Cerebral aneurysms and subarachnoid haemorrhage

Avoiding the diagnostic pitfalls

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Patients with atypical or mild symptoms of subarachnoid haemorrhage due to a ruptured cerebral aneurysm are more difficult to diagnose than patients who present with typical symptoms of sudden severe headache and associated symptoms. Patients without typical symptoms are more likely to have low volume haemorrhages that may not be seen on a CT scan.

Headache, a common presentation to primary care physicians, usually has a benign cause and course but can occasionally be a symptom of a neurological condition requiring urgent neurosurgical treatment. Secondary headache (headache due to an underlying pathological cause, including structural, vascular, infective, inflammatory or drug-induced) therefore always needs to be considered in patients with new, severe or persistent headache or atypical presentations of headache.1,2

Patients presenting with sudden severe ‘thunderclap’ headache, with or without associated symptoms (which may include nausea and vomiting, loss of consciousness and a focal or global neurological deficit) clearly need immediate investigation for cerebral aneurysms and subarachnoid haemorrhage.

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with brain CT for probable subarachnoid haemorrhage (SAH) due to a ruptured cerebral aneurysm. However, patients with atypical or mild symptoms of SAH are more difficult to diagnose; these are often the patients with low volume haemorrhages that may not be seen on a CT scan. Further investigations are required in these patients to exclude SAH as the aetiology. As some of these investigations are invasive and involve risk (e.g. lumbar puncture and digital subtraction angiography), significant decisions and recommendations have to be made in patients who often have the mildest headache.\textsuperscript{2,5}

Ruptured cerebral aneurysms have a significantly different natural history to unruptured aneurysms. The re-rupture rate in the first two weeks after haemorrhage is 20\% (with mortality of 10\%), and in the first six months is 50\%.\textsuperscript{6,7} The annual rupture rate of a small (less than 10 mm) unruptured aneurysm is in the order of 1\% per year or less.\textsuperscript{8}

There are, of course, structural causes of headache and nontraumatic SAH other than a ruptured cerebral aneurysm, and these are generally discovered on further investigation.\textsuperscript{9,10} The critical issue is the diagnosis of the haemorrhage itself as a missed haemorrhage can have devastating effects for the patient.\textsuperscript{11}

The investigation and management options for patients with SAH due to a ruptured cerebral aneurysm are discussed in this article, and a case study of a low volume haemorrhage is presented to demonstrate some of the difficulties and complexities surrounding diagnosis (Box and Figures 1a and b). The presentation and management of unruptured cerebral aneurysms is also discussed.

KEY POINTS

- Subarachnoid haemorrhage (SAH) due to a ruptured cerebral aneurysm is a potentially fatal condition that often presents as headache.
- A raised index of suspicion must exist for all patients with sudden, severe headache with or without associated symptoms.
- Initial investigation of suspected SAH should be a noncontrast CT scan; most haemorrhages will be apparent on this test.
- Management of SAH is increasingly becoming endovascular.
- The outcome for patients with no or only a mild global or focal neurological deficit on presentation has improved, but prehospital mortality and disability from the initial haemorrhage remains a significant problem.
- Unruptured asymptomatic aneurysms are not an urgent scenario and can be dealt with in consultative outpatient fashion.
RUPTURED CEREBRAL ANEURYSM WITH LOW VOLUME HAEMORRHAGE

Case study
A 41-year-old woman who was previously well presented to her local hospital’s accident and emergency department with a sudden severe headache associated with nausea and vomiting but no loss of consciousness. She had a CT scan, as there was suspicion that this may represent subarachnoid haemorrhage (SAH). The CT scan was normal and the patient was offered a lumbar puncture as a further investigation. By then her headache was improving, and she declined the test and discharged herself against advice.

She presented to her GP over two weeks later with some mild, variable ongoing headache and some general malaise. The GP was concerned enough about the history given to arrange review by a neurosurgeon. Following this consultation, a CT angiogram (CTA) was performed, which showed a right carotid bifurcation aneurysm (Figure 1a). It had ruptured at such low volume as to be not seen on initial CT scan.

Following case conferencing between the endovascular surgeon and neurosurgeon, it was determined that the most effective treatment for this lesion was microsurgical repair. The anatomy of the aneurysm was such that simple coiling was not possible, and stenting/coiling was thought to be higher risk than microsurgical repair.

The woman was counselled that this aneurysm was likely to have ruptured and that her risk of re-rupture was high. It was felt that at this stage after the event lumbar puncture had little to offer in terms of definitively excluding haemorrhage.

She agreed to undergo microsurgical repair (Figure 1b). The surgery was uneventful, and she was discharged home within five days.

Investigations for SAH
CT scan
A noncontrast CT scan of the brain is the best and most widely available test for the investigation of severe headache. It is the most sensitive imaging modality for detecting blood, and will reveal blood in over 95% of patients with SAH, although the sensitivity decreases over time (sensitivity is 98 to 100% in the first 12 hours after SAH, reducing to 93% at 24 hours and to between 57 and 85% six days after SAH). Other methods to differentiate SAH and contaminated collection are examination on spectrophotometry of the supernatant from centrifuged CSF samples (presence of xanthochromia indicating SAH) and laboratory assessment of CSF samples for extracellular blood products such as bilirubin (detection of bilirubin indicating SAH).

The majority of patients with suspected SAH appear to have ‘traumatic taps’. Needless to say a clear result is comforting and as definitive as possible in the acute period after possible haemorrhage. CSF samples that are heavily contaminated with blood will often be problematic despite the above considerations.

CT angiography
CT angiography (CTA) is often used as an adjunct to CT and lumbar puncture, and can be a powerful tool for the non-invasive assessment of anomalies in the larger vessels of the circle of Willis. CT angiography (CTA) is often used as an adjunct to CT and lumbar puncture, and can be a powerful tool for the non-invasive assessment of anomalies in the larger vessels of the circle of Willis.

However, this assessment of vascular anatomy does not exclude that there has been haemorrhage.

The sensitivity of CTA is less than that of digital subtraction angiography.

MRI and magnetic resonance angiography
MRI is less sensitive than CT at finding acute haemorrhage and therefore is not recommended as a primary screening modality patients with suspected SAH. Magnetic resonance angiography (MRA) also, in general, has somewhat less anatomical resolution than CTA, although new techniques for optimising images and defining pathology are promising.

Digital subtraction angiography
Digital subtraction angiography (DSA) is the gold standard to assess vascular anatomy but has restricted availability and an attendant risk as it is an invasive test. It is uncommonly used to investigate patients with SAH, unless haemorrhage or vessel anomaly is seen on noninvasive tests.

Lumbar puncture
Lumbar puncture is the test required to exclude SAH when CT imaging is unhelpful. It is best performed more than 12 hours after SAH.

The results of lumbar puncture can be confounded by ‘traumatic taps’, where the cerebrospinal fluid (CSP) is stained by blood contamination during puncture. Traditionally there should be clearing of the blood staining if it is traumatic, and consistency over tube collection if SAH is present. Other methods to differentiate SAH and contaminated collection are examination on spectrophotometry of the supernatant from centrifuged CSF samples (presence of xanthochromia indicating SAH) and laboratory assessment of CSF samples for extracellular blood products such as bilirubin (detection of bilirubin indicating SAH).

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Nonhaemorrhagic presentation of cerebral aneurysms

Incidental finding

Most aneurysms are discovered incidentally in an outpatient setting. Increased access to noninvasive, relatively accurate imaging such as CTA and MRA has meant that the volume of incidental findings of cerebral aneurysm, which has a community prevalence of about 2%, is significant.\(^{23,24}\)

Several multicentre and prospective trials have attempted to determine the natural history of cerebral aneurysms and the morbidity of therapy.\(^{8,25-29}\) Unfortunately, the heterogeneity of aneurysms in terms of size, morphology and site, along with the moving target of microsurgical and endovascular morbidity, conspires to cloud risk–benefit analysis for repair.\(^{30}\)

In general terms, the larger the aneurysm, the greater the risk of haemorrhage.\(^{8,27}\) Certain sites, such as the posterior circulation and the posterior communicating artery (which is part of the circle of Willis), appear to have increased haemorrhage risk independent of the size of the aneurysm.\(^{8,27}\) Aneurysms with unfavourable morphology, such as daughter sacs, are also thought to be higher risk.\(^{27}\) It is generally agreed that the haemorrhage rate for small (less than 7 mm) aneurysms with no unfavourable features is less than 1% annually.\(^{27}\) Giant aneurysms (greater than 25 mm) have an adverse event rate of more than 2% annually.\(^{8,27,31}\)

Mass effect

Mass effect generally relates to large aneurysms or those in specific sites.\(^{22}\) Large or giant aneurysms may have a mass effect on neural structures, causing focal or global neurological deficits or seizures.\(^{2,33}\) Standard imaging will usually make this diagnosis, although other lesions may be mimicked – for example, thrombosis of an aneurysm can lead to acute swelling and embolus that may mimic a brain tumour on imaging.\(^{34}\)

Sites notorious for nerve deficit even with small or medium aneurysms are the posterior communicating artery, affecting the third cranial nerve (oculomotor nerve), and the anterior communicating artery, affecting the second cranial nerve (optic nerve) or chiasm.\(^{35}\) Compression of these or other nerves can cause acute or chronic deficit.

Acute cranial neuropathies related to aneurysms are an emergency in that they indicate a change in the aneurysm, which signals an increase in risk of haemorrhage.\(^{36}\) Differentiation of aneurysmal compression and diabetic mononeuropathy can be problematic (e.g. pupil involvement in aneurysmal compression), and is a classic pitfall.\(^{37}\)

Therapy and outcome

Outcome following SAH due to a ruptured cerebral aneurysm remains concerning despite advances in therapy.\(^{2}\) A rough guide is that after SAH, one-third of patients will be treated and recover ‘back to normal’, one-third will survive and be disabled, and one-third will not survive.\(^{2}\) Prehospital mortality is up to 15%, which is probably an underestimation as many causes of sudden death in the elderly may go uninvestigated.\(^{38}\)

The immediate treatment of patients presenting with haemorrhage involves basic resuscitation in poor grade patients (i.e. patients with a global or focal neurological deficit).\(^{39}\) A significant cause of ongoing coma following a SAH is hydrocephalus relating to the blood in the subarachnoid space. A patient with a decreased level of consciousness following a haemorrhage is not considered optimised until this pressure is relieved with external ventricular drainage. As an extension of this, no patient should be considered unsalvageable on the grounds of clinical condition without CSF diversion.\(^{40,41}\) Careful control of the patient’s blood pressure (by medication) is vital initially to decrease the risk of rebleeding.\(^{39}\)

Securing of the aneurysm by the most appropriate means should be achieved at the earliest opportunity, when the multidisciplinary team required can be assembled.\(^{42}\) The patient’s current use of anticoagulant and antiplatelet medication has to be dealt with as effectively as possible with the administration of procoagulants, blood products and platelets as appropriate. The presence of irreversible or significant bleeding diathesis may be a relative indication for endovascular treatment (i.e. coiling) rather than microvascular treatment (i.e. clipping).

Vasospasm is a significant complication of SAH and typically has onset five to 10 days after the bleeding event.\(^{43}\) Treatment of vasospasm includes pharmacotherapy with nimodipine and induction of hypertension and hypervolaemia, to increase cerebral blood flow.\(^{44,45}\) Endovascular angioplasty (chemical and balloon) is being used increasingly to manage this problem, with great benefit.\(^{45,46}\) A typical stay in an intensive care or high dependency unit of a patient with vasospasm can be more than two weeks due to the intensity of this therapy.

Patients with unruptured aneurysms that are being repaired microsurgically require the same presurgical assessment and management of antiplatelet or anticoagulant therapy as those undergoing any other major craniotomy. In those undergoing elective coiling, there is often no need to stop antiplatelet medication, and in those requiring complex repairs and/or stents, preloading with significant antipatelet medication prior to the procedure is often appropriate.\(^{47}\)

Endovascular therapy

The greatest change in the care of patients with this condition in recent times is the emergence of endovascular therapy as the dominant modality for treating ruptured cerebral aneurysms (Figure 2).\(^{29}\) The use of different embolic material and coils, along with stents and technical tricks of repair such as balloon-assisted coiling, have rapidly changed the deployment of endovascular therapy.\(^{48}\)

Dedicated neuroradiologists are now working in large centres and repairing the majority of ruptured aneurysms.\(^{49}\)
Microvascular repair

Microvascular repair (surgical aneurysm clipping) remains an important modality for lesions unfavourable for endovascular intervention. Unsuitability for coil repair often relates to the ratio of the fundus to the neck of the lesion in locations where the use of stent or balloon assistance for the coiling may be inappropriate or unsafe. The most common location where coiling is not favoured is the middle cerebral artery bifurcation.

Repair of a ruptured carotid bifurcation aneurysm is discussed in the case study (Box and Figures 1a and b).

Open surgery

Open surgery may also be used where there are associated surgical benefits, such as removal of intracerebral clot.

Treatments compared

In general, endovascular therapy has lower upfront risk than other treatments, and is without the associated morbidity of craniotomy (brain retraction, wound, access risk). It does, however, have less durability overall, and also a greater possibility of persistent/recurrent aneurysm necks (filling of the aneurysm where it pouches out from the vessel wall), which present an ongoing risk of haemorrhage, although the magnitude of that risk overall and in the previously ruptured versus the unruptured cohort is not well known. Securely coiled aneurysms that are stable after two years rarely re-rupture.

A relevant factor in stent-assisted repair of aneurysm is that the patient usually requires indefinite antiplatelet therapy. This and issues of durability will become more clear as our longitudinal experience with this therapy improves with time.

The ideal therapeutic scenario is where a balance and good working relationship exists between neuroradiologists and neurosurgeons. Care decisions should not be based on treatment availability in a condition such as this, and collaboration is vital; however, access limitations to endovascular care can influence intervention decisions.

In the presented case study, the anatomy of the aneurysm was such that simple coiling was not possible and stenting/coiling was thought to be higher risk than microsurgical repair.

Conclusion

Cerebral aneurysms remain a diagnostic and therapeutic challenge. Although SAH can be excluded in most patients with headache by simple history taking, in those where doubt remains (e.g. when the haemorrhage is low volume and does not show on CT imaging) the consequences of failure to diagnose need to be weighed against the inconvenience and possible morbidity of further investigation.

When an aneurysm has been diagnosed, the decision to treat and the mode of treatment to use has become multidisciplinary in a way analogous to that in many other medical conditions. This has led to better outcomes and the emergence of specialised neurovascular centres.

References

A list of references is included in the website version of this article (www.medicinetoday.com.au).

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Figure 2. Cerebral angiogram showing endovascular repair of two cerebral artery aneurysms, with metal coils in place (arrows).
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References