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SPONTANEOUS INTRACRANIAL HYPOTENSION; THREE CASE REPORTS WITH SIMILAR CLINICAL MANIFESTATIONS, TREATED SUCCESSFULLY USING DIFFERENT MANAGEMENT TECHNIQUES.

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ABSRACT

Introduction: Spontaneous Intracranial hypotension (SIH) is a rare neurological disorder, characterized by orthostatic headaches. Due to the complicity of its diagnosis and lack of awareness amongst physicians, SIH remains an under-diagnosed disease and its true prevalence remains unknown. It is a reversible condition, if diagnosed early. Delay in diagnosis can result in life threatening complications.**Case Reports:**We present a case series of three patients who presented with typical symptoms of SIH. But management course of each patient varied. The first patient responded well to the EBP (epidural blood patch) while the second improved with conventional symptomatic treatment. The third patient needed a surgical intervention for complication developed due to SIH. This case series hence covers a variety of treatment options for patients with SIH.**Conclusion:**SIH is an emerging challenge for neurologists worldwide. Awareness amongst physicians regarding this disease along with a high level of suspicion and good history skills will allow early diagnosis of the disease and prevent delay in treatment and hence complications.

Introduction:

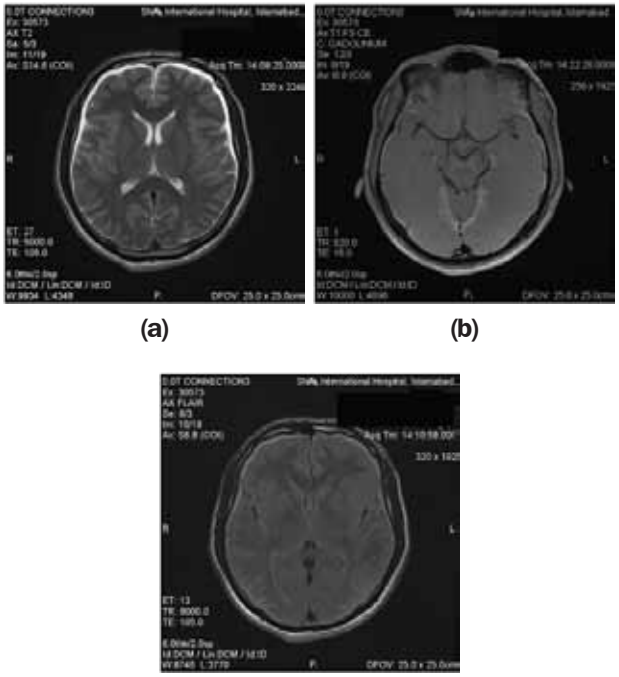
Spontaneous Intracranial hypotension, SIH, is a diagnostic and management dilemma for the physicians today. There are different methods being used by the neurologists to treat SIH, but complications and different response of individuals to treatment makes it challenging. We present three cases of spontaneous intracranial hypotension, with almost similar clinical presentation, treated successfully using different approaches.

Case 1:

A 40 years old male, noticed orthostatic headache 4 weeks before presenting to Neurology clinic at Shifa International Hospital. The headache was intermittent, moderate in intensity, 7/10 on pain scale, gradual in onset and progressively worsening. It was exacerbated by standing and walking and relieved on lying flat. He tried painkillers but were noteffective. For last 2 weeks he had also experienced intermittent blurring of vision on standing. He had no episodes of similar headaches in the past. There was no history of trauma or any procedure including spinal anesthesia or lumbar puncture done. He had no complains of nausea, vomiting, focal weakness, neck stiffness, dizziness or loss of consciousness. His systemic and neurological examinations were unremarkable. He had no motor weakness or any sensory deficit, all cranial nerves were intact and gait was normal. Eye examination and fundoscopy were normal. MRI brain with and without contrast was done, which showed bilateral fronto parietal subdural hygroma, droopy penis sign i.e. downwards droppingof splenium of corpus collosum and pachymeningeal enhancement on contrast, as shown in Figure 1 below. These findings are consistent with spontaneous intracranial hypotension.

The patient was initially advised bed rest, plenty of fluids and painkillers, but he didn't improve with conventional treatment and pain continued to interfere with his daily work routine. Hence other options were discussed with the patient and after consent, epidural blood patch (EBP) was performed. EBP was done as a day-care procedure. During the procedure, 30 ml of fluid (blood) was inserted in the epidural space. Post procedure, patient was observed for six hours and then

discharged home. On one week follow up, his headache had resolved. Since then, he has been pain free and doing well.



(a) (b)

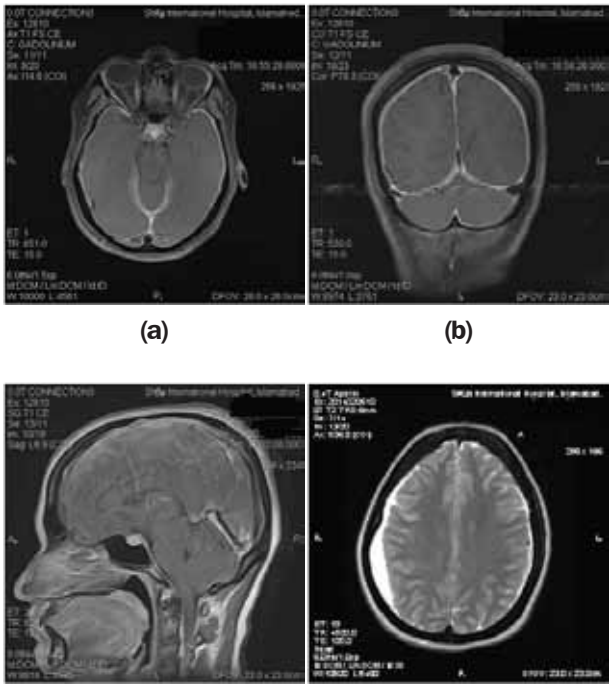
(c)

Figure 1 shows MRI Brain images; a) Axial T2 b) Axial T1 & c) FLAIR. The images above shows bilateral fronto- parietal subdural abnormal signal with intense post contrast enhancement and enhancing meninges, all of which are consistent with intracranial hypotension.

Case 2:

A 33 years old female, with no known comorbidities, presented to neurology clinic with headache for past 20 days. It was an early morning, throbbing headache, which started off from neck and radiated to the occipito-temporal region. It was worse on standing and sitting and partially relieved on lying flat. The pain was 6/10 on pain scale. She had no complaints of nausea, vomiting, photophobia, focal weakness, seizures, fever or weight loss. There was no history of trauma or similar headaches. Her family history was negative for malignancies. She was married, hadfour children, all born via spontaneous vaginal delivery (SVD), without any spinal or epidural anesthesia. Her systemic and neurological examination was normal. She had normal gait with a good power in all four limbs and no sensory deficit. Planters were bilaterally down going, eye examination was normal and fundoscopy was unremarkable.

MRI brain, with and without contrast, was done which showed bilateral fronto-parieto-temporal subdural abnormal signals with intense post contrast leptomeningeal enhancement, associated with mass effect and impending trans-tentorial and tonsillar herniation. Figure 2 below shows her MRI findings.



(a) (b)

(c) (d)

Figure 2: it shows MRI, with and without contrast, of a patient with intracranial hypotension. Figure (a) shows T1 axial, (b) T1 coronal and (c) T1 sagittal view. Figure 2 (d), shows Flair image. T1 images above, show post contrast enhancement of lepto-meninges, extending into sulci over vertex. Severe mass effect with increase intracranial pressure causing small ventricle compression can also be seen. Lateral, 3rd and 4th ventricles are relatively smaller in size. There is subtle prominent abnormal lepto-meningeal enhancement extending into sulci over vertex which is occupied by bright signal on FLAIR image. Tonsils are low lying. Development of subdural hemorrhages in parietal regions bilaterally, larger on right, can also be seen in images above. Based on clinical symptoms and MRI findings, she was diagnosed with spontaneous intracranial hypotension and treated with conventional therapy. She was advised bed rest, plenty of fluids and painkillers (Acetaminophen two tablets thrice daily for two weeks). Two weeks later, on follow up, her symptoms had improved and she had resumed daily life activity. After six months, follow up MRI brain was done which showed subtle interval decrease in mass effect and leptomeningeal enhancement. Currently, she is symptom free and doing well.

Case 3:

A 45 years old scientist, known case of migraine for more than a decade, well controlled on medications, presented to Neurology outpatient department with complaints of "a different type of headache", worse on standing and better on sitting, for the past 1 year. He had no other associated symptoms. He had visited several doctors during this time but was never diagnosed. The intensity of his headaches had progressively worsened from 5/10 initially to 8/10 currently on pain scale. He was advised MRI brain by the last doctor he had visited. He presented to us with his MRI reports. On examination he looked very distressed and anxious due to the headache. He had no focal weakness, sensations were intact and rest of the neurological examination was also unremarkable. His general physical and eye examination was normal. MRI brain showed downward drooping splenium, upward elevation of tentorium, low lying cerebellar tonsils and persistent extensive mucosal thickening. The MRI brain along with his history, was suggestive ofSIH.

He was started initially on conventional therapy and advised bed rest, hydration, caffeine and painkillers. He was advised to return if pain did not settle.

A week later, he presented to emergency department at Shifa International Hospital, with severe, unbearable headache. On examination, he had no focal deficit, neurological examination was normal. He was however very anxious and in distress due to the pain. A CT scan head was done immediately, which showed right sided large subdural hematoma, as shown in Figure 3 below.

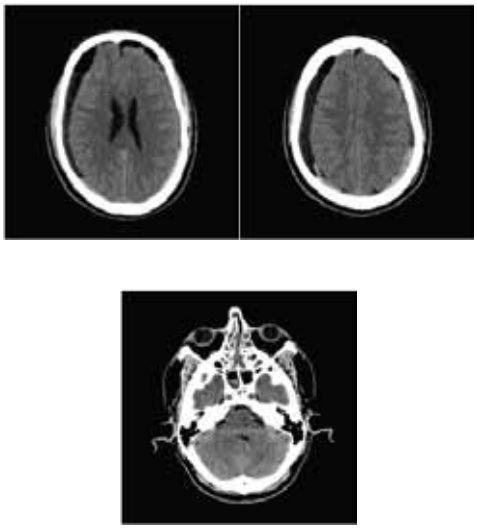


Figure 3: CT scan head showing right large subural hematoma

After a discussion with the patient and his family, neurosurgery was planned and a bur hole was done to drain the hematoma. There were no per or post-operative complications. The headache resolved after surgery and the patient was discharged home within a week. Two days post discharge, he again presented to emergency department with severe headache and monoparesis (right-sided weakness). His examination showed decrease power on right side, 4/5 in both right upper and lower limbs with up going right plantar, the rest of the neurological examination was unremarkable. All cranial nerves were intact. A CT scan head (as shown in figure 4 below) was done, which showed an intra cranial bleed on left mid parietal, the place corresponding to the site of craniotomy.

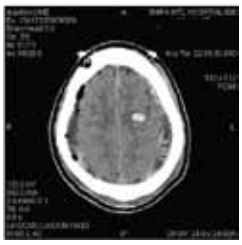


Figure 4 shows a CT scan brain. An ICB in the left parietal lobe can be seen along the burr hole craniotomy tract. No mid line shift or tonsillar herniation seen. Post-surgical air can be seen in the bilateral subdural hematoma.

Figure 4

He was admitted and treated as a stroke. Stroke pathway was activated and he was started on aggressive physiotherapy. A few days later his headache resolved and he was discharged on rehabilitation. Within one month's time, his power of all 4 limbs was 5/5. Currently he is doing well, has no headache or any residual weakness.

Discussion:

Spontaneous intracranial hypotension (SIH) is a difficult diagnosis due to the variable and non-specific manifestation of the disease. It is a benign neurological condition causing postural headaches resulting from low cerebral spinal fluid (CSF) pressure. It was first described by Georg Schanltenbrand, a German neurologist. [1] A revised diagnostic criteria for SIH, as shown in table 1 below, was given in 2011 which made diagnosis easier and simpler[2]. It is the same diagnostic tool used to diagnose patients in our case report.

	Diagnostic Criteria for SIH
1	Orthostatic headache i.e. worst on standing and relieved on lying flat
2	Any one of the following;
	a) low CSF pressure
	b) symptoms improve on epidural blood patch (EBP)
	c) active spinal CSF leak demonstrated
	d) MRI shows changes corresponding to spontaneous intracranial hypotension.
3	No recent history of dual puncture
4.	Symptoms not explained by any other diagnosis

Table 1: Shows the revised diagnostic criteria for SIH, first given in 2011

SIH is twice more common in females as compared to males and more likely to peak at the age of 40 years[3] .The mechanism of the disease is well explained by Monro- kellie doctrine. The CSF leak results in low CSF pressure and hence volume. This leads to a compensatory increase in blood volume causing dilation of veins in brain, spine and meninges. This dural vasodilation and engorgement results in dural enhancement. Such changes can lead to structural abnormalities like mass effect on thecal sac, disc herniation and nerve root compression . All the changes described above can be seen on MRI brain of patients with SIH, hence facilitating the diagnosis. There are different treatment approaches in patients with spontaneous intracranial hypotension. The most conventional approach is still considered as the first line treatment option, is non-invasive and focuses on symptomatic relief. Patients are usually advised bed rest and prescribed painkillers for headache. Also, they are advised Trendelenburg positioning and increase caffeine intake, both of which help increase CSF pressure and hence relieve headache. EBP (Epidural Blood patch) has recently gained international recognition and now being considered as first line treatment. Recent gain in popularity of EBP in treatment for SIH is due to its high success rate of around 90% and a low recurrence. In EBP, a certain volume of blood is injected into the CSFspace, creating an acute compression of the thecal sac and hence increase in CSF pressure, resulting in an immediate relief of pain. It also causes long term relief by formation of dual tamponade which helps seal the CSF leak and maintain the CSF pressure. Most EBP's are placed in the lumbar spine. Thoracic and cervical blood patches are associated with higher risks of cord and nerve compression, neck stiffness, chemical meningitis and cord puncture. SIH, if not treated in time, can lead to various complications. Some of these can be life threatening. Subdural hematoma is one of the well- known complication. The high pressure in cortical veins due to intracranial hypotension causes them to rupture resulting in small subdural collections. These small collections can give rise to a large subdural hemorrhage. This can cause deterioration in the condition of patient, drowsiness and drop in GCS. MRI can show mass effect and midline shift.

Large subdural hematomas need to be surgically evacuated. In a few cases, subdural collections can result in normalization of the low intracranial pressure hence associated with disappearance of the headache[1°]. Other well recognized complications of SIH include cerebral venous thrombosis (CVT), cerebral displacement and dural enhancement.

Conclusion:

SIH is an emerging challenge for neurologist worldwide. It is an under diagnosed disease. If diagnosed early, SIH is treatable and has an excellent prognosis. A delay in diagnosis can subject a patient to a lot of unnecessary and painful procedures and complications.

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Author's contribution:

Dr. Ali Ilyas : Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review
Dr. Wasim Tariq: Study concept and desig, data analysis, manuscript writing, manuscript review
Dr. Anam Abrar: data collection, data analysis, manuscript writing, manuscript review
Dr. Muhammad Amjad: Study concept and design, manuscript writing, manuscript review
Muhammad Azhar Saeed: Study concept and design, data analysis, manuscript writing, manuscript review
Inayatullah Khan: Study concept and design, data collection, data analysis, manuscript writing, manuscript review