International Journal of Medical Science in Clinical Research and Review
Online ISSN: 2581-8945
Available Online at https://ijmscrr.in/ Volume 7|Issue 06 (November-December) |2024 Page: 1132-1138

Case Report

Primary Intracranial hypotension and MRI – Case Reports

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Article Received: 20- September -2024, Revised: 10-October-2024, Accepted: 30-October-2024

ABSTRACT:
Intracranial hypotension (IH) is an uncommon, benign and self-limiting condition caused by low cerebrospinal fluid pressure and presents with orthostatic headache as chief complaint but these symptoms are not specific to intracranial hypotension and therefore misdiagnosis is common. The most prominent feature of intracranial hypotension on MRI is "brain sagging". Further evaluation in most cases reveal CSF leakage through a dural defect with no identifiable cause resulting in low CSF pressure and volume. With the use of different diagnostic measurements and angles, the MRI detection of Intracranial hypotension has improved.

Keywords: Intracranial hypotension, Magnetic resonance imaging, cerebrospinal fluid pressure

INTRODUCTION:

Intracranial hypotension is of primary or secondary origin. Secondary intracranial hypotension is related to previous injury which may be iatrogenic like secondary to lumbar puncture or surgery along the neuraxis. Primary intracranial hypotension can occur as a result of trivial trauma and weakness of the dura resulting in the dural tears causing CSF leaks [3], [4]. In many cases of primary intracranial hypotension, the site of CSF leak is occult. SIH is estimated to affect 5 per 100,000 people per year, with predominance for women [5].

Patients with primary intracranial hypotension present with orthostatic headache as chief complaint which worsens in upright position, coughing and Valsalva manoeuvre. Other clinical features include hearing impairment, dizziness, vomiting, diplopia, visual defects, backache and neck pain. Women are more commonly affected than men in the third to fifth decade [3].

With the use of different diagnostic measurements and angles, magnetic resonance imaging plays an important role in accurate diagnosis of intracranial hypotension and also plays an important role in follow-up of these patients [2]. The specific MRI features include sagging of the brain, diffuse meningeal enhancement, subdural fluid collections and reversible pituitary enlargement[3], [5].

We describe two patients with spontaneous intracranial hypotension examined with MRI Brain studied retrospectively after receiving permission from concerned authorities (Head of the Department of Radiodiagnosis and Hospital Director) of MGM Hospital, Navi Mumbai and review their characteristic imaging features.

CASE1:
A 45-year-old female patient came with complaints of headache, dizziness and vomiting.

Figure-1:

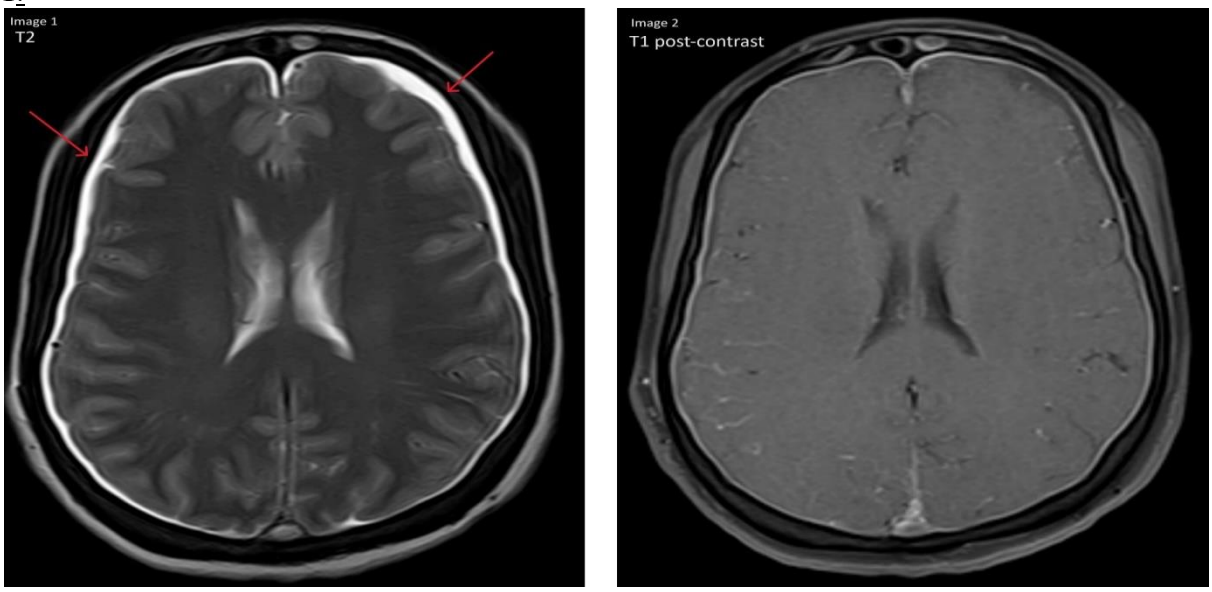
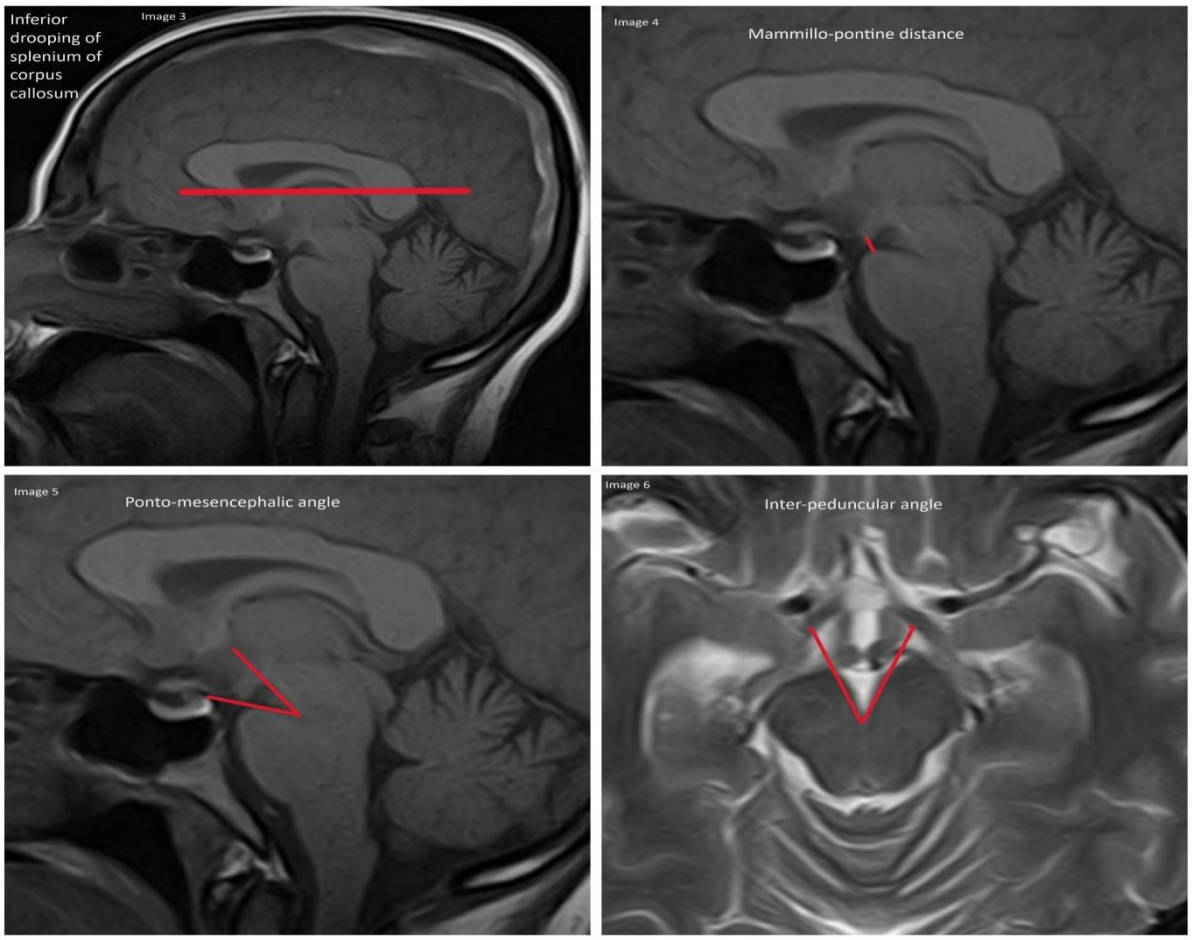


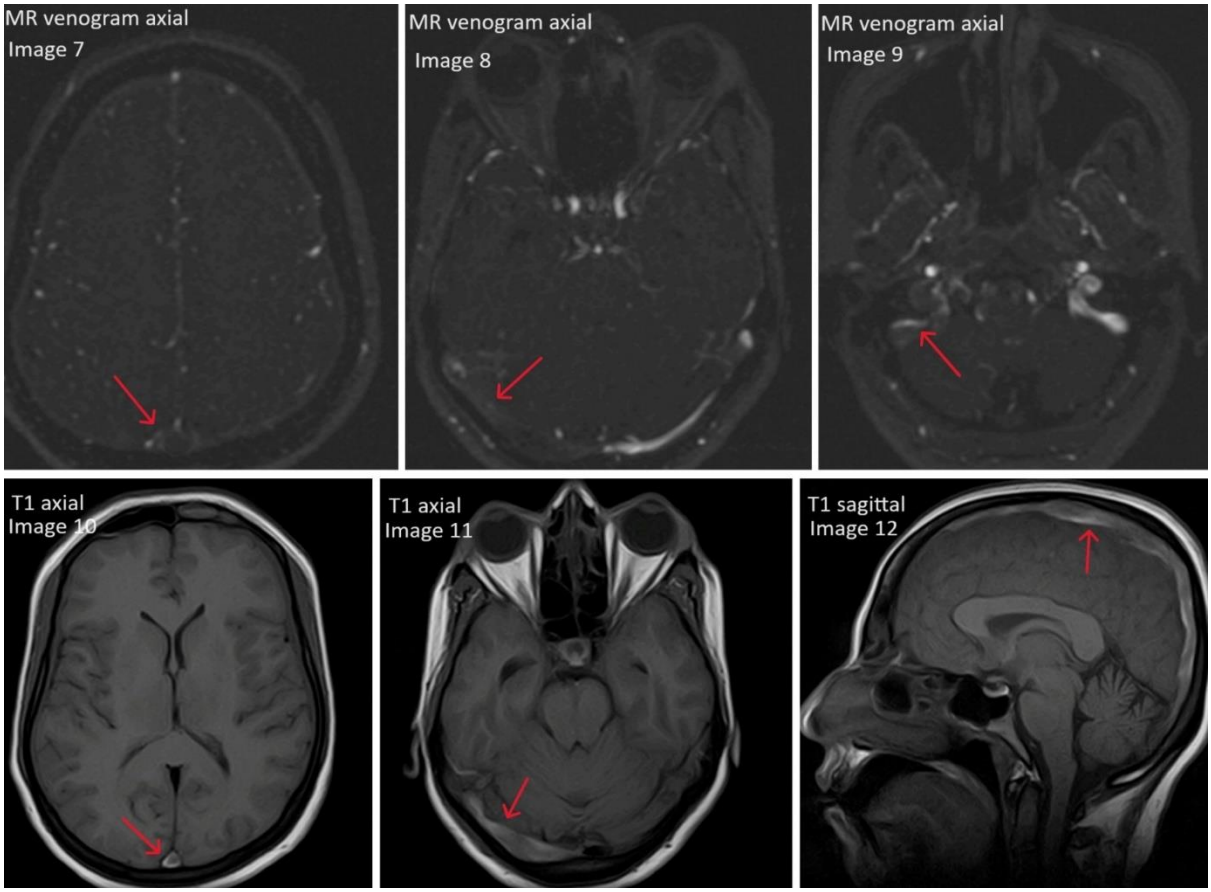
Image 1 – T2 axial image shows subdural effusions along the bilateral cerebral hemispheres.
Image 2 – T1 post contrast axial image shows diffuse pachymeningeal thickening and enhancement involving the bilateral cerebral hemispheres and along the inter-hemispheric fissure

Figure-2:



There was inferior drooping of splenium of corpus callosum (Image 3) and sagging of midbrain demonstrated by reduced mamillo-pontine distance measuring 3.3mm (Image 4) and reduced ponto-mesencephalic angle measuring 38°(Image 5) and interpeduncular angle measuring 38° (image 6).

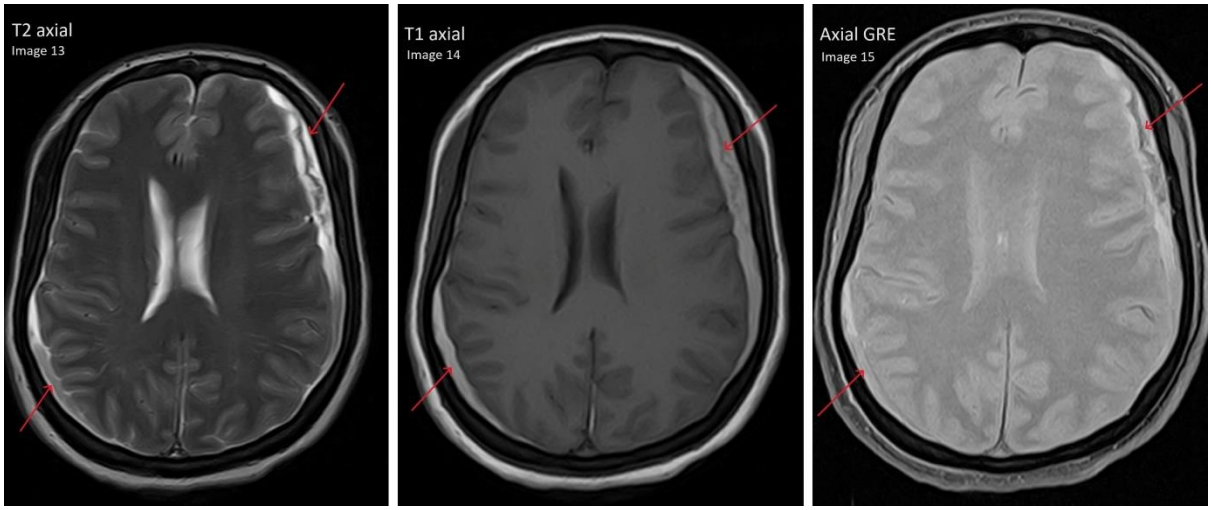
Figure 3:



There was distention of superior sagittal sinus (image 10 and 12) and right transverse sinus (image 11) with T1 hyper intense signal within.
MR venogram shows loss of flow related signal intensity with filling defects involving the superior sagittal (image 7), right transverse (image 8), right sigmoid sinuses (image 9) and proximal portions of right internal jugular vein suggestive of dural venous sinus thrombosis. There was also involvement of cortical veins of bilateral high parietal lobes.

After 10-15 days the patient got worsened and presented with altered sensorium. The MRI brain was repeated.

Figure 4:



There were large extra-axial collections along the left fronto-parieto-temporo-occipital regions and right parieto-occipital regions appearing hyperintense with hypo-intense areas within on T2W images (image 13) and hyperintense with iso-intense areas within on T1W images (image 14) with patchy areas of blooming within on GRE (image 15) suggestive of acute on late subacute subdural haemorrhages.

The patient was followed up with CT Myelogram (done outside) which revealed leak of contrast opacified CSF along multiple lumbar neural foramina on left side from L1-L2 to L4-L5 levels maximum at L3-L4 level with the extravasated contrast reaching up to the medial edge of left psoas major muscle.
Patient was treated with epidural blood patch at the site of CSF leakage.

CASE 2:
A 39-year-old female patient with complaints of headache and diplopia.

Figure-5:

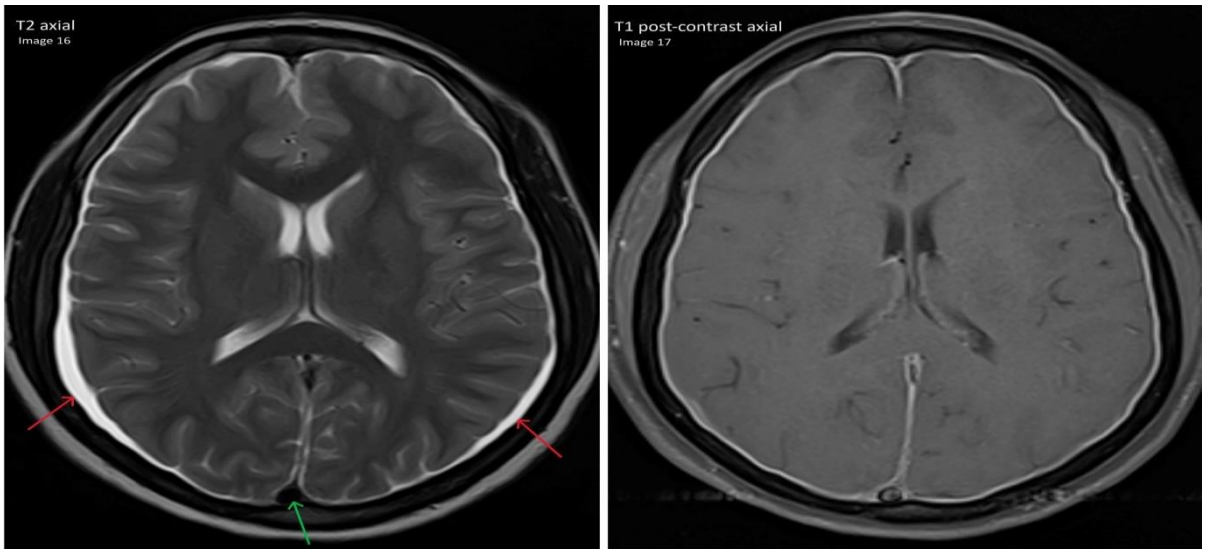
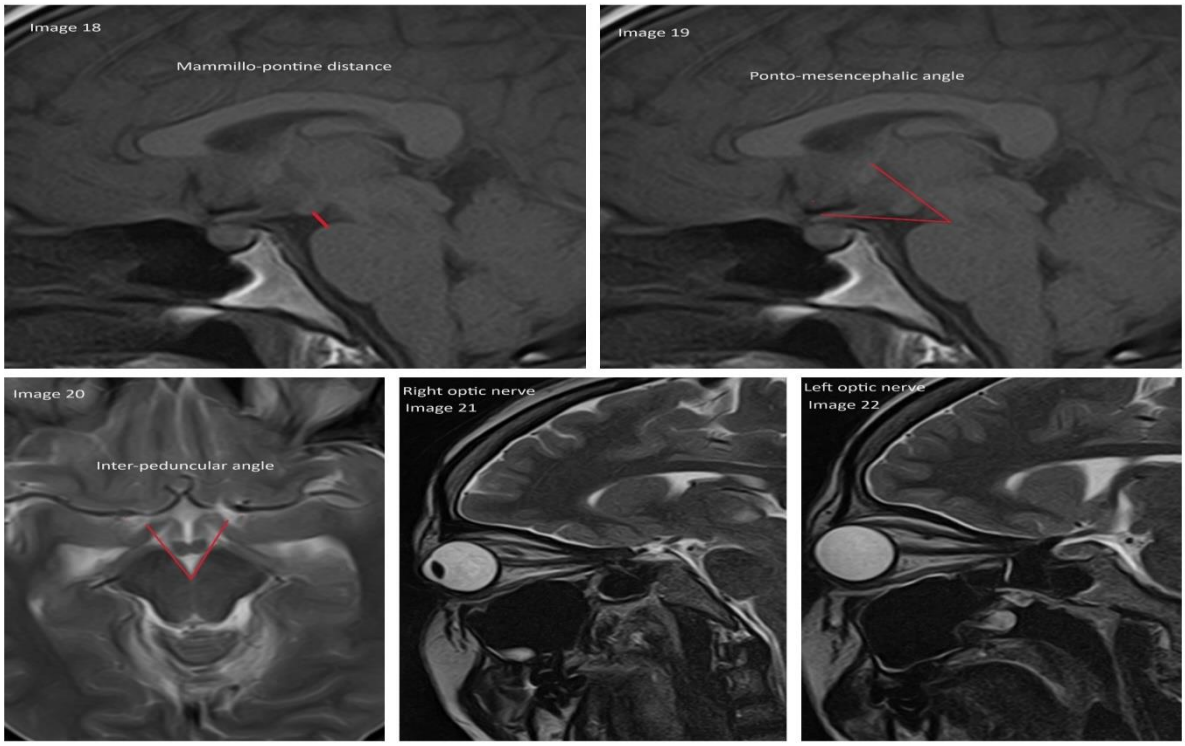


Image 16 – T2 axial image- red arrows show subdural effusions in the bilateral parietal regions.
Green arrow shows distention of superior sagittal sinus with loss of its normal trifoliate shape.
Image 17 – T1 post contrast axial image shows diffuse pachymeningeal thickening and enhancement involving the bilateral cerebral hemispheres and along the inter-hemispheric fissure.

Figure 6:



There was sagging of midbrain demonstrated by reduced mamillo-pontine distance measuring 4.2mm (Image 18) and reduced Ponto-mesencephalic angle measuring 40°(Image 19). However, the interpeduncular angle was normal measuring 50° (image 20).
T2 sagittal images of right orbit (image 21) and left orbit (image 22) show reduction in peri-optic CSF spaces.

DISCUSSION:

In patients with suspected spontaneous intracranial hypotension, MRI with and without gadolinium contrast remains an initial imaging examination of choice. The most common qualitative finding is diffuse pachymeningeal thickening and enhancement followed by features of increased venous blood volume presenting as engorgement and distention of dural venous sinuses with loss of normal trifoliate shape on the cross-sectional imaging and prominence of the inferior inter-cavernous sinus. Intracranial venous thrombosis is a rare but well recognized complication and may involve dural venous sinuses and cortical veins.

MR findings also include subdural effusions and eventual subdural hematomas. Reduced CSF volume results in reversible enlargement of pituitary gland due to engorgement of pituitary vasculature [1], inferior drooping of splenium of corpus callosum, reduced CSF within the optic nerve sheath and may result into sagging of midbrain, pons and acquired tonsillar ectopia.

Few quantitative signs have been described to evaluate the sagging of midbrain and pons which includes;[2]

Mamillo-pontine distance <6.5mm - measured on the mid-sagittal T1W image between the inferior margin of mammillary bodies and superior surface of the pons.

Ponto-mesencephalic angle <50° - measured on the mid-sagittal T1W image and it is the angle between two lines drawn along anterior margin of the midbrain and antero-superior margin of the pons.

Interpeduncular angle <40.5° - it is the angle formed by posterior half of the cerebral peduncles measured on an axial T2W image at the level of mammillary bodies.

The common cause of spontaneous intracranial hypotension is a CSF leak which is more common at spinal level than skull base [10], [11]. In majority of patients CSF leak is occult or no epidural CSF leak is detected despite extensive imaging and follow-up. In some cases, follow up with CT myelogram demonstrates a CSF leak site from a dural defect, a CSF-venous fistula [3], [4] or a spinal meningeal diverticula [12]. If the site of CSF leak is identified, it can be sealed with the help of a targeted epidural blood or fibrin glue patches [3], [4], [6], [7].

Spontaneous intracranial hypotension is a diagnosis of exclusion and can mimic various other conditions [5]. Sub-dural effusions should be differentiated from subdural hematoma resulting from rupture of bridging cortical veins, subdural hygroma. Diffuse meningeal thickening and enhancement can also be present in conditions like hypertrophic pachymeningitis due to IgG4 related diseases, neuro-sarcoidosis, rheumatoid arthritis, temporal arteritis, polyangitis with granulomatosis and meningeal tuberculosis [17], [18], [19]. Acquired tonsillar ectopia secondary to spontaneous intracranial hypotension should be differentiated from Chiari malformation [13], [14], [15], [16]. Clinical mimics of spontaneous intracranial hypotension include orthostatic hypotension, vestibular migraine and cervicogenic headache [5], [8], [9]. The reversible enlargement of pituitary gland should be differentiated from small non-functioning pituitary adenomas and pituitary apoplexy [23], [24], [25]. In patients presenting with cerebral venous sinus thrombosis should be assessed for possible underlying spontaneous intracranial hypotension [5], [20], [21], [22]. CSF venous fistulas also show a good clinical response to the targeted fibrin glue injections [6], [7].

CONCLUSION:

Spontaneous intracranial hypotension is an exotic diagnosis of exclusion, based on the clinical suspicion, history, physical examination and MRI results. It is often mis-diagnosed due to the wide range of clinical mimics and secondary changes from spontaneous intracranial hypotension can give rise to symptoms that imitate other conditions. Qualitative MRI findings are sometimes equivocal and can be absent in some cases. Therefore, quantitative assessments help in more accurate diagnosis of spontaneous intracranial hypotension which can be further investigated with the help of CT Myelogram or Cisternogram to look for the cause and exact site of CSF leak and can guide clinicians to plan for appropriate treatment.

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