CLINICAL CASE - TEST YOURSELF Neuroradiology

An unusual cause of thunderclap headache with intracranial hemorrhage in a 40-year-old woman

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PART A

A 40-year-old woman with no significant medical history presented to the emergency department due to a sudden onset of severe bilateral frontal and parietal headache with vomiting, with no history of trauma. Neurological examination revealed no meningeal or focal neurological signs. A CT scan was performed (Fig. 1), with a subsequent CT angiography revealing no aneurysm or vascular malformation (not shown). Five days after admission, an MRI and MR angiography of the head were performed (Fig. 2). After four months, follow-up MRI and MRA were performed (Fig. 3).

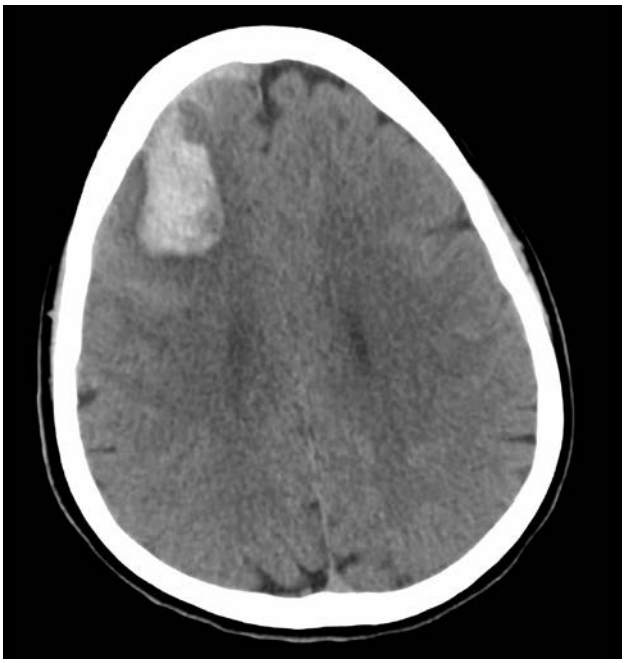

Fig.1: *Non contrast head CT*

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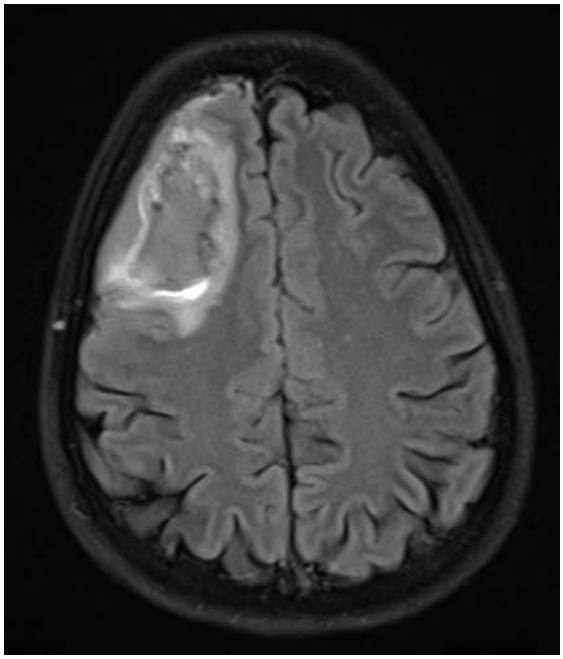

Fig.2a: *T2 FLAIR sequence*

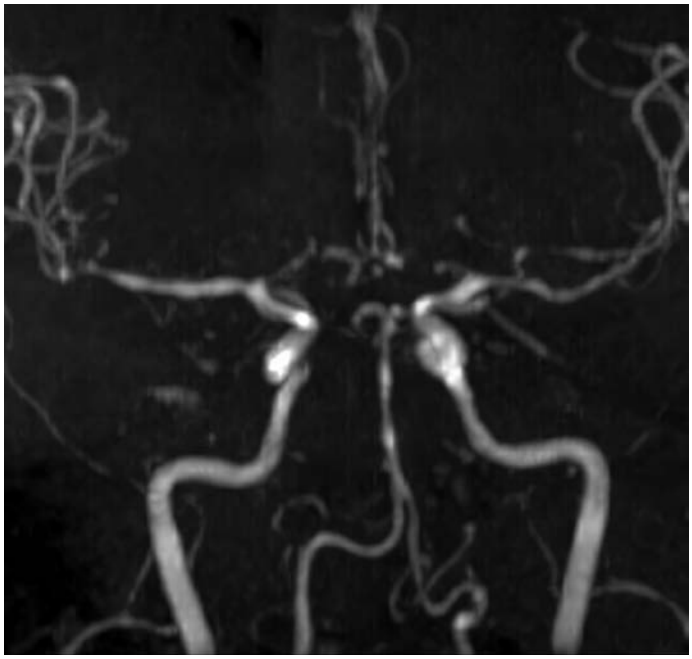

Fig.2b: *TOF MIP*

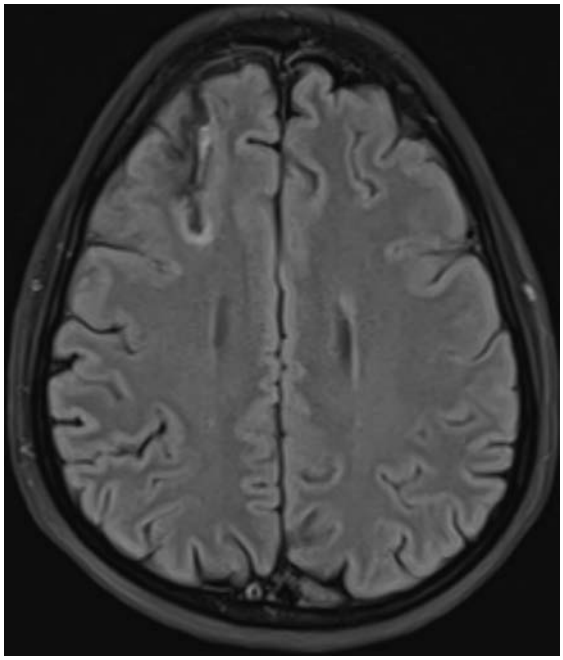

Fig.3a: *T2 FLAIR sequence*

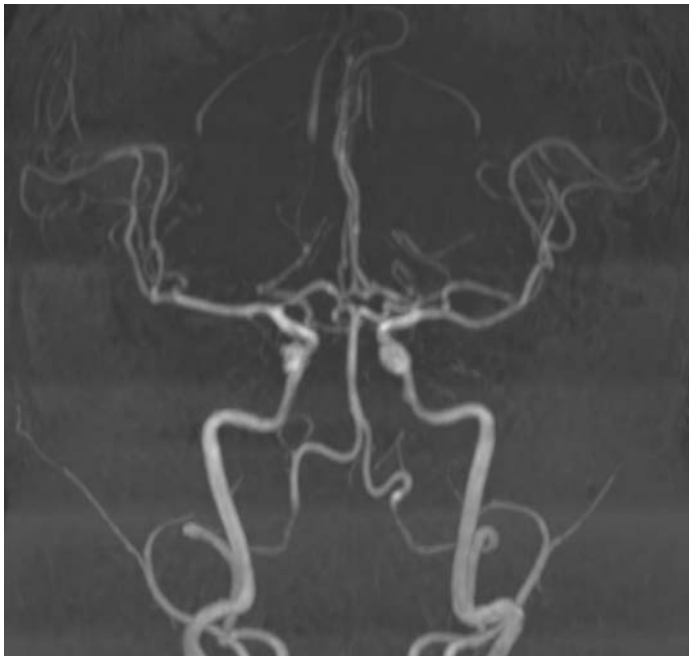

Fig.3b: *TOF MIP*

PART B

Diagnosis

Intraparenchymal hematoma with concomitant small subdural and subarachnoid hemorrhage (SAH) in a patient with Reversible Cerebral Vasoconstriction Syndrome (RCVS).

A plethora of possible underlying conditions have to be included in the differential diagnosis of a spontaneous intraparenchymal hemorrhage (IPH). The two commonest causes of IPH are hypertension and cerebral amyloid angiopathy (CAA). Hypertensive hematomas tend to occur in the basal ganglia, thalami, pons, midbrain, and cerebellum, due to chronic stress, causing the eventual rupture of small intraparenchymal brain vessels [1].

Similarly, CAA weakens cortical blood vessels due to β-amyloid deposition, leading to bleeds in lobar, cortical, or cortical-subcortical regions, usually in older patients. [2] Other causes of IPH include coagulopathy, vascular malformation rupture, cerebral venous thrombosis, mycotic aneurysm rupture, moyamoya, tumor, hemorrhagic conversion of an ischemic stroke, or vasculitis [2].

The presentation of an IPH varies depending on size, location, and time of onset. The chief complaint is most commonly headache; however, patients with small or deep hematomas rarely have this symptom. Superficial hematomas may exhibit headache close to their location (i.e., forehead in frontal lobe hematomas) as well as meningeal signs.

Nausea and vomiting may occur due to elevated intracranial pressure or stimulation of medullary vomiting centers. A decrease in the level of consciousness may be present; however, it is rare with small, supratentorial, unilateral hematomas. Seizures may be present, usually either at onset or within two weeks. Focal neurologic signs such as aphasia, hemiparesis, ataxia, hemianopsia, etc, may be present depending on the affected area [3].

The role of imaging is essential, with a non-contrast CT scan being the first-line diagnostic modality in the acute phase. CT and MR angiography are appropriate initial investigations to detect macrovascular bleeding sources, with DSA being reserved for further investigation of cases with an expected high yield in detecting a macrovascular source [4].

Several CT features have prognostic utility, such as the presence of intraventricular hemorrhage and hematoma volume (>32 ml supratentorially or >21 ml infratentorially), and are incorporated in the ICH score [1]. The spot sign in CTA may be moderately predictive of hematoma expansion [4].

Treatment consists of medical management, which includes blood pressure regulation, coagulation disorder reversal with vitamin K and Fresh Frozen Plasma (FFP) or Prothrombin Complex Concetrates (PCC) infusion, platelet transfusion, withholding of anticoagulants and antiplatelet medication, thromboprophylaxis measures in appropriate patients (with intermittent pneumatic compression, IVC filters and low molecular weight or unfractionated heparin after cessation of bleeding in immobile patients), intracranial pressure (ICP) regulation with mannitol and hypertonic saline administration, glucose correction, antipyretic and antiepileptic medication.

Surgical management includes hematoma evacuation for patients with cerebellar or supratentorial hemorrhage with neurological deterioration, decompressive craniectomy for supratentorial hemorrhage with GCS score < 8, large hematomas with significant midline shift or medically intractable ICP, and ventricular drainage for hydrocephalus with or without IVH [5].

In our case, the patient did not indicate neurosurgical intervention, so she was admitted to the neurology department for conservative treatment and further investigation.

The MRA performed on the 5th day of admission revealed focal reduction in diameter of multiple intracranial arteries, mainly the basilar, anterior, middle, and posterior cerebral arteries. The ECG revealed mild right frontal-temporal cerebral dysfunction; thus, antiepileptic medication was initiated. A comprehensive serological immune panel was ordered, which revealed no abnormal test values. No elevated blood pressure was recorded.

The patient was discharged in good clinical condition with no neurological deficits, with instructions for antiepileptic and conditional antihypertensive medication. Follow-up MRI-MRA performed after four months revealed absorption of the hematoma with normal-width intracranial vessels.

Based on the above, as well as the location of the hemorrhage, the age, history of the patient, and the clinical presentation, this was deemed a probable case of Reversible Cerebral Vasoconstriction Syndrome. Post-sub-

arachnoid hemorrhage vasoconstriction was also a consideration; however, the small extent of the SAH as well as the pattern of vessel narrowing deemed it unlikely.

RCVS encompasses a group of conditions marked by alternating narrowing and dilation of medium-to-large cerebral arteries. Clinically, it presents with repeated episodes of sudden, severe "thunderclap" headaches (TCH), sometimes accompanied by focal neurological deficits.

The vasoconstriction is usually reversible, resolving spontaneously within approximately three months [6]. RCVS diagnosis is based on key clinical features of "thunderclap" headache (reaching peak intensity within ≤1 minute) or severe recurrent headache, cerebral vasoconstriction on imaging in at least 2 different arteries and resolution of vasoconstriction by 3 months, with no evidence of vasculitis on laboratory analysis. RCVS2, a diagnostic score, was developed, which is calculated based on the presence of a single or recurrent episodes of TCH, lack of carotid intracranial artery involvement, presence of a vasoconstrictive trigger, female gender and presence of SAH [7].

Imaging, apart from the pattern of narrowing of the intracranial arteries, may demonstrate SAH or areas of ischemia.

Additional findings can include intraparenchymal hemorrhage, subdural bleeding, and cerebral edema—the latter occasionally displaying features similar to posterior reversible encephalopathy syndrome (PRES), a condition that may co-occur with RCVS [6].

Vasoactive substances are the most frequent trigger factors for RCVS, followed by pregnancy and sexual intercourse. Surgery or trauma may also be a potential trigger, associated with a higher risk of residual neurological deficits [8].

While the overall prognosis is often favorable, patients who develop neurological deficits may face more serious outcomes [6]. Treatment is mostly symptomatic and includes analgesics, oral calcium channel blockers, removal of vasoconstrictive factors and avoidance of glucocorticoids, because they may significantly worsen outcomes [9].

This case highlights the importance of considering RCVS in the differential diagnosis of non-traumatic intracerebral hemorrhage, particularly in younger patients with a clear medical history.

Recognition of this condition by the radiologist can significantly aid clinical investigation and further management. **R**

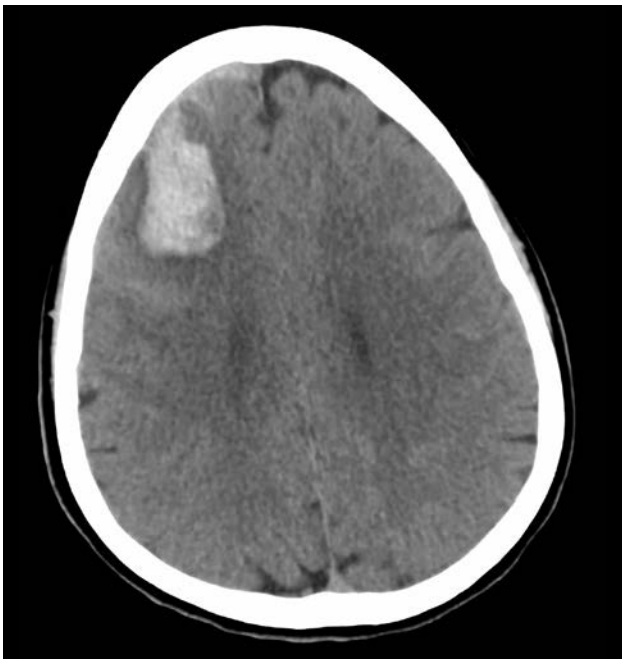

Fig.1: *Right frontal lobe intraparenchymal hematoma with concomitant small subdural hematoma and subarachnoid hemorrhage in the adjacent sulci.*

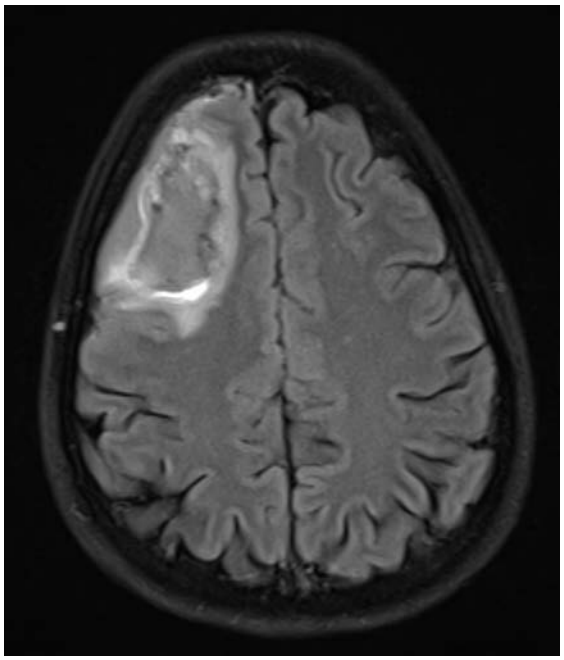

Fig.2a: *Right frontal lobe intraparenchymal hematoma.*

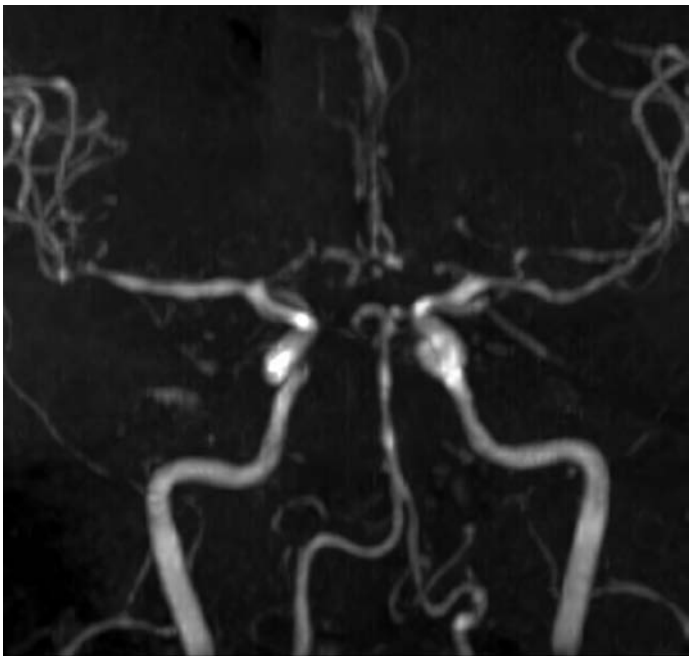

Fig.2b: *Segmental narrowing of multiple cerebral vessels, mainly of the basilar, anterior and middle cerebral arteries.*

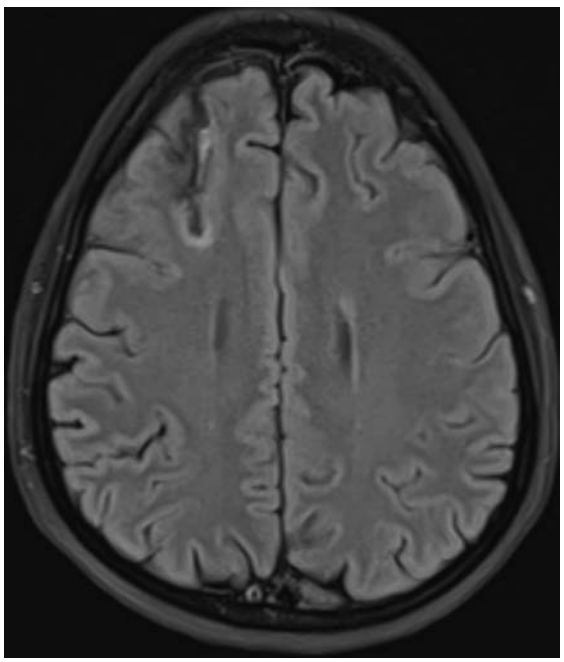

Fig.3a: *Resolution of the hematoma.*

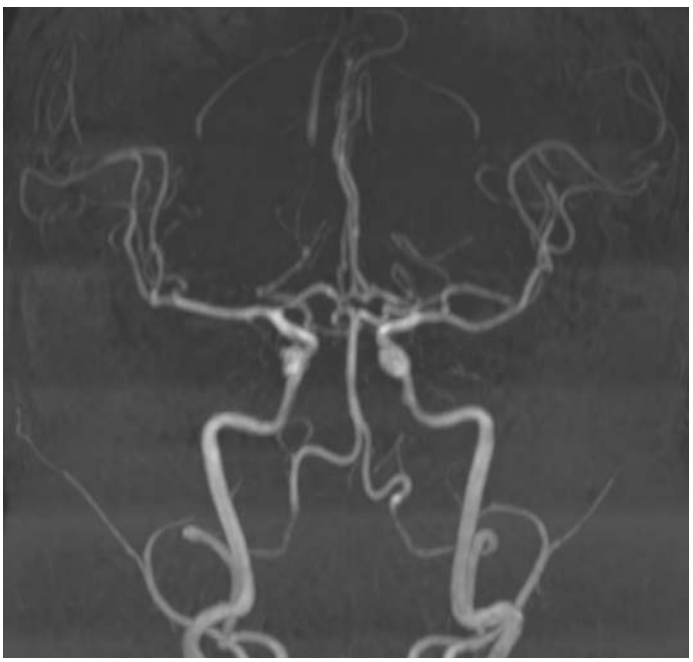

Fig.3b: *Normal caliber cerebral vessels.*

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