

Case 31

A cerebral vascular catastrophe

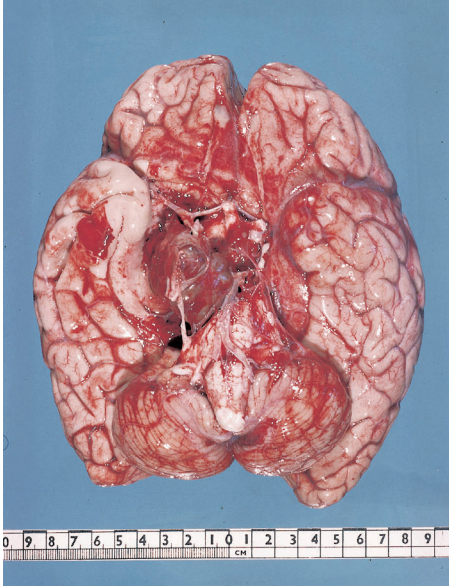


Figure 31.1

A previously healthy schoolmaster, aged 41 years, collapsed in his classroom. He was taken by ambulance to hospital, where he was found to be deeply unconscious, not responding to painful stimuli, breathing stertorously, and with marked neck stiffness. Shortly after arrival, he had respiratory arrest and was intubated and put on a ventilator. Four days later his brainstem reflexes were found to be absent and he was pronounced dead; following his premortem wishes he became a multiorgan donor.

Figure 31.1 shows the appearance of the inferior aspect of his brain at autopsy.

Describe the pathological findings revealed in this specimen

There is a large right-sided aneurysm of the circle of

Willis* which has ruptured. There is blood in the sub-arachnoid space.

Describe the circle of Willis; what is its importance?

The arterial circle of Willis is a rich anastomosis between the anterior, middle and posterior cerebral arteries on both sides. The posterior cerebral links to the middle via the posterior communicating artery, while the anterior cerebral links to its opposite via the anterior communicating artery. The middle and anterior cerebral arteries on each side are formed by the bifurcation of the internal carotid artery (Fig. 31.2).

Its importance is that it provides a highly effective anastomosis between the carotid system on either side. For example, in the healthy subject, ligation of the internal carotid artery can be performed without infarction of the brain on that side.

What is the commonest cause of this condition?

The great majority of circle of Willis aneurysms are congenital. About 20% are multiple and 85% occur on the anterior half of the circle. They may be associated with polycystic kidneys and with collagen diseases such as the Ehlers–Danlos syndrome.† Rarely, they result from arteriosclerosis, trauma or infection (mycotic aneurysm).

What is the natural history of the course of the untreated disease?

About a quarter of patients with haemorrhage from rupture of the aneurysm die without recovering consciousness, as in this case. Untreated, more than half will

*Thomas Willis (1621–1675), physician and anatomist, Oxford and then London.

†Edward Lauritz Ehlers (1863–1937), dermatologist, Copenhagen; Alexandre Danlos (1844–1912), dermatologist, Paris.

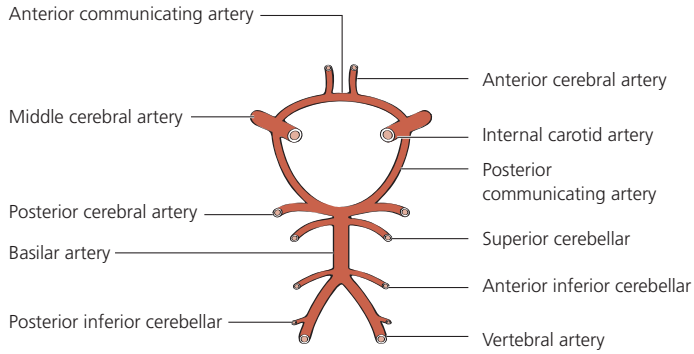
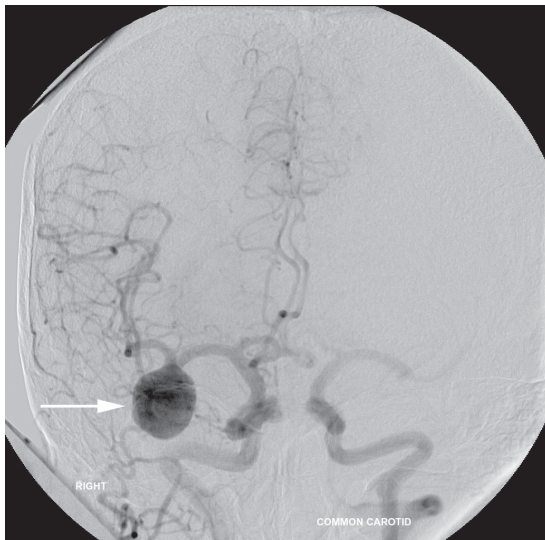
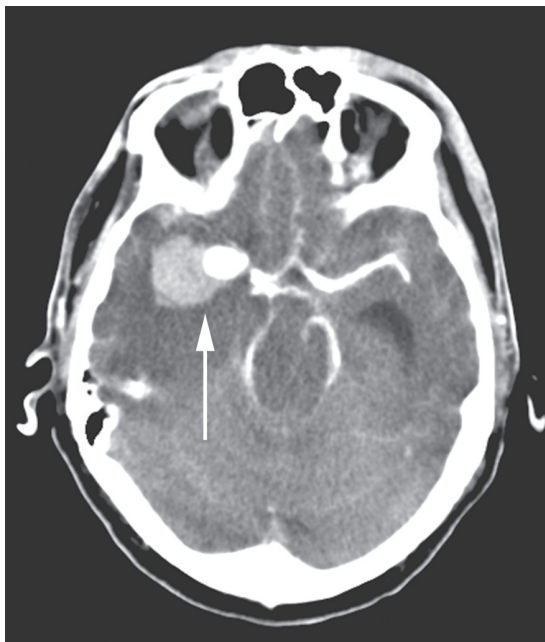


Figure 31.2 Anatomy of the circle of Willis.



(a)



(b)

Figure 31.3 Middle cerebral artery (MCA) aneurysm. (a) An angiogram illustrating a large right MCA aneurysm (arrowed). (b) This is also visible on the contrast-enhanced CT, which also shows a recent haemorrhage (light grey, arrowed), with surrounding oedema, some midline shift to the left, and obliteration of the right lateral ventricle.

bleed again within 6 weeks of the initial bleed and the mortality of this is high. After 6 weeks, the risk of a further bleed becomes less, but is still present.

Outline the management of this condition

When the patient is in coma or has a dense hemiplegia, nursing care only is indicated. If the patient recovers from the initial bleed, CT, MR or cerebral angiography are

performed to localize the aneurysm (Fig. 31.3). If an aneurysm is demonstrated, surgical treatment is indicated. This may be possible by percutaneous transarterial embolization or open operation with ligation or clipping of the neck of the aneurysm. About one-third of the angiograms are negative. These probably indicate that thrombosis has taken place in a small aneurysm. In such cases, the treatment is conservative and the prognosis is good.