Imaging "worst headache of my life" Part 2: Conditions where initial CT is often normal, but other imaging may be diagnostic

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Headache is a common indication for brain imaging studies performed in the emergency department (ED). When patients present with the "worst headache of my life," imaging often plays a major role in differentiating benign causes of headache from life-threatening causes, and noncontrast computed tomography (NCT) is the most commonly performed imaging test.[1] NCT can detect many emergent causes of the "worst headache of my life," such as hemorrhage, hydrocephalus, ischemia, tumors, and posterior reversible encephalopathy syndrome. However, not all emergent causes of headache are excluded by a negative head CT. Identifying which patients should undergo further imaging work-up, as well as the precise nature of that evaluation, can be a challenge for emergency room physicians and radiologists alike.

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In the first part of this 2-part series, the authors reviewed potentially life-threatening causes of headache that commonly result in visible abnormalities on NCT. In the second part of this series, we consider causes of headache in which the initial NCT usually or often fails to detect the disease, but other imaging may ultimately allow one to make the correct diagnosis.

Diagnosing these particular conditions can be difficult, as physicians in the ED may not pursue further diagnostic imaging if the initial NCT is negative. Nonetheless, some of these conditions may in fact be life-threatening, requiring urgent treatment. Correctly establishing the diagnosis, then, requires appropriate levels of clinical suspicion, and also requires that the radiologist be aware of the potential limitations of the initial NCT and be prepared to suggest alternative imaging strategies for appropriate patients.

Dural sinus thrombosis

Dural sinus thrombosis (DST) causes headache by virtue of impairment of venous outflow from the brain with resultant increase in intracranial pressure. Occasionally the headache is accompanied by altered mentation, decreased level of consciousness, and papilledema. The diagnosis can be difficult to establish on NCT. As a result, patients are often not diagnosed until subsequent imaging studies are performed.

The most specific feature on NCT is that of a hyperdense dural sinus. However, thrombosed dural sinuses commonly do not show this characteristic hyperdensity, which can lead to a false-negative diagnosis (Figure 1).[2] The sensitivity of the hyperdense sinus sign has been reported as low as 20%, and therefore the diagnosis may be missed in a large proportion of NCT examinations.[3] To compound the problem, clinicians may not consider the diagnosis because DST is a relatively uncommon entity. As a result, neither clinician nor radiologist may pursue further imaging.

On contrast-enhanced CT, DST is seen as lack of opacification of the thrombosed dural sinus. More sensitive noninvasive imaging techniques for diagnosis of DST include magnetic resonance (MR) imaging (showing

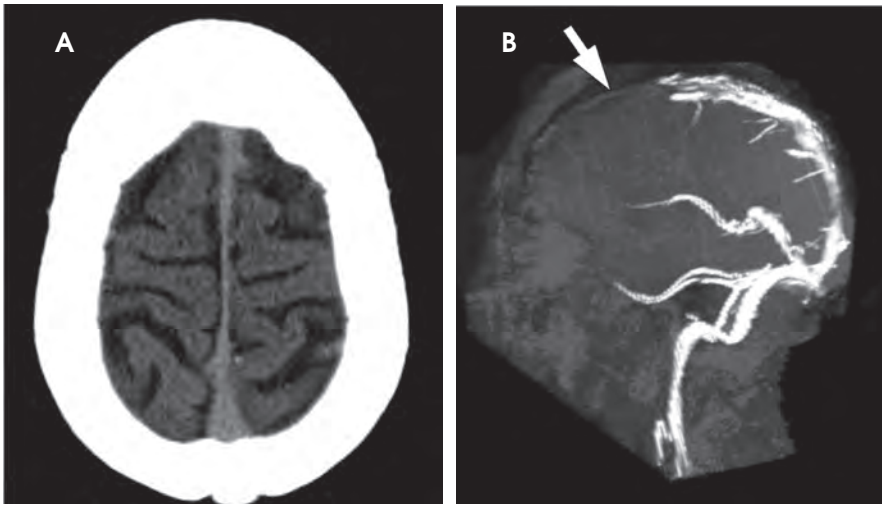

FIGURE 1. *55-year-old man presenting with severe headache. (A) Unenhanced axial CT image is normal; the superior sagittal sinus does not appear hyperdense. (B) Maximum intensity projection of MR venogram demonstrates thrombosis of the anterior portion of the superior sagittal sinus (arrow).*

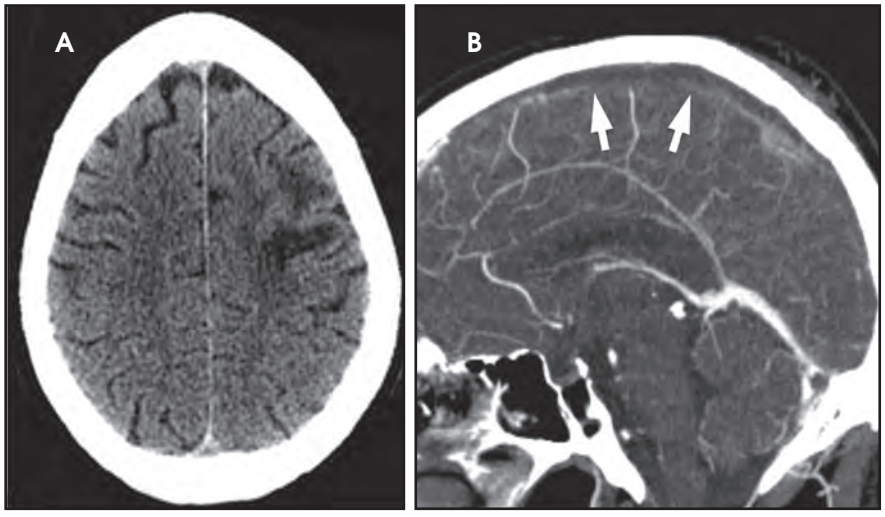

FIGURE 2. *48-year-old man with headache developing over a period of 1 week. (A) Unenhanced axial CT image shows a focal region of subcortical hypodensity region in the left hemisphere with sparing of the overlying cortex. This pattern may mimic vasogenic edema, but can also be seen with ischemia due to venous sinus thrombosis. (B) Sagittal maximum intensity projection image from CT venogram confirms the presence of thrombus within the superior sagittal sinus (arrows).*

loss of a flow void within the affected dural sinus), MR venography (showing loss of flow-related enhancement), and CT venography (in which the thrombus will be seen as a filling defect).

Abnormalities of the brain parenchyma may be present in some cases of DST. Venous infarction results from thrombosis of dural venous sinuses and/or cortical veins. Although venous infarction can be associated with evidence of thrombosis on NCT (ie, a hyperdense sinus), this is not always the case. In some cases of DST, the parenchymal findings may be the only abnormality seen on NCT. It is therefore useful to consider the appearance of venous infarction on NCT, as its appearance may mimic other disease processes.

On initial inspection, venous infarction may mimic the appearance of arterial infarction. Importantly, however, venous infarctions will not be confined to the usual arterial vascular territories, which may provide a clue to the correct diagnosis. Furthermore, in contrast to arterial infarction, venous infarction characteristically involves the subcortical white matter first and may initially spare the overlying cortex, although cortical infarction may follow if venous occlusion is severe or prolonged.[4] As a result of this cortical sparing, in some cases, the initial NCT appearance may also mimic vasogenic edema associated with a neoplasm, such as a metastasis (Figure 2). There may be associated hemorrhage in the brain parenchyma in some patients with DST, although this is not present in all cases. Patients presenting with headache who demonstrate subcortical edema or hemorrhage on initial CT imaging should therefore undergo further investigation of the venous sinuses.

Arterial dissection

Headache is present in approximately 70% of patients with dissection of the carotid or vertebral artery (co-called cervicocephalic arterial dissection).[5] Similar to DST, cervicocephalic arterial dissection is a cause of worst headache of my life that is often clinically unsuspected, which causes delay in performance of the appropriate imaging study. The most common locations of cervicocephalic arterial dissections are in the cervical portion of the internal carotid artery (within a few centimeters of the carotid bifurcation) and the segment of the vertebral artery that extends around the C1-C2 vertebral body complex. Although arterial dissection can result in stroke, ischemia may be present in fewer than half of patients with acute dissection at initial presentation.[6] Thus, it is not uncommon that brain imaging does not detect any abnormality (Figure 3).

Unenhanced CT imaging of the neck is of limited help in establishing the diagnosis of arterial dissection; the dissection is inconspicuous against the background of soft tissue. Instead, CT angiography or MR angiography are sensitive imaging techniques for the diagnosis. Findings potentially seen

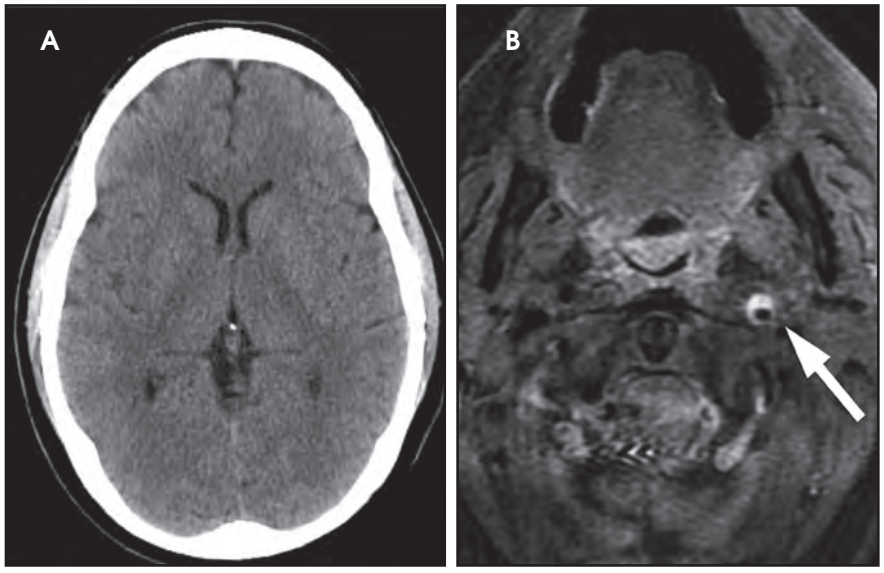

FIGURE 3. *55-year-old man with 3 weeks of headache, initially treated for sinusitis without improvement. He subsequently developed blurred vision and on exam was found to have Horner syndrome. (A) Unenhanced axial CT image is normal. (B) Axial T1-weighted image with fat suppression through the neck shows crescent-shaped hyperintense signal in the wall of the left internal carotid artery (arrow) with associated luminal narrowing due to carotid dissection.*

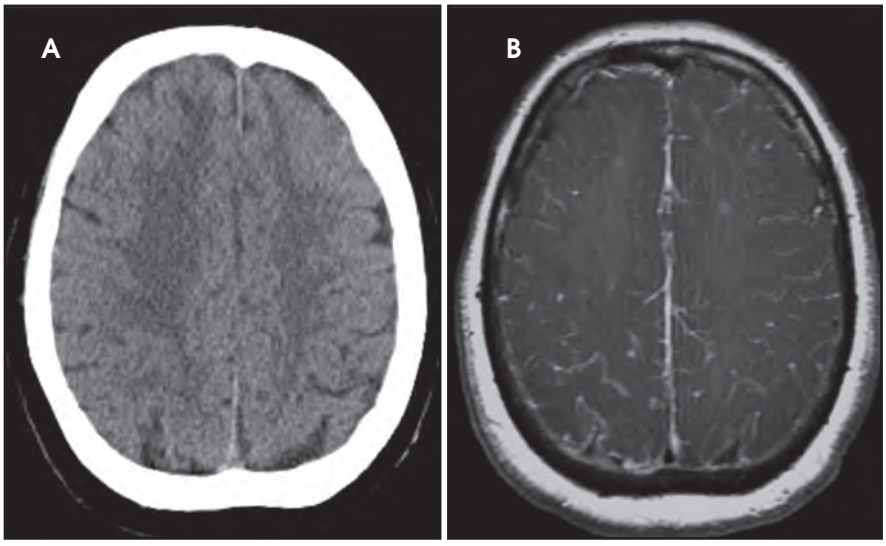

FIGURE 4. *54-year-old woman with headaches, fevers, and confusion. (A) Unenhanced axial CT image is normal. (B) Postcontrast axial T1-weighted image demonstrates leptomeningeal enhancement due to streptococcus pneumoniae meningitis. The absence of such enhancement should not be interpreted as sufficient evidence to exclude meningitis.*

using either technique can include luminal narrowing due to the intramural hematoma or pseudoaneurysm. When findings on these studies are equivocal, catheter angiography may be indicated. Particular clinical features that should prompt imaging investigation for the possibility of dissection include the presence of neck or facial pain, orbital pain, or Horner syndrome.[6]

Meningitis

Although it can be a cause of severe headache, the diagnosis of meningitis is established by cerebrospinal fluid (CSF) analysis and clinical symptoms. In uncomplicated cases of meningitis, imaging is not necessary.[7] In such cases, CT is usually normal and does not influence management.[8] When an abnormality is present, it usually takes

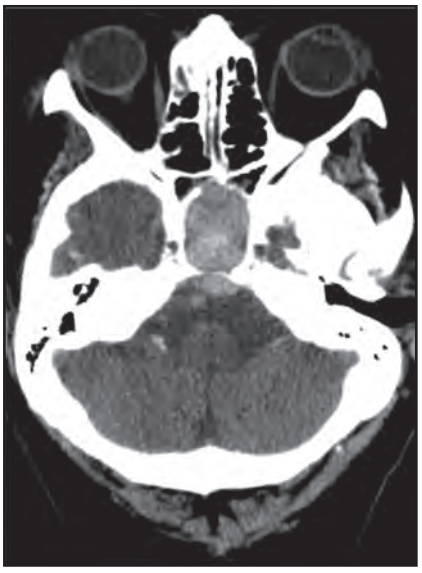

FIGURE 5. *77-year-old woman with sudden onset of severe headache and visual changes. Unenhanced axial CT image shows high density in the sella due to hemorrhage (ie, pituitary apoplexy). Note the smooth bony expansion of the sella, the result of the presence of a slowly growing macroadenoma.*

the form of mild, transient hydrocephalus.[8] Occasionally, leptomeningeal enhancement can be seen on postcontrast imaging (Figure 4); the absence of such enhancement does not exclude meningitis.

Uncommonly, other complications can arise in the setting of more severe infections, and imaging can play an important role in these cases. Complications of severe meningitis can include ventriculitis, extra-axial fluid collections (empyemas and subdural effusions), parenchymal infection (cerebritis or abscess), and secondary infectious vasculitis.[7] Imaging with CT or MR may help to identify these complications. Additionally, imaging may help to identify the cause of meningitis in cases where there is a direct pathway for spread of infection, such as in the case of complicated sinusitis, skull base fractures, or prior surgery.

Pituitary Apoplexy

Pituitary apoplexy is an uncommon cause of worst headache of life, and denotes symptomatic hemorrhage within the pituitary gland, usually into

FIGURE 6. *20-year-old woman with chronic headaches. Maximum intensity projection from a MR venogram shows narrowing at the junction of the transverse and sigmoid sinuses (arrow), a finding that has been reported in association with idiopathic intracranial hypertension.*

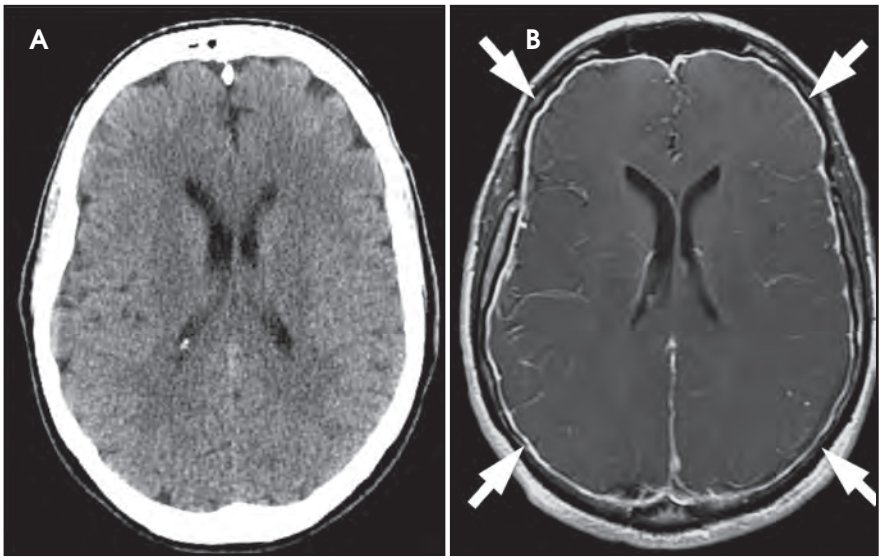

FIGURE 7. *58-year-old man who developed a severe postural headache while lifting furniture. (A) Unenhanced axial CT image is normal. (B) Postcontrast axial T1-weighted image demonstrates diffuse, smooth dural enhancement (arrows) characteristic of intracranial hypotension.*

a pituitary adenoma. In many patients who present with pituitary apoplexy, the adenoma has not been previously diagnosed, and therefore the diagnosis is not often suspected initially.[9] Clinical symptoms most commonly consist of headache, vomiting, and visual disturbance.[9, 10]

Despite what may be pronounced clinical symptoms, initial imaging may fail to reveal an abnormality. In a previous series of patients presenting with pituitary apoplexy, intrasellar hemorrhage was not recognized on initial NCT in the majority of cases, although intrasellar tumors were visible in almost all cases on NCT.[10,11] Failure to detect intrasellar hemorrhage on the initial CT potentially could result from partial volume averaging, and in some cases multiplanar reformation may prove helpful in evaluating the sella when clinical suspicion is high.

When abnormalities of the pituitary gland are visible, CT may reveal expansion and hyperdensity of the gland, which may be accompanied by mass effect on the suprasellar structures, including the optic chiasm (Figure 5). MRI, by contrast, has demonstrated very high sensitivity for the detection of pituitary apoplexy, and can be used to confirm the diagnosis.[10] Careful scrutiny of the sella and suprasellar region should be a routine part of the search pattern when reviewing the initial imaging of patients presenting with the "worst headache of my life," then, especially in patients presenting with headaches accompanied by visual disturbances.

Disorders of CSF pressure

Headaches caused by disorders of CSF pressure are not uncommon. In some patients with headache, CSF pressure may be too high, as in the case of idiopathic intracranial hypertension (IIH, aka, pseudotumor cerebri), or too low, as in the case of intracranial hypotension.

Patients with IIH may present to the ED acutely with severe headache, but often have a preexisting history of chronic headaches. They most commonly are young, obese women.[12] On physical exam, these patients may have papilledema as a result of the increased intracranial pressure. The discovery of papilledema may result in a head CT being performed to exclude an intracranial mass, but in most cases of IIH the initial CT is normal.

Patients with papilledema often undergo additional imaging of the venous sinuses in order to exclude venous sinus thrombosis as a cause for elevated intracranial pressure. Once thrombosis is excluded, one sign suggesting the presence of IIH that may be visible on venous sinus imaging is narrowing at the junction of the transverse and sigmoid sinuses (Figure 6). This finding is hypothesized to be due either to intrinsic stenosis of the vessel or collapse of the sinus due to increased CSF pressure.[13] On MRI, other potentially subtle findings associated with IIH include flattening of the posterior globe, dilation, and tortuosity of the optic nerve sheath, and an empty sella.[12] It should be emphasized that these signs might suggest the diagnosis of IIH, but that ultimately the diagnosis is established by lumbar puncture with measurement of opening pressure.

Patients with intracranial hypotension usually present with headache that is worse when standing and improves when lying down. Intracranial hypotension may be iatrogenic (due to lumbar puncture or surgical penetration of the thecal sac), but in many cases is due to spontaneous leakage of CSF from around the spine.[14] In cases of spontaneous intracranial hypotension, the onset of symptoms may be abrupt, resulting in presentation to the emergency room with severe headache.

Initial clinical misdiagnosis of intracranial hypotension is common, which is compounded by the fact that initial CT imaging in these patients is often normal (Figure 7).[15] When present, subtle features of cranial sagging and cerebellar tonsillar ectopia may be appreciated on CT. Bilateral subdural hygromas or hematomas may also accompany intracranial hypotension, and can be identified on the initial scan, although the cause of the collections may not be immediately recognized.[16] In patients without a history of trauma, bilateral subdural collections should prompt consideration of the diagnosis of intracranial hypotension, and MRI should be pursued as the next appropriate imaging step.

Diffuse smooth dural enhancement is the most suggestive MR imaging feature. Other variably present findings include crowding of the basilar cisterns due to brain sagging, pituitary enlargement, cerebellar tonsillar ectopia, and distension of the venous sinuses.[17] Diagnosis is ultimately determined through the combination of clinical symptoms, imaging findings, and CSF pressure measurement.[18]

Conclusion

Although NCT is the most common modality for evaluating the "worst headache of my life," several potentially life-threatening diseases will commonly fail to show abnormalities on the initial NCT. These conditions may be easily missed if they are not actively considered by treating ED physicians and radiologists. While certain details of patients' presentations may provide clues to suggest the presence of these conditions, appropriate selection of additional imaging is also critical to successful diagnosis. By understanding the strengths and limitations of various imaging modalities, the radiologist may be uniquely positioned to anticipate conditions at risk of being missed on initial imaging, and to make suggestions to help avoid this potential hazard.

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