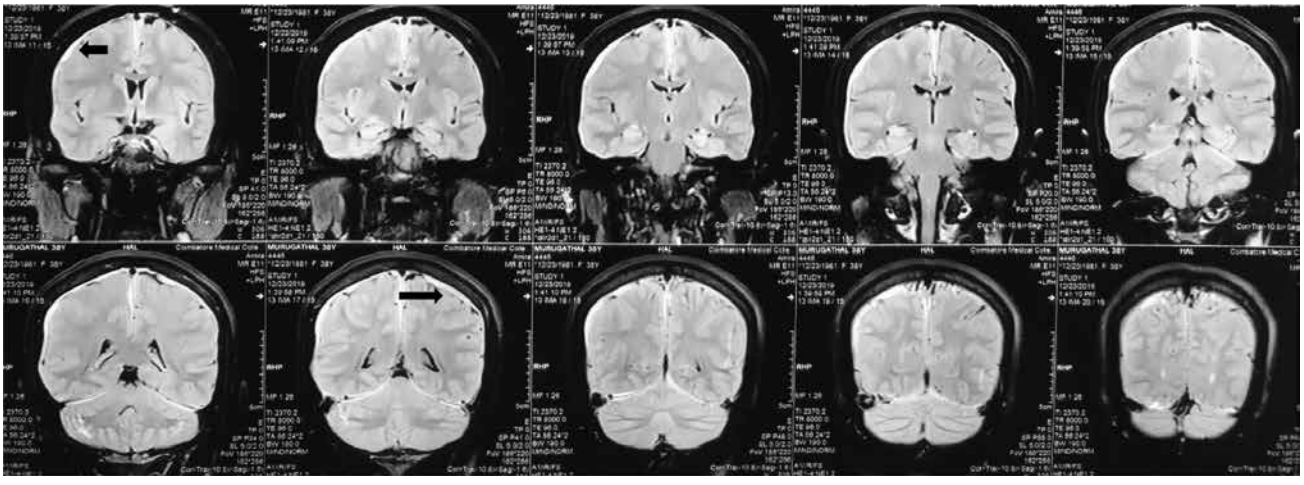

Figure 1. T2 Flair coronal section of MRI of brain - Bilateral subdural hygroma.

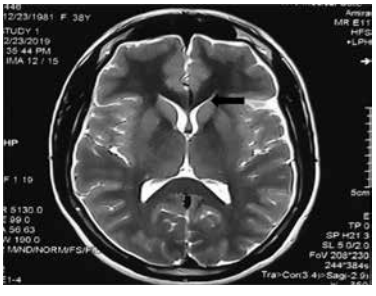

Figure 2. T1 Axial section of MRI of brain – Slit-like frontal horn of left lateral ventricle.

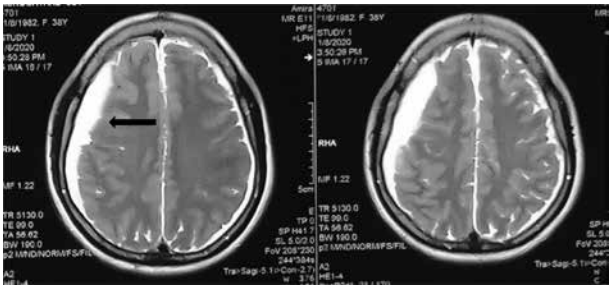

Figure 3. T2 Axial section MRI of brain - Increase in volume of subdural collection in right frontoparietal convexity and left frontoparietal, occipital convexity (follow-up scan).

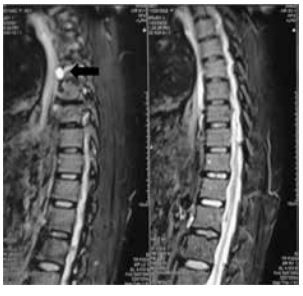

Figure 4. MRI of spine with CSF flow sequence showing leak at T7 vertebral level.

blood patch with patient's own blood was put. After the procedure, her headache reduced tremendously within 2 days.

DISCUSSION

There are various causes of intracranial hypotension. The CSF leak can be caused by lumbar puncture (CSF trickles into the paravertebral muscles), spinal surgery and spinal trauma (tear in the arachnoid surrounding a nerve root). Spontaneous intracranial hypotension can be caused due to low intracranial pressure by leakage of CSF with unknown cause. It is a rare disease occurring in 1 in 50,000 people. It is more common in women than men. The most common cause of spontaneous intracranial hypotension is the leak of CSF through a tear in spinal dura. Marfan and Ehlers-Danlos syndrome, autosomal dominant polycystic kidney disease are genetic risk factors for spontaneous CSF leak.

Orthostatic headache is cardinal presentation of spontaneous intracranial hypotension. It may be associated with diplopia (due to 6th cranial nerve palsy or self-audible bruit from turbulence in the intracranial venous system). There can be sagging of the frontal lobes in low CSF pressure caused by leakage of the CSF (brain sagging syndrome), which can lead to brainstem lesions with stupor, gaze palsies and cranial nerve palsies. Patients are apathetic and disinhibited with prominent daytime somnolence.

To localize the site of the leak, radionuclide cisternography or CT (computed tomography) myelography can be done. CT myelography is the preferred diagnostic test. Dynamic CT myelography is useful for detecting high flow leaks. As there are technological advancements and availability of MRI, MRI of brain with gadolinium contrast is done, which shows prominent dural enhancement (due to dural venous dilation) or diffuse pachymeningeal enhancement described by Fishman and Dillon et al. Other additional features seen are subdural effusions on cerebral convexities, temporal lobes, optic chiasma or cerebellar tonsils. In order to find leak, MRI of spine can be done which may show spinal fluid collection, dural enhancement, dilated epidural veins, enlarged epidural venous plexus, attenuation of spinal canal or compression of spinal cord and contrast

extravasation. MRI of spine helps us find the site of leak present in the spinal dura.

A diagnostic criteria for headache in spontaneous intracranial hypotension was framed by Schievink et al as:

- Orthostatic headache
- The presence of at least one of the following: low opening pressure (≤60 mm water), sustained improvement of symptoms following epidural blood patching, demonstration of an active spinal CSF leak, cranial MRI changes suggestive of intracranial hypotension (brain sagging or pachymeningeal enhancement)
- No recent history of dural puncture
- Not attributed to another disorder.

CONCLUSION

Our patient had orthostatic headache, with relief of headache after blood patch and MRI of brain showing dural enhancement.

The definitive treatment is epidural blood patch with approximately 20 mL of patient's blood at the site of leak, which relieves the headache. After blood patch, there has been no recurrence; very few cases had repeated episodes of orthostatic headache.

As there is increase in availability of MRI, spontaneous intracranial hypotension can be detected easily. It can be treated easily, preventing long-term morbidity as well as preventing the inappropriate usage of analgesics.

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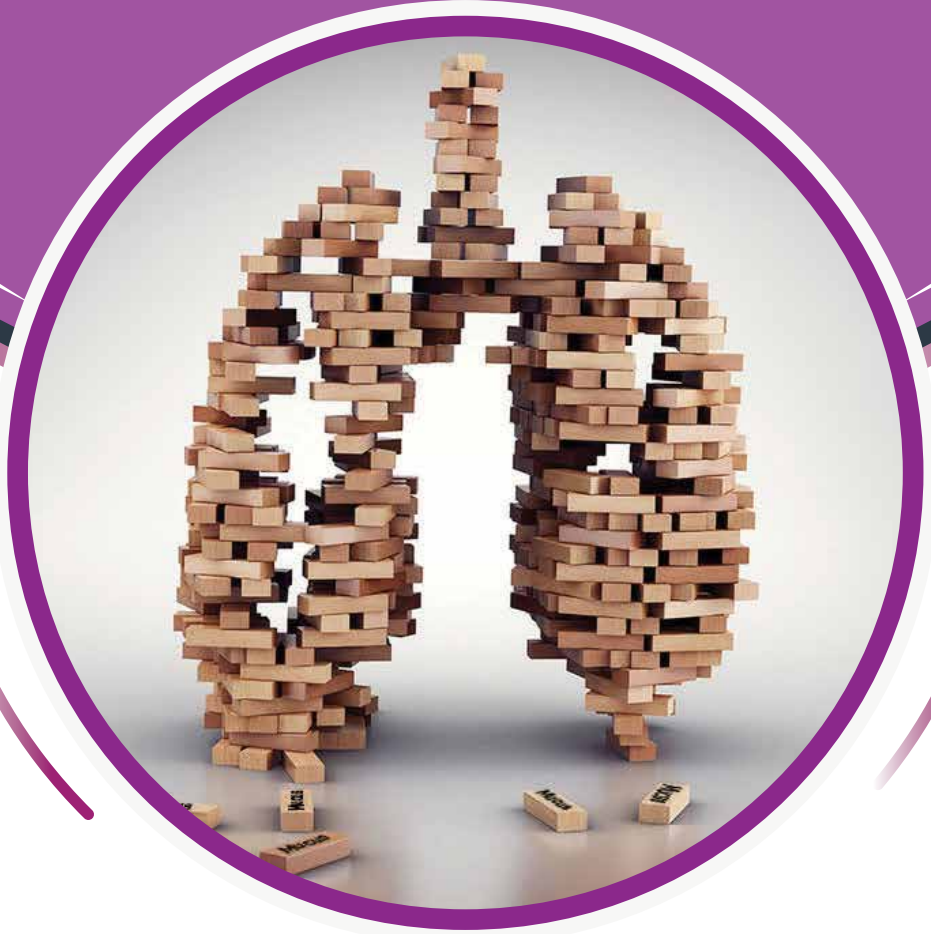

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