Non-Aneurysmal Perimesencephalic Subarachnoid Haemorrhage with Associated Pontine Haemorrhagic Infarction

A Case Report and Subject Review

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Summary

Non-aneurysmal perimesencephalic subarachnoid haemorrhage is characterized by an accumulation of blood in the perimesencephalic and prepontine cisterns identified on sectional imaging together with persistently negative cerebral angiography. Magnetic resonance imaging usually contributes no further information on the possible cause of the bleed but may occasionally show further features including associated parenchymal infarcts such as the pontine haemorrhagic infarct seen in the case described here.

Introduction

Approximately 85% of cases of acute subarachnoid haemorrhage (SAH) are due to rupture of an intradural cerebral aneurysm. There is a small number of other recognized pathologies of the brain and spine that can also result in acute SAH, amounting to a further 5% of cases. In the remaining 10% of cases of acute SAH, the haemorrhage appears confined to the perimesencephalic and prepontine cisterns on CT and typically there is an associated negative angiogram. MR imaging usually fails to contribute any further significant information in these cases. This subset of SAH patients tends to follow a benign clinical course and generally has a very good outcome. This clinico-radiolo-

gical entity has been termed non-aneurysmal perimesencephalic subarachnoid haemorrhage (NPSAH). We describe a single illustrative case that had evidence on MR imaging of an associated pontine haemorrhagic infarction and yet still had a very good clinical outcome, and review the history and features of this condition.

Case report:

A 55-year-old man presented with an acute severe headache and truncal ataxia at a peripheral hospital. Three months prior to this presentation he had been treated with thrombolytic agents for an acute occlusion of a femoro-distal bypass graft, and had subsequently been placed on oral warfarin therapy. He was mildly hypertensive which was well controlled with a single ACE inhibitor agent.

An unenhanced computed tomographic scan performed at this stage showed acute focal subarachnoid haemorrhage with the blood localised within the perimesencephalic and prepontine cisterns. Furthermore there was an area of diminished radiodensity seen in the adjacent pons together with smaller focal hyperintense areas indicating both oedema or infarction and haemorrhage within the substance of the pons (figure 1 A). The patient was then transferred to our institution for angiography and further management. A magnetic resonance (MR) scan

Figure 1 A) Axial unenhanced CT scan cut showing moderately dense clot in the prepontine and perimesencephalic cisterns with some hyperintensity within the truncus itself as well.

with diffusion imaging was performed upon arrival which again showed the presence of clot in the perimesencephalic and prepontine cisterns together with high signal intensities in the upper pons on T1-weighted images and high signal areas within the upper pons and lower mesencephalon on T2-weighted images (figures 1 B, C). A bright signal was seen within the same area on diffusion imaging. No abnormal signal was seen within the basilar artery to suggest dissection with intramural thrombus. At this stage, he was found clinically to have a left-sided intranuclear ophthalmoplegia and dysarthria and left-sided cerebellar ataxia.

Cerebral angiography was performed which showed slight circumferential narrowing of the mid to distal basilar artery only. No aneu-

rysm was demonstrated. The basilar narrowing was thought initially to be due to vasospasm. The rest of the cerebral circulation appeared normal.

He was managed medically with intravenous fluids and piracetam (Nootropil, UCB), and recovered remarkably rapidly from his original presenting neurological deficits.

Repeat cerebral angiography was performed six weeks later. The angiographic appearance of the basilar artery and its branches was identical to that seen during the initial angiogram. Again no aneurysm or other cerebrovascular abnormality was demonstrated. He was discharged on oral antiplatelet agents in the place of warfarin for the peripheral vascular disease.

Figure 1 B) Sagittal T1 weighted midline MR image showing the pre-truncal clot and high signal intensity haemorrhage within the truncus.

Figure 1 C) Axial T2 weighted MR image showing the extensive high signal intensity within the truncus. No evidence of basilar arterial dissection was seen on any sequence.

Discussion

Between 15 and 20% of cerebral angiograms performed on patients presenting with acute subarachnoid haemorrhage (SAH)

will show no evidence of any aneurysm or other causative lesion¹. In a number of these, aneurysms may still be present as the cause for the haemorrhage but may not be visualised for a number of reasons including an inadequate number of radiographic views, movement artefact in a restless patient, misinterpretation of vessel loops or infundibula, spontaneous thrombosis of the aneurysm, or reduced arterial flow due to severe vasospasm². For cases with an initial negative angiogram, repeat angiography is advocated two to six weeks after the initial haemorrhage. This has a variable positive yield of between 2% and 22%^{3,4,5}. Some authors have even advocated a third angiogram at between one and six months if the second is negative⁶, whereas others regard this as superfluous⁷. However, provided that the initial angiogram is technically good and properly evaluated the rate of false-negative findings can be reduced to below 2%^{4,8,9}. Even with a negative angiogram some neurosurgeons would be prepared to explore a region of the brain looking for an aneurysm based upon the pattern of localization of blood in the subarachnoid space on CT. Seven out of nine

Figure 2 Right (A) and Left (B) vertebral arterial digital subtraction angiograms at the time of presentation showing narrowing of the distal basilar artery which was originally thought to represent vasospasm.

patients thus explored surgically by Tatter, et al, showed three lesions with small aneurysm sacs # 4 mm in size that were clipped and four micro-aneurysms too small for clipping¹⁰. Other groups have also reported aneurysms found at exploratory surgery that were missed on initial angiographic studies¹¹.

There are other non-aneurysmal causes of acute SAH including trauma, dural arteriovenous fistula, spinal tumour or vascular malformation, vasculitis, coagulopathy, cocaine abuse, sickle cell disease, pituitary apoplexy, intracranial arterial dissection and intracranial neoplasia or infarction^{1,2,12,13}.

Van Gijn and co-workers were the first to describe a particular subgroup of patients who presented with an acute SAH with the clot seen on computed tomography (CT) confined to the perimesencephalic cisterns and adjacent subarachnoid spaces.

All of the 13 patients in this series had negative cerebral angiograms and followed a benign clinical course with excellent outcome¹⁴.

They coined the term “non-aneurysmal perimesencephalic subarachnoid haemorrhage”. Since this first description, other authors have independently confirmed this as a distinct entity with consistent imaging findings and a universally benign clinical course and good outcome^{15,16}.

The CT pattern typically shows blood localized to the interpeduncular, prepontine and ambient cisterns as well as the basal Sylvian fissures. No blood is seen in the lateral Sylvian or interhemispheric fissures and rarely within the ventricles^{17,18}.

The clots themselves are usually mildly hyperintense. This perimesencephalic CT pattern is not pathognomonic, however, for this condition as it can be seen in between 1% and 10% of ruptured posterior fossa aneurysms^{19,20}.

This means that although the chances of finding an aneurysm at angiography are small in patients with the typical perimesencephalic CT pattern, cerebral angiography is still indicated^{11,13,19}. Between 30% and 68% of cases with negative cerebral angiograms will show blood in the perimesencephalic distribution on CT scans^{3,5,12,21}.

Non-aneurysmal perimesencephalic SAH (NPSAH) has thus been estimated to account for around 10% of all cases of SAH²².

Schievink, et al, coined the term “pretruncal non-aneurysmal SAH” in 1997 as they realised that the pattern of cisternal blood or clot accumulation was, in fact, sited in front of the brain stem (truncus cerebri) in the interpeduncular and prepontine cisterns and around the basilar artery and not simply around the mesencephalon²³.

Figure 3 Right (A) and Left (B) vertebral digital subtraction angiograms done over two months later showing an identical narrowed appearance to the basilar artery to that seen on the original angiogram.

Yet another variant of PNSAH occurs with accumulation of blood mainly in the quadrigeminal cistern with no pretruncal blood accumulation. These patients also seem to run an uncomplicated clinical course²⁴.

Complications related to the blood in the subarachnoid space such as vasospasm and hydrocephalus do occur but tend to be uncommon and generally mild in effect. Vasospasm occurs in between 3% to 20% of patients with PNSAH^{21,25,26}.

It is only seen in the posterior circulation. It is usually mild and focal probably due to the relatively small blood load in the subarachnoid space, but occasionally can be diffuse and severe, although such cases still seem to have a good outcome clinically²⁷.

Hydrocephalus can develop in a number of cases of PNSAH but it is also usually mild and tends to resolve spontaneously. Rarely it may be severe enough to necessitate shunt placement^{25,28}.

Although originally not felt to be a feature of PNSAH, intraventricular blood can be found on CT in some cases and its presence may be a good indicator for the development of acute hydrocephalus as well as a possible indicator for a higher complication rate and poorer outcome¹⁸. The risk of rebleeding in PNSAH appears to be very small^{11,15,16,25,29,30}.

In general, the short and long term clinical outcome in PNSAH is very good. Most patients seem to be discharged in good clinical condition with little or no long-term neurological deficits^{13,14,15,25,29,30,31,32}. A number of more recent studies have shown that patients with PNSAH can suffer considerable long term psychological sequelae including depression, irritability, tiredness and diminished endurance, as well as a higher incidence of minor cognitive dysfunction and headaches^{15,29,33}. Although PNSAH has become such a distinct entity with its own imaging features and clinical course, the cause of this condition still remains largely speculative as few patients ever seem to reach surgery or post mortem. The pathological mechanisms proposed include vertebrobasilar dissection, rupture of a traversing mesencephalic or pontine perforating artery, rupture of a prepontine vein or bleeding from a small subpial vascular malformation^{2,34,35,36,37}. As with our case, anticoagulation may also predispose to PNSAH³⁸. Hypertension has also been shown to be an independent risk factor for PNSAH³⁹.

The role of magnetic resonance imaging appears to be controversial in the setting of PNSAH. As the localisation of the blood clot in the pretruncal region has usually already been made on CT, MR imaging has been shown to add little further information concerning the

source of the bleeding^{17,40}. One important exception to this has been the recently described finding of acute perforator territory infarcts involving the caudate, putamen or thalamus in a number of patients with PNSAH^{36,41}. The theory associated with this finding is that perforator arterial thrombosis and infarction can lead secondarily to rupture of the affected artery as it traverses the subarachnoid space. There is at least one autopsy report published 11 years before the entity of PNSAH was described that describes a case of fatal SAH due to rupture of a small perforator artery arising from the basilar artery. This ruptured vessel was shown to have a medial deficiency suggesting that rupture of these perforating arteries may be dependant upon an intrinsic vessel wall weakness.

Ongoing improvements in imaging technology have led to the widespread availability of high-resolution CT angiography (CTA). Some authors are already advocating the use of spiral CTA as the follow-up investigation of choice after a negative initial diagnostic cerebral angiogram⁷. CTA has been shown to detect aneurysms not seen out conventional angiography, particularly anterior communicating and posterior fossa aneurysms. Its role in the setting of PNSAH is primarily for the exclusion of a vertebrobasilar aneurysm. Ruigrok, et al, suggests that conventional angiography might be omitted in patients with PNSAH and a negative vertebrobasilar CTA⁴².

One other new imaging modality not yet mentioned with respect to PNSAH but which offers a potentially greater positive yield for angiographically occult aneurysms is 3D rotational digital subtraction angiography. This provides three-dimensional angiographic images of higher definition than either CT or MR angiography⁴³.

In the typical clinical and radiological setting of PNSAH, many authors concur that second or further follow-up cerebral angiograms are probably unnecessary^{3,12,16,30,31}.

Where equivocal or non perimesencephalic SAH patterns exist then repeat angiography or, where available, high quality CTA, is indicated. The timing of a repeat study is probably best delayed until after the period of expected vasospasm, usually two to six weeks after the initial bleed.

As mentioned above, some authors feel that MR imaging is probably of little benefit in ca-

ses of typical PNSAH (40). This may be true given a retrospective analysis of cases in a series but many cases with access to MR imaging will undoubtedly still undergo MR examination looking for angiographically or CT occult vascular or other lesions. MR has shown cases of associated perforator territory infarction, and given that one of the possible causes of PNSAH is vertebrobasilar dissection, the presence of intramural haematoma may be shown on high-resolution T1 weighted MR images. MR imaging of the spine would be indicated where there is clinical suspicion of a spinal lesion⁴⁰.

Given all of the facts reviewed above, we feel that our case represents an example of PNSAH in that the CT scan showed concentration of blood of moderate hyperintensity in the pretruncal cisterns and two cerebral digital subtraction angiograms were negative except for the unexplained narrowed appearance of the basilar artery. This we initially thought to be due to vasospasm although the basilar calibre remained more or less constant on the second angiogram for which we have no definite explanation. One other possible cause for this narrowed appearance could have been basilar arterial dissection³⁴ although the MR scan obtained did not show intramural haematoma within the basilar artery. A further finding in our case is the presence of haemorrhage and oedema within the substance of the pons. The larger area of high signal intensity within the pons on the T2 weighted images indicates a more extensive area of oedema and infarction as confirmed by the positive diffusion scan. The extent of this oedema was out of keeping with the degree of neurological deficit sustained. Furthermore, there was a relatively rapid and complete recovery from this event. These features are possibly more in keeping with a venous infarction rather than an arterial haemorrhage. As with most cases of PNSAH, however, the exact cause of the SAH remains speculative, and the postulated mechanisms must still include perforator haemorrhage or thrombosis or spasm, venous rupture and infarction, or limited basilar dissection. The use of oral anticoagulants probably also contributed to the occurrence of this haemorrhage but it is surprising that it had not occurred during the earlier thrombolytic therapy when the risk of bleeding was potentially greater. Hypertension may also have been a

contributing factor. Our patient has done very well clinically and has been discharged with very little residual neurological deficit.

Perimesencephalic or pretruncal non-aneurysmal subarachnoid haemorrhage thus indicates a distinct subcategory of acute subarachnoid haemorrhage having a typical pattern of blood distribution on CT with associated negative cerebral angiography, a benign clinical course and very good overall outcome. Reco-

gnition of this condition from onset may help to avoid unnecessary risk and expense due to repeat angiography. The role of MR imaging remains controversial although this modality may help to identify cases with associated deep cerebral or other perforator territory infarction or vertebral dissection. Our case shows that PN-SAH may also be associated with brainstem infarction but still seems to run the usual benign clinical course associated with this condition.

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