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CASE REPORT - VASCULAR

Perimesencephalic subarachnoid hemorrhage with a positive angiographic finding: case report and review of the literature

Ahmad Hafez[1] · **Jussi Numminen**[2] · **Raj Rahul**[1] · **Juha Järveläinen**[2] · **Mika Niemelä**[1]

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Abstract The vast majority of perimesencephalic subarachnoid hemorrhage cases are reported as negative-finding etiologies. Recently, high-resolution images allowed us to overcome the previous difficulty of finding the source of bleeding, which underlies the concept of a "negative finding". We discovered a venous etiology, hidden behind the tip of the basilar artery; namely, the lateral pontine vein. Here, we review the literature on perimesencephalic subarachnoid hemorrhage and on venous aneurysm. We highlight this type of aneurysm as a candidate source of perimesencephalic hemorrhage. This case may change our way of dealing with what we have termed a negative finding of subarachnoid hemorrhage.

Keywords Perimesencephalic subarachnoid hemorrhage · Venous aneurysm

Introduction

The etiology of perimesencephalic subarachnoid hemorrhage (pSAH) has long been debated, with most studies presenting negative imaging findings [2, 3, 5, 6, 8, 13–15, 20, 23, 25–27, 30]. Patients with pSAH and negative radiological findings often have a better prognosis than patients with an identifiable arterial aneurysm as the cause of subarachnoid hemorrhage (SAH). To identify potential etiologies of pSAH, we have to think outside the box; possibilities include venous aneurysms or pouches, thrombosed arterial aneurysms or pseudo-aneurysms, and small perforator vessel fistulas. Investigation of these is, however, often beyond the scope of our daily practice.

We report a rare case of pSAH with a positive, non-arterial aneurysmal, radiological finding of what might represent a ruptured venous aneurysm. We review the case-specific details that led us to discover this rare etiology of pSAH.

Case report

Initial presentation

A 66-year-old female was referred to our tertiary neurosurgical center (Helsinki University Hospital, Helsinki, Finland) after complaining of acute severe headache. The headache started acutely in the evening, lasting for a couple of seconds before decreasing in intensity. There were no deficits in level of consciousness, although she felt somewhat drowsy. Time from start of headache to admission was below 4 h. The patient had a positive medical history of hypertension, hypercholesterolemia, diabetes mellitus type II, and hypothyreosis. She had no verified history of tobacco smoking, alcohol abuse, or previous head trauma.

The primary head computerized tomography (CT) revealed SAH in a typical perimesencephalic pattern: blood in the perimesencephalic cisterns anterior to the brainstem with an extension into the ambient cisterns and basal parts of the Sylvian fissures (Fig. 1a).

Upon presentation, the patient was totally alert and oriented, but suffering from headache, stiff neck, photophobia, and vomiting (Hunt & Hess 2, WFNS grade 1). The primary cerebral CT angiography (CTA) was negative for arterial aneurysms or other cerebrovascular malformations (Fig. 1b and c). The

✉ Ahmad Hafez
ext-ahmad.hafez@hus.fi

[1] Department of Neurosurgery, University of Helsinki and Helsinki University Hospital, Topeliuksenkatu 5, PB-266, 00029 Hus, Finland

[2] Department of Radiology, University of Helsinki and Helsinki University Hospital, Topeliuksenkatu 5, PB-266, 00029 Hus, Finland

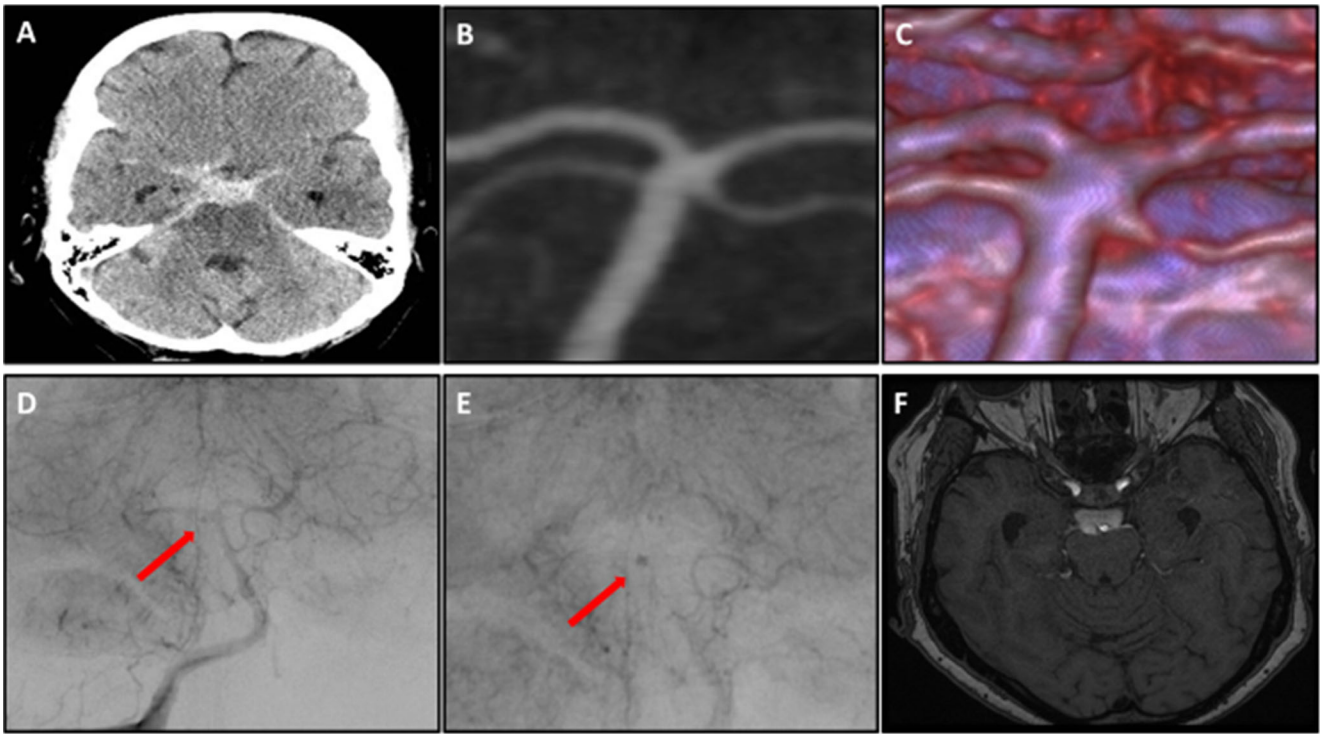

Fig. 1 Non-contrast CT at presentation, axial cut (**a**) shows a subarachnoid hemorrhage limited to the interpeduncular, ambient cisterns, base of Sylvian fissures. **b**, **c** CTA shows a normal basilar artery with its branching area viewed anteriorly. DSA, late arterial time (**d**, **f**) shows an aneurysm (*red arrow*) on the right side near the tip of the basilar artery. Time of flair MR with contrast (F) shows an aneurysm, the cause of the perimesencephalic hemorrhage

patient was admitted to the neurosurgical intensive care unit. A digital subtraction angiography (DSA) was performed 3 days later. The DSA showed an aneurysm at the junction of the superior cerebellum artery (SCA) and the posterior cerebellar artery (PCA). The aneurysm was not acting as a normal arterial aneurysm, as it filled in late arterial phase or early venous phase (Fig. 1d and e). To exclude the possibility of a partially thrombosed arterial aneurysm, magnetic resonance imaging (MRI) was performed. MRI revealed a small aneurysm in the same area; however, the aneurysm seemed to be located outside of the arterial tree (Fig. 1f). High-resolution FIESTA sequence supported the location (Fig. 2a). This led to a second CTA in the late venous phase. The venous-phase CTA suggested that the aneurysm was related to the venous system (Fig. 2b and c). Seven days after admission, a second DSA was performed, which indicated a small venous aneurysm belonging to the transverse pontine vein (close to the SCA) as the most plausible source of the patient's pSAH (Fig. 2d, f, and e).

Post-discharge course and follow-up

The patient was transferred back to a regional step-down hospital after 3 weeks. At that time, she had recovered well, with only minor headache and slight disorientation. The patient's last DSA was performed at their post-discharge outpatient control visit (3 months after pSAH) at which time the angiography revealed a normal wall vascular structure (Fig. 3). The patient recovered well and was neurologically intact with some minor headaches.

Discussion

We report a rare case of pSAH with a positive image finding, indicating a ruptured venous aneurysm as the source of the bleeding. Intracranial venous aneurysms most often represent a vascular anomaly, hence, coexisting with other vascular anomalies such as arteriovenous malformations (AVMs), dural arteriovenous fistulas (DAVFs), vein of Galen malformations, or cavernomas [18, 22, 24, 28, 29]. In isolation, without any coexisting vascular anomaly, venous aneurysms are considered rare. An external cause of isolated venous aneurysms is head injuries, although these venous aneurysms are most often located at the brain surface and not deeper in the venous system as in our case [12].

To the best of our knowledge, this is the first report on a saccular intracerebral venous ruptured aneurysm causing pSAH. Van et al. published their first 13 pSAH cases in 1985, with unexplained negative angiographic findings. Capillary or venous sources were considered the most likely etiologies of

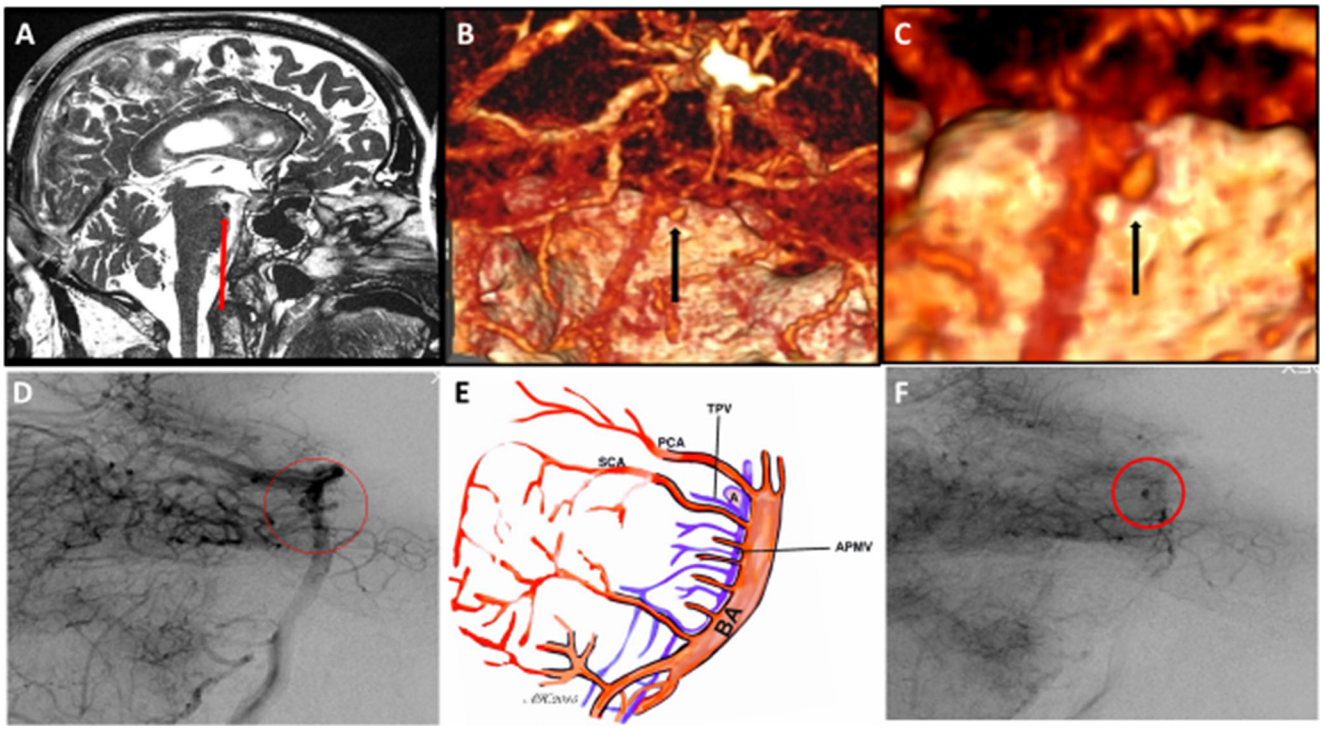

Fig. 2 Fast imaging employing steady-state acquisition (FIESTA) MRI (**a**) sagittal cut shows an aneurysm (*red arrow*) isolated from the arterial tree. CT venography 3D reconstruction of the perimesencephalic veins (**b**, **c**) shows the aneurysm (*black arrows*) located at the right transverse pontine vein after bifurcation from the anterior pontine mesencephalic vein. Late arterial phase DSA 7 days following admission (**d**) clearly shows a small aneurysm (*inside red circle*) not connected to the basilar artery or one of its branches. The accompanying diagram (**e**) displays how this aneurysm of the transverse pontine vein (TPV), which is a branch of the anterior pontine mesencephalic vein (APMV), clearly overlaps with the superior cerebellar artery (SCA) in the anterior posterior view. Very late arterial, almost total venous phase DSA (**f**) shows the aneurysm (*inside red circle*) filling well in venous time

this benign elusive disease [30]. In contrast, Alexander et al., who studied 140 cases of angiography-negative SAHs, suggested ventriculostriate and thalamoperforating vessels as the source of the bleeding [2]. Recently, Aqqawal et al. [1] presented a case with a clipped cervicomedullary junction ruptured venous aneurysm. Extra-cranial, however, true venous aneurysm is often encountered [4, 11].

The pathogenesis of intracranial venous bleeding involves many actions. The process mainly combines tissue hypoxia and physiological shunt, which lead to aberrant angiogenesis after long-standing venous hypertension [19].

The clinical course and outcome of patients with pSAH have recently been extensively investigated [8, 13, 15]. Most previous studies have found that non-aneurysmal SAH with a perimesencephalic pattern of bleeding has a less aggressive course than an aneurysmal pattern of bleeding, and patient outcome is better [3, 5, 6, 8]. However, identification of a plausible etiology remains challenging. In the literature, pSAH is

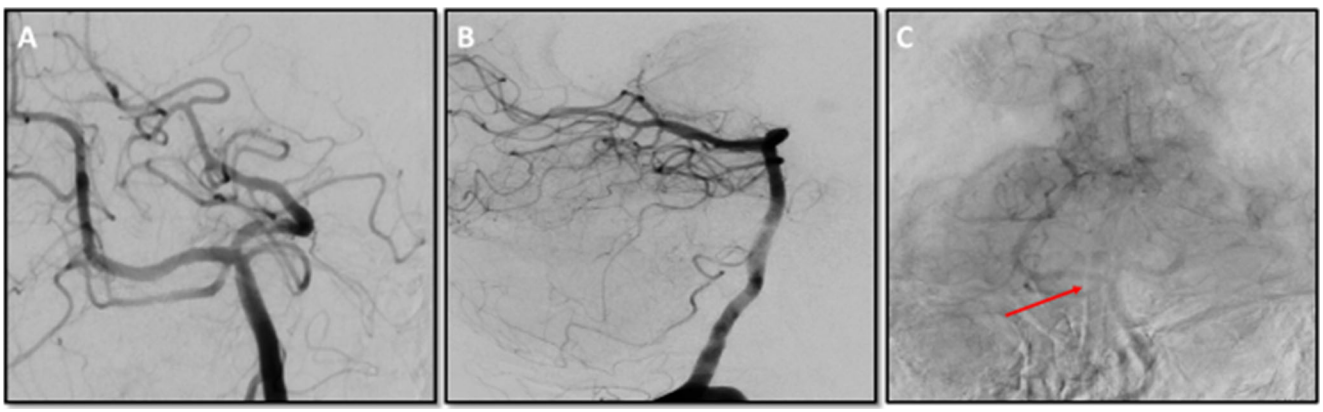

Fig. 3 DSA 3 months after presentation as viewed anteriorly (**a**) and laterally (**b**). No aneurysm is seen, and even in late arterial stage (**c**) no pathology is apparent in the area in which the venous aneurysm was observed (*red arrow*)

described in many ways, e.g., pSAH of unknown origin or etiology, non-aneurysmal SAH, negative angiography SAH, idiopathic pSAH, and benign pSAH [3, 6, 8, 9, 13, 15]. Noteworthy is that despite no identifiable source of the bleeding, some "negative angiography SAHs" re-bleed and markedly worsen patient outcome [8, 21]. It is in these re-bleeding cases that identification of a bleeding source is of crucial importance. Recent developments in radiological techniques (e.g., high-contrast and 3D angiography reconstructions) now offer improved chances of identifying small lesions responsible for pSAH. The next step after confirming a negative arterial phase DSA in a patient with SAH should be another angiography in late venous phase to rule out AVMs and DAVFs as well as rare etiologies such as venous pouch and saccular venous aneurysm.

Known risk factors associated with SAH are female sex, high blood pressure, smoking, elevated cholesterol levels, diabetes, alcoholism, drug abuse, and Finnish heritage [8, 16, 17]. Our patient is a 66-year-old Finnish female suffering from high blood pressure, diabetes mellitus type II, and high cholesterol. She had no known history of tobacco smoking, alcohol abuse, or head trauma. She presented with a typical pSAH distribution on the primary head CT. The initial CTA and DSA were both negative for any arterial aneurysms and other cerebrovascular malformations. The first DSA and a second late venous phase CTA revealed a saccular venous aneurysm in the area of the superior cerebellar artery as a plausible bleeding source.

Our case demonstrates that in the case of pSAH a ruptured saccular venous aneurysm is a potential etiology. Other differential diagnoses that should be considered are partially thrombosed aneurysms, capillary fistulas, and basilar perforator aneurysms [10].

The decision on how to treat ruptured saccular venous aneurysms is challenging, with little evidence to support different approaches. Naturally, all treatment decisions should also be made in favor of the patient. The prognosis of patients with arterial aneurysmal SAH has been extensively studied, but prognosis after venous SAH remains obscure. Our patient recovered well. She recovered to an independent state, being completely neurologically intact. This is, however, only one case and does not generalize to all patients with venous aneurysm SAH or other possible etiologies of pSAH.

Patients with pSAH of unknown etiology are primarily treated conservatively [7, 8, 14, 25–27]. However, SAH complications, such as acute and chronic hydrocephalus, may occur, often requiring surgical interventions, including temporary external ventricular drainage and cerebrospinal fluid shunting. The most feared complications of SAH (apart from re-bleeding) are vasospasm and delayed cerebral ischemia [8, 23, 27]. These complications occur in pSAH, although less frequently than in arterial aneurysmal SAH [8]. The underlying reasons remain unknown. In our institution, we treat pSAH and arterial aneurysmal SAH patients initially similarly and will continue to do so until further evidence regarding the treatment of non-arterial aneurysmal SAH or venous aneurysmal SAH emerges. A question that every treating neurosurgeon should consider is when to stop imaging. If, for example, the first two angiographies are negative, whom does the control angiography serve—the patient or the curious neurosurgeon?

Conclusions

We report a case report of a patient treated for perimesencephalic SAH with a positive angiographic finding suggesting a ruptured saccular venous aneurysm originating from the lateral pontine vein. Our case suggests that venous aneurysms might be a plausible source of pSAH of unknown origin. Thus, careful analysis of "negative angiographies" should be done to identify this uncommon etiology. Multicenter collaborations are required to elucidate the optimal diagnostic strategy and management of these rare ruptured saccular venous aneurysms.

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Compliance with ethical standards

Informed consent Informed consent was obtained from the patient.

Conflict of interest All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers, bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patient-licensing agreement), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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