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CE - MEDICAL ILLUSTRATION

Intracranial hypotension: diagnosis by trial of Trendelenburg positioning and imaging

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An 80-year-old woman with well-controlled hypertension, hypothyroidism, and an atraumatic fall 3 weeks prior, presented with complaints of altered mental status and left eye drooping for 1 day, without headache. On initial examination, she was noted to be lethargic with a partial left third nerve palsy, with preserved but sluggish pupillary light reflex. A CT head scan was done, revealing bilateral acute on chronic subdural hematomas with an admixture of hypodense material in the subdural space and some hyperdense components (Fig. 1). Given her third nerve palsy, there was concern for pathology other than the subdural hematoma. An MRI brain scan with and without contrast was done, and revealed diffuse pachymeningeal enhancement, in addition to bilateral frontoparietal subdural collections. (Fig. 2d–f) The collections were heterogeneous and thought to represent a mixture of proteinaceous CSF material and acute blood (Fig. 2d–f). A lumbar puncture was performed, with an opening pressure of 9 cmH_2O. CSF studies showed: glucose 64 mg/dL, protein 129 mg/dL, 4444 red blood cells, 167 white blood cells (24 % neutrophils, 76 % lymphocytes), and a negative Gram's stain. The pachymeningeal enhancement, new cranial nerve palsy, and elevated CSF white blood cell count were thought to be consistent with chronic subdural hematoma versus lymphoma, inflammatory process, or other malignant process. While CSF cultures, cytology and flow cytometry were pending, the patient's course improved, and she was able to maintain upright posture and feed herself. However, within a day of this improvement, she was again noted to have fluctuating mental status, ranging from baseline intact mental status to lethargic and obtunded. She also developed a new right third nerve palsy, which intermittently resolved and recurred spontaneously. EEG monitoring was done and revealed only diffuse slowing with no epileptiform activity. In addition, she developed episodes of hypertension with bradycardia and irregular breathing, which are the symptoms of Cushing's triad, a syndrome associated with increased intracranial pressure.

Given a fluctuating course and concern for increased intracranial pressure with the signs of Cushing's triad, imaging was reviewed again. On second review, further abnormalities were noted, including engorged cortical veins and venous sinuses and signs of brain sagging. Brain sagging describes the downward displacement of the brain that was demonstrated in our patient with: ventricles smaller than expected for age, increased anterior–posterior and decreased bipeduncular diameter of the mesencephalon, an effaced interpeduncular cistern, enlarged pituitary gland, and a sagging corpus callosum with a down-pointing splenium (Figs. 1, 2) These imaging findings raised concern for spontaneous intracranial hypotension (SIH). The patient was subsequently placed in Trendelenburg position, and her mental status improved dramatically. She did, however, have a persistent partial left third nerve palsy. Although this was thought to support the diagnosis of SIH, there was some concern that the clinical improvement may have been coincidental, as her mental status had been fluctuating throughout her hospital course. A CT myelogram was done, revealing CSF extravasations at T11/T12, supporting the diagnosis of SIH (Fig. 3). As the patient's examination continued to fluctuate with position, a lumbar epidural blood patch was performed. The patient showed significant

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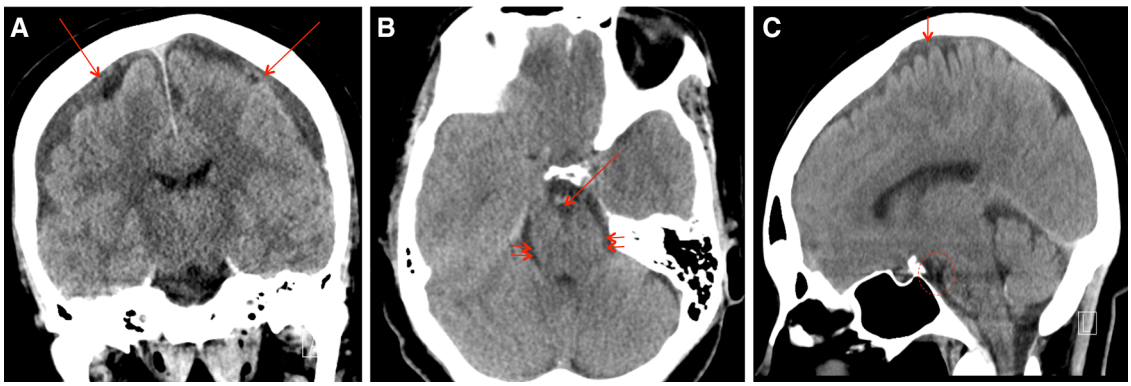

Fig. 1 CT brain without contrast. **a** Coronal view shows bilateral subdural, heterogeneous collections. **b** Axial view shows small interpeduncular (*arrow*) and ambient (*double arrow*) cisterns. **c** Sagittal view again shows subdural collection (*arrow*) and effaced interpeduncular cistern (*dotted circle*)

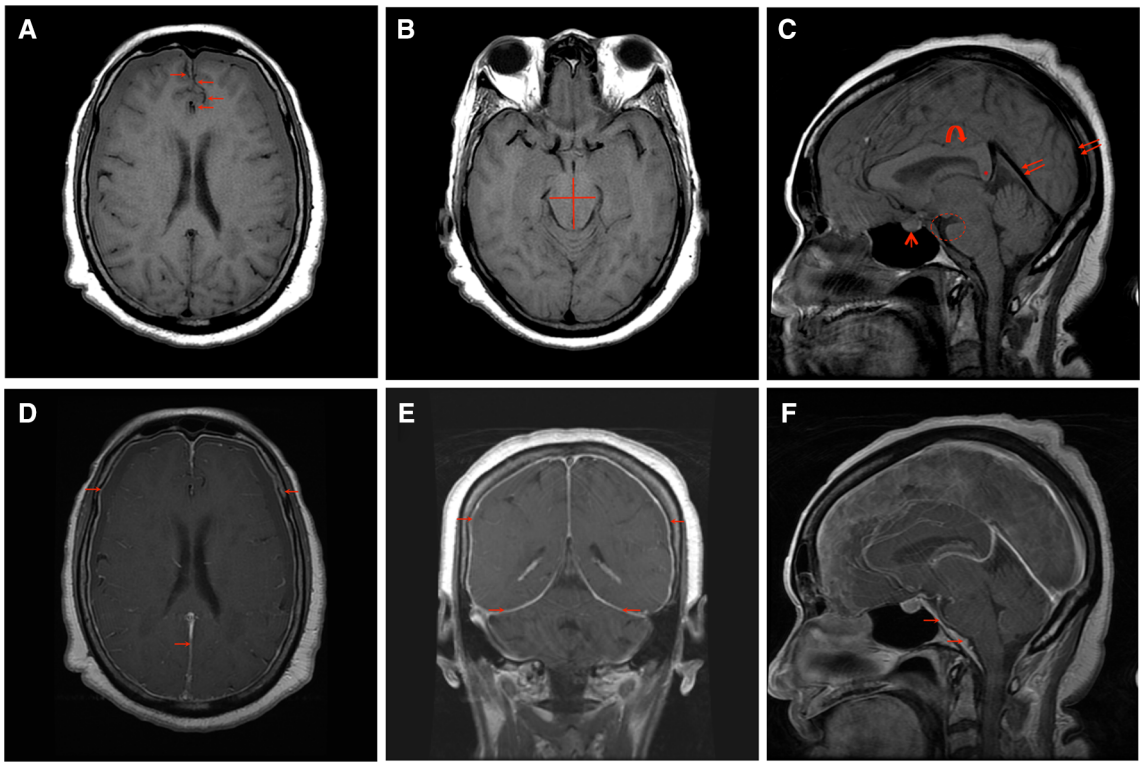

Fig. 2 MRI brain with and without contrast. **a** T1 axial view shows subdural collections and engorged cortical veins (*arrows*). **b** T1 axial view shows increased anterior–posterior and decreased bipeduncular diameter of the mesencephalon (*cross*). **c** T1 sagittal view shows engorged venous sinuses (*double arrows*), effaced interpeduncular cistern (*dotted circle*), enlarged pituitary gland (*single straight arrow*), and sagging corpus callosum (*curved arrow*) with down-pointing splenium (*asterisk*). Post contrast images show diffuse pachymeningeal enhancement (*arrows*) in axial (**d**), coronal (**e**), and sagittal (**f**) views

improvement with stable, baseline mental status, resolution of the right third nerve palsy, resolution of the Cushing's triad, and return of ability to ambulate following the procedure. She was transferred to a rehabilitation facility, and after a short stay, was discharged home with persistent, but improving left partial third nerve palsy and no other neurological deficits. Her CSF cultures, cytology, and flow cytometry all were negative, excluding alternative diagnoses. It was thought that her initial injury may have been her atraumatic fall, causing dural tearing at T11/T12 and leading to her SIH.

Our patient's symptoms of altered mental status, cranial nerve palsy and pachymeningeal enhancements were initially attributed to chronic subdural hematoma, as prior cases with this entity have been thought to be causative. There was also some concern for lymphoma or other

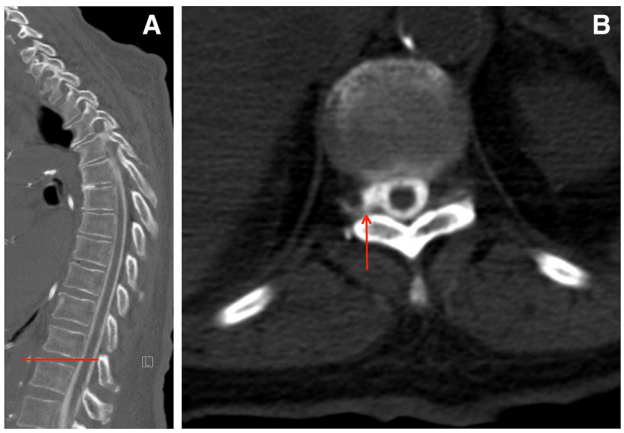

Fig. 3 CT myelogram. **a** Sagittal view shows level (*line*) of axial cut. **b** Axial view showing CSF extravasations (*arrow*)

malignant processes, which have also shown similar MRI and clinical findings. However, our patient's imaging was concerning for downward displacement of the brain, with smaller than expected ventricles and cisterns, and low opening pressure on lumbar puncture, all consistent with a diagnosis of spontaneous intracranial hypotension (Figs. 1, 2) SIH is an uncommon phenomenon, occurring at an incidence of 5 per 100,000 people per year, most often in the fourth and fifth decade, with women affected more than men in a 2:1 ratio. SIH is classically described as presenting with a postural headache associated with meningeal signs [1, 2]. The proposed etiology is a CSF leak through a spontaneous dural defect, frequently in the spine at the cervico-thoracic junction or thoracic regions. SIH symptomatology is typically described as a postural headache with meningeal signs, vertigo, hearing loss, and diplopia [1]. However, it can have heterogeneous presentations, including cranial nerve palsies, cortical vein and sinus thromboses, depressed mental status, weakness, ataxia, bulbar weakness, postural tremor, chorea, vasoconstriction, galactorrhea, radiculopathy, and myelopathy [3]. Changes in hearing, vertigo and dizziness are thought to be from vestibulocochlear nerve deformation and low perilymphatic pressure [4]. Cranial neuropathies, pituitary dysfunction, and brain stem signs are thought to be secondary to brain sagging, which generates downward pressure. More severe pressure may be associated with ataxia, bulbar signs, parkinsonism, and chorea. Radiculopathy and myelopathy are thought to be secondary to local CSF collections [4].

Intracranial imaging findings of SIH include brain sagging, pachymeningeal enhancement, venous engorgement, enlarged pituitary and subdural collections, with or without hemorrhage from stretching of bridging veins [3]. Spinal imaging may reveal distended epidural veins, epidural fluid collections, abnormal nerve root visualization, and meningeal diverticula. CSF leakage site may be detected with either CT or MRI myelography, both of which involve injection of contrast into the epidural space to look for contrast extravasations [3]. Treatment options range from conservative measures such as bed rest, hydration and caffeine to epidural blood patch, epidural fibrin patch, and targeted blood/fibrin patch. These measures are curative in about 75 % of cases [3]. In refractory cases, surgery with meningeal suturing or epidural packing may be attempted. Intrathecal saline injection to restore CSF volume until leak is repaired may be warranted in cases of depressed mental status [1]. However, there are no consensus guidelines on surgical intervention for subdural collections [3].

Spontaneous intracranial hypotension (SIH) is an uncommon but often serious illness that may result in diverse clinical presentations. Unique imaging findings, including brain sagging, subdural collections, and pachymeningeal enhancement should raise concern for the presence of SIH. The use of Trendelenburg positioning for greater than 24 h to assess improvement may be used in evaluation of such cases. When the concern for SIH is high, prompt CT or MRI myelography should be used to confirm diagnosis and guide treatment. A variety of treatment options exist and may provide definitive cure.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Statement of human and animal rights All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

Informed consent As with all case reports from my institution, waiver of informed consent was granted by the IRB.

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