Non-Aneurysmal Subarachnoid Hemorrhage

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ABSTRACT

The majority of spontaneous subarachnoid hemorrhages arise from the rupture of saccular aneurysms. The concept of a non-aneurysmal form of subarachnoid hemorrhage with a distinct radiographic appearance was first presented by van Gijn and colleagues in 1985. The differential diagnoses of this type of hemorrhage include Non-aneurysmal perimesencephalic subarachnoid hemorrhage, arterial dissection, cerebral arteriovenous malformations (AVMs), dural arteriovenous fistulae, cervical AVMs, saccular aneurysms of spinal arteries, mycotic aneurysms, pituitary apoplexy and use of anticoagulants. In this review, we discuss the different causes of non-aneurysmal subarachnoid hemorrhages.

Keywords: Non-aneurysmal subarachnoid hemorrhage; Subarachnoid hemorrhage; Perimesencephalic subarachnoid hemorrhage

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INTRODUCTION

Approximately 85% of all spontaneous subarachnoid hemorrhages (SAH) arise from the rupture of saccular aneurysms [1, 2]. The term non-aneurysmal SAH with a distinct radiographic appearance was first coined by van Gijn and colleagues in 1985 [2]. Of the 15% of SAHs not attributable to saccular aneurysms, two-thirds (10% of the total) are caused by non-aneurysmal SAH and the remaining 5% by a variety of rare conditions (Table 1) [3]. A literature search with the indexing term "non-aneurysmal subarachnoid hemorrhage" in MEDLINE and Google Scholar was performed, the literature was reviewed, and different causes of non-aneurysmal were evaluated.

PERIMESENCEPHALIC NONANEURYSMAL HEMORRHAGE (PMSAH)

Perimesencephalic nonaneurysmal hemorrhage (PMSAH) is characterized by accumulation of subarachnoid blood predominantly around the midbrain and absence of an aneurysm or other source of bleeding on angiography [10, 11]. PMSAH was first described by van Gijn et al in 1985 [4]. Perimesencephalic hemorrhage constitutes 10% of all episodes of spontaneous SAH and two thirds of those with a normal angiogram [1, 3, 5]. PMSAH appears to have a distinct etiology and natural history than aneurysm rupture, with good clinical outcomes and minimal risk of rebleeding [1, 6]. PMSAH patients are younger and less likely to be female or hypertensive than other SAH patients, with a trend towards fewer smokers among the PMSAH group. A thorough investigation of PMSAH risk factors is limited by a small number of patients within this category [1, 7].

CLINICAL PRESENTATION

Clinically, there is little to distinguish idiopathic perimesencephalic hemorrhage from aneurysmal hemorrhage. The headache onset is more often gradual (minutes rather than seconds) than with aneurysmal hemorrhage, but the predictive value of this feature is poor [4, 8]. Loss of consciousness and focal symptoms are exceptional and then only transient; a seizure at onset virtually rules out the diagnosis [8]. On admission, all patients are in perfect clinical condition, apart from their headache [4, 8]. Transient amnesia is found in about one-third and is associated with enlargement of the temporal horns on the initial CT scan [9].

RADIOLOGICAL FEATURES

The radiographic pattern of PMSAH is relatively distinct, with hemorrhage centered anterior to the midbrain or pons, with or without extension of

Table 1: Causes of subarachnoid hemorrhage

1.	Ruptured aneurysm (85%)
2.	Non-aneurysmal perimesencephalic hemorrhage (10%)
3.	Rare conditions (5%)
	a) Arterial dissection (transmural)
	b) Cerebral arteriovenous malformation
	c) Dural arteriovenous fistula
	d) Vascular lesions around the spinal cord
	e) Septic aneurysm
	f) Pituitary apoplexy
	g) Cocaine abuse
	h) Trauma (without contusion)

blood around the brainstem, into the suprasellar cistern, or into the proximal Sylvian fissures [6, 12, 13]. There may be some sedimentation of blood in the posterior horns of the lateral ventricles, but frank intraventricular hemorrhage or extension of the hemorrhage into the brain parenchyma indicates arterial hemorrhage and rules out this particular condition [10]. A posterior variant of PMSAH with hemorrhage primarily in the quadrigeminal cistern has also been described [14]. Because aneurysm rupture occasionally produces a PMSAH-like pattern of bleeding, diagnostic cerebral angiography continues to be recommended for these patients [12].

CLINICAL COURSE

Typically, the early course is uneventful; rebleeds and delayed cerebral ischemias do not occur. Approximately 20% of patients have enlarged lateral ventricles on their admission brain CT scan, associated with extravasation of blood in all perimesencephalic cisterns, which probably causes blockage of the CSF circulation at the tentorial hiatus [15]. Only few have symptoms from this ventricular dilatation and even then an excellent outcome can be anticipated [10, 16]. The period of convalescence is short and almost invariably patients are able to resume their previous work and other activities [10, 17].

Arterial dissection: It is unknown precisely what proportion of all SAH arises from a dissected vertebral artery [18]. Dissection is more often recognized in the carotid than in the vertebral artery, but SAH from a dissected artery occurs mostly in the vertebral artery [19, 20]. Neurological deficits that may accompany SAH from vertebral artery dissection are palsies of the ninth and tenth cranial nerves by subadventitial dissection [21] or Wallenberg's syndrome [22]. Rebleeds occur in between 30 and 70% of cases [22, 23]. Dissection of the intracranial portion of the internal carotid artery or one of its branches as a cause of SAH is much less common than with the vertebral artery. Reported cases have affected the terminal portion of the internal carotid artery [24, 25], the middle cerebral artery [26] and the anterior cerebral artery [27].

Cerebral arteriovenous malformations (AVMs)*:* Non-aneurysmal subarachnoid bleeding at the convexity of the brain may occur from superficial AVMs. In 5% of all ruptured AVMs there is extravasation only in the subarachnoid space, without intracerebral hematoma [28]. These superficial lesions can be visualized on computerized tomography (CT) scan.

Dural arteriovenous fistulae*:* Dural arteriovenous fistulae of the tentorium can give rise to a basal hemorrhage that is indistinguishable on CT from aneurysmal hemorrhage [29, 30]. The anomaly is rare and can be found from adolescence to old age and after a first rupture, rebleeding may occur [31]. In majority of the patients there may be a history of skull fracture.

Cervical AVMs: Spinal AVMs present with SAH in ~10% of cases; in 50% of these patients, the first hemorrhage occurs before the age of 20 years [32, 33]. Clues pointing to a cervical origin of the hemorrhage are onset with a sudden and excruciating pain in the lower part of the neck, or pain radiating from the neck to the shoulders or arms [34]. In the absence of such symptoms, the true origin of the hemorrhage emerges only when spinal cord dysfunction develops, after a delay that may be as short as a few hours or as long as a few years [33, 35]. Rebleeds may occur, even repeatedly [36]. CT scanning of the brain in patients with a ruptured cervical AVM may show blood throughout the basal cisterns and ventricles [34]. If a cervical origin of the hemorrhage is suspected, magnetic resonance imaging (MRI) or magnetic resonance angiography (MRA) angiography are the first line of investigation, because spinal angiography is impractical without localizing signs or symptoms.

Saccular aneurysms of spinal arteries: As with AVMs of the spinal cord, the clinical features of spinal SAH may be accompanied by those of a

transverse lesion of the cord, either partial or complete. Patients with these lesions present with pain in the lower part of neck or in the back with radicular pain and/or cord deficit, with blood mainly seen in the basal cisterns. Saccular aneurysms of spinal arteries are extremely rare, with recorded incidents in ~12 patients [37, 38].

Anticoagulants: Anticoagulant drugs are seldom the sole cause of SAH [39]. Severe coagulopathy other than by anticoagulant drugs, e.g. congenital deficiency of factor VII, is also a rare cause of hemorrhage confined to the subarachnoid space [40]. If aneurysmal hemorrhage occurs in a patient on anticoagulants, the outcome is relatively poor [41].

Sickle cell disease: Thirty per cent of patients with sickle cell disease and SAH are children. CT scans in these children show blood in the superficial cortical sulci; angiograms show no aneurysm, but often show multiple distal branch occlusions and a leptomeningeal collateral circulation. The SAH is attributed to rupture of these collaterals [42].

DIFFERENTIAL DIAGNOSIS

If angiography is negative, it is essential to take account of the pattern of hemorrhage on the initial CT scan. If this pattern is perimesencephalic, the diagnosis of non-aneurysmal hemorrhage is established and no repeated studies are needed given the absence of rebleeds and the invariably good outcome. Such patients need no longer be on an intensive or medium care unit and can be transferred to a regular ward [43, 44]. A perimesencephalic pattern of hemorrhage may occasionally (in 2.5–5% of cases) be caused by rupture of a posterior fossa aneurysm [10, 45]. A formal decision analysis based on these observations indicates that a strategy where CT angiography is performed and not followed by conventional angiography, if negative, results in a better utility than a strategy where CT angiography is always followed by conventional angiography or if all patients are initially investigated by conventional angiography [43-45].

CONCLUSION

Non-aneurysmal SAH is a well-recognized entity and can be due to various etiologies. In patients with a negative angiogram for saccular aneurysms, one should keep in mind all these possibilities for appropriate management.

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