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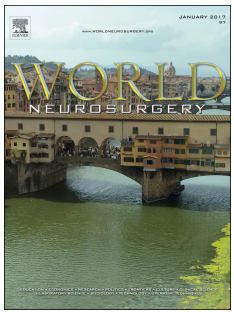

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Acute convexity subarachnoid hemorrhage: what the neurosurgeon needs to know

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Non-traumatic convexity/convexial/cortical subarachnoid hemorrhage, cerebral amyloid angiopathy, intracerebral hemorrhage, ischemic stroke, cortical superficial siderosis, stroke

When faced with a nontraumatic subarachnoid hemorrhage (SAH), neurosurgeons typically search for the commonest cause: a ruptured aneurysm. However, an increasingly recognized and differential diagnosis is acute convexity subarachnoid hemorrhage (cSAH). In cSAH – which accounts for about 6% of all nontraumatic SAH[1]- bleeding is limited to the subarachnoid space over the convexities of the brain and not extending into the sylvian fissures, parenchyma, ventricles, or basal cisterns,[2] a pattern readily distinguished from the more diffuse pattern seen in aneurysmal SAH. The two commonest causes of cSAH typically affect different age groups and cause characteristic syndromes. In older patients (>60 years) the commonest cause is cerebral amyloid angiopathy (CAA), presenting with transient focal neurological symptoms (TFNE) of unilateral spreading sensory or motor symptoms[3,4] attributed to cortical irritation from the overlying acute hemorrhage. In younger people (<60 years) the commonest cause is reversible cerebral vasoconstriction syndrome (RCVS), typically presenting with recurrent thunderclap headaches[5,6]. Other, much less common, causes of cSAH include posterior reversible encephalopathy syndrome (PRES), cerebral venous thrombosis (CVT), infections (e.g. endocarditis), coagulopathies (e.g. disseminated intravascular coagulation) and Moya Moya syndrome. Unlike other patterns of SAH, cSAH is hardly ever due to an underlying macrovascular pathology such as a ruptured aneurysm, arterio-venous-malformation (AVM), or tumor[5,6].

The sensitivity of unenhanced CT is very high in diagnosing acute cSAH and is usually the first investigation[7]. cSAH can also be readily detected on MRI FLAIR sequences. Over time cSAH evolves into cortical superficial siderosis (cSS) due to deposition of hemosiderin (generated by blood breakdown) in subpial cortical layers; gradient-recalled echo and susceptibility-weighted images can therefore detect evidence of prior episodes of cSAH as sulcal hemosiderin or cSS (Figure 1)[5].

Convexity SAH has received limited attention from neurosurgeons, perhaps because of the low yield of macrovascular causes and a perceived benign prognosis. Patients with cSAH might therefore not be routinely followed-up and are usually considered of interest mainly for neurologists. But is this really true? In recent years it has become clear that cSAH has distinct and readily recognized causes, which do not always have a benign course.

RCVS is the most likely etiology in patients under the age of 60[8]. Multifocal and synchronous cSAH can be seen in RCVS, a feature that is not typical of CAA. RCVS typically presents with thunderclap headache identical to that seen in aneurysmal SAH: the key distinction is that in RCVS headaches are usually recurrent, a feature said to be almost diagnostic of this entity[6]. Nevertheless, patients with RCVS are often referred directly to neurosurgeons from the emergency department. Although cerebral vasculitis is considered when either noninvasive or catheter angiography shows the radiological appearance of multiple areas of arterial caliber abnormality, in our experience the clinical picture of recurrent thunderclap headache is highly suggestive of RCVS and should help clinicians to avoid unnecessary tests for cerebral vasculitis (e.g. lumbar puncture or cerebral biopsy).

In patients over the age of 60, cSAH is frequently associated with imaging markers of CAA[4], an age-related process of accumulation of amyloid-beta in superficial leptomeningeal and cortical arterioles near the brain's cortical surface. In CAA, cSAH results from bleeding into the subarachnoid space from abnormally fragile amyloid-laden vessels[9]. The typical clinical presentation is with TFNE, typically stereotyped recurrent attacks of sensory or motor disturbance (numbness, tingling, weakness) affecting face, arm or leg but without thunderclap headache (unlike typical aneurysmal SAH)[10]; a less severe or gradual onset headache can occur. Such patients might therefore present to TIA clinics or emergency departments before referral to neurosurgery. In cSAH due to probable CAA it is increasingly recognized that the course might not be as favorable as previously believed[11], with a high risk of future symptomatic ICH of up to 19% per patient-year in patients fulfilling the Boston criteria for probable CAA[11,12]. Gold-standard for CAA diagnosis is neuropathological confirmation, but the modified Boston criteria have emerged as a reasonable tool instead. They include CMB and cSS to noninvasively diagnose CAA with good diagnostic accuracy[13]: Therefore, once a diagnosis of SAH is confirmed, the next most useful diagnostic tool for CAA is MRI with blood-sensitive sequences to detect biomarkers of CAA. Probable CAA (with highest diagnostic certainty) is diagnosed with the following criteria: age ≥55 years; multiple ICH restricted to lobar, cortical or corticosubcortical regions or single lobar, cortical or corticosubcortical ICH in addition to focal or disseminated SS; with no other underlying cause present.

DSA is rarely required in patients with cSAH if clinical and radiological features are supportive of either RCVS or CAA (Figure 2) but remains essential in cases of diagnostic uncertainty.

The treatment of cSAH depends on the cause. In RCVS, nimodipine is often used, and might reduce frequency and severity of thunderclap headaches though controlled trials are not available[14]. Steroids do not seem to be helpful and might even be harmful[15]. A careful search for known precipitating factors (e.g. sympathomimetic drugs, SSRIs, etc.) is important and patients should be advised to avoid any triggers identified. For cSAH due to CAA, typically TFNE recur over a short time period[16]. These attacks seem to respond to both anticonvulsant drugs (e.g. levetiracetam) or migraine preventives (e.g. topiramate)[17], though there are no controlled trials. Patients should be reassured that although these attacks may be distressing, each attack does not usually reflect new bleeding, and the natural history is usually of improvement and remission over days to weeks. There are no interventions proven to reduce ICH or recurrent cSAH risk. However, extrapolating data from patients with CAA presenting with ICH, blood pressure reduction to a target of 130/80 is reasonable, as well as avoiding exposure to antithrombotic (antiplatelet and anticoagulant) drugs wherever possible.

Further research is needed to understand the underlying pathophysiology of cSAH in both RCVS and CAA, which should allow rational treatments proven to reduce complications and improve outcomes in both of these conditions. In the meantime, neurosurgeons should be aware of the highly characteristic clinical-radiological syndromes associated with cSAH because these require a different approach to investigation to the more common aneurysmal form of nontraumatic SAH.

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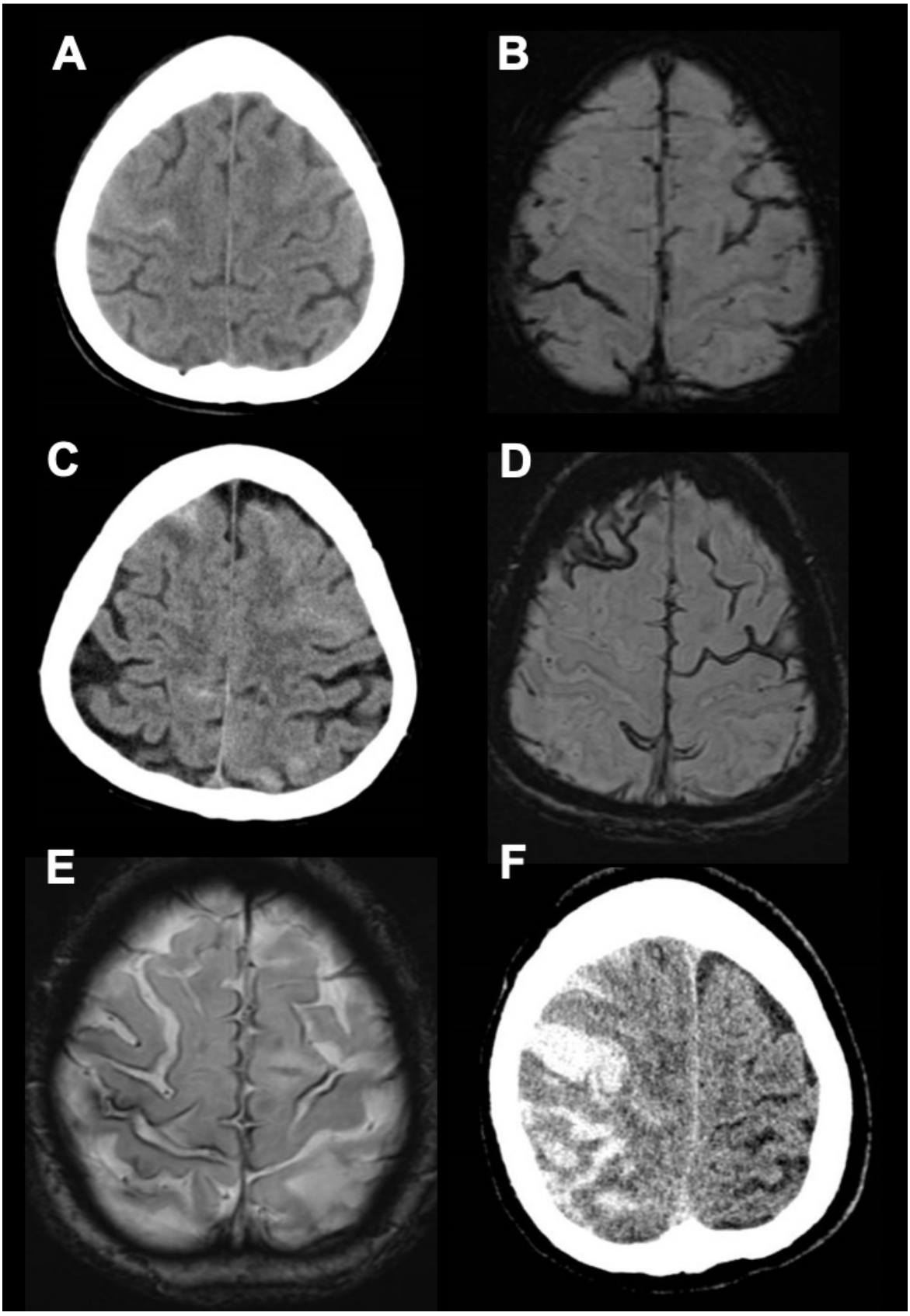
FIGURE LEGEND

Figure 1. Axial images of 3 patients with cSAH: A) CT scan showing acute cSAH in a patient with left sided migratory sensory symptoms; B) susceptibility-weighted image (SWI) of the same patient 11 days after the acute cSAH clearly showing sulcal hemosiderin and evolving cSS in the areas of previous cSAH; C) CT scan of a patient 3 days after presentation with thunderclap headache; D) SWI image of the same patient 6 months after the acute cSAH, showing cSS in the areas of previous cSAH; E) T2*-weighted gradient-recalled echo (GRE) who three years earlier had cSAH associated with clumsiness and tingling of the left hand, showing typical disseminated cSS; F) CT scan of the same patient who went on to develop an ICH 3 months after the MR scan shown in panel E).

Figure 2. Suggested diagnostic pathway for acute, nontraumatic cSAH.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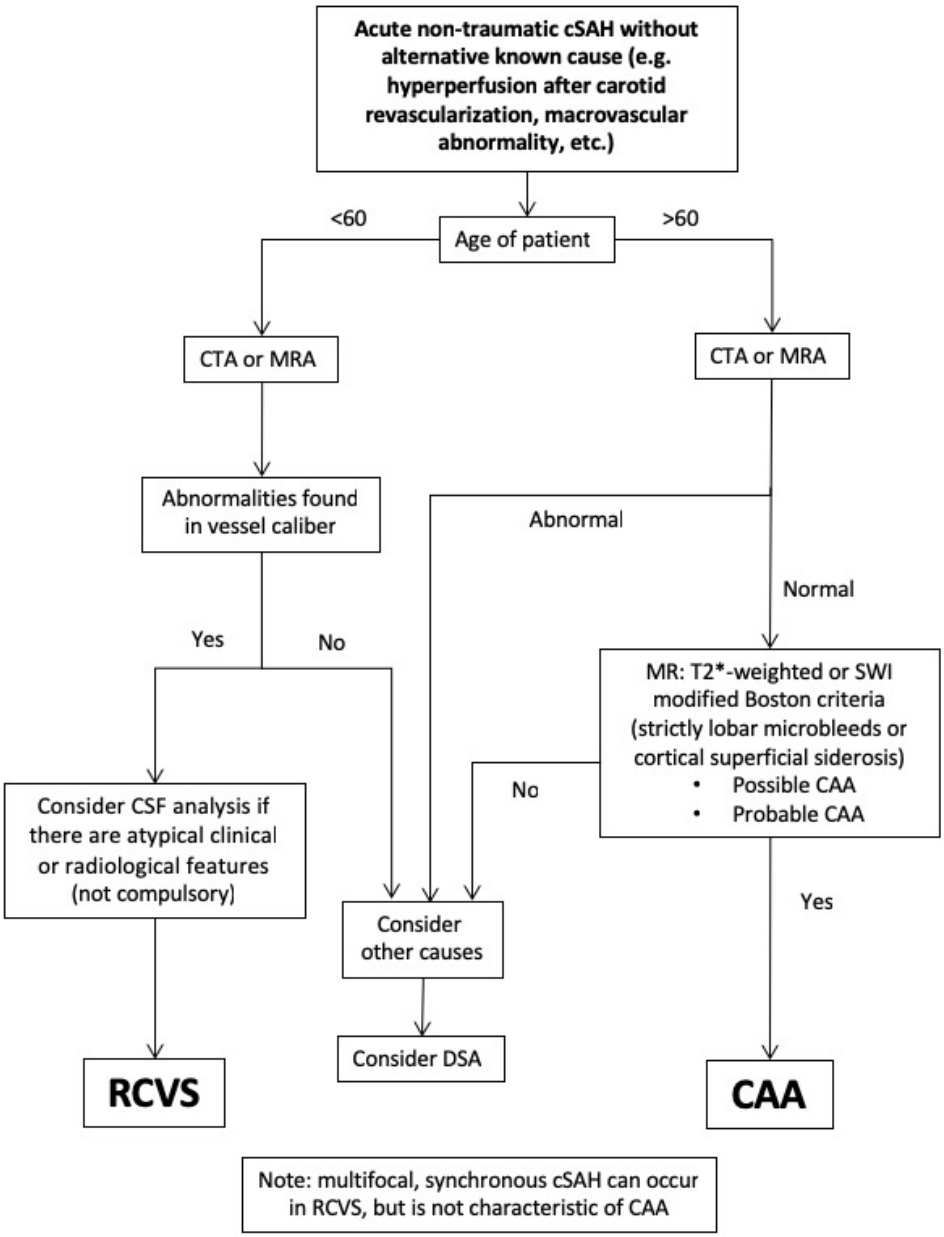
Acute non-traumatic cSAH without alternative known cause (e.g. hyperperfusion after carotid revascularization, macrovascular abnormality, etc.)
Age of patient
<60
>60
CTA or MRA
CTA or MRA
Abnormalities found in vessel caliber
Abnormal
Normal
Yes
No
MR: T2*-weighted or SWI modified Boston criteria (strictly lobar microbleeds or cortical superficial siderosis)
• Possible CAA
• Probable CAA
No
Yes
Consider CSF analysis if there are atypical clinical or radiological features (not compulsory)
Consider other causes
Consider DSA
RCVS
CAA
Note: multifocal, synchronous cSAH can occur in RCVS, but is not characteristic of CAA