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CASE REPORT

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Ruptured fusiform cerebral aneurysm in a neonate

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Abstract An 11-day-old male infant presented with subarachnoid and intraventricular haemorrhage caused by a ruptured intracranial aneurysm. Magnetic resonance (MR) angiogram and digital subtraction angiography (DSA) revealed a fusiform aneurysm 10 mm in diameter supplied by the proximal segment of the anterior cerebral artery (A1), with both distal segments (A2) arising from the aneurysm. The right A1 was aplastic. There have been 13 previous case reports of neonatal cerebral artery aneurysms, but only 1 of these has been fusiform. None of the earlier reports has mentioned dysplastic segments or other anomalies of the circle of Willis.

Key words Cerebral aneurysm · Neonate · Intracranial haemorrhage · Circle of Willis

Introduction

Ruptured neonatal intracranial arterial aneurysms are extremely rare. Only 13 such cases have previously been reported in the literature, all but 1 of which were of the saccular type. Fusiform aneurysms may occur in dysplastic vascular segments.

Case report

An 11-day-old male infant presented with a 2-day history of irritability, drowsiness, vomiting and reduced urine output. On examination, he was hypertonic with bulging anterior fontanelle, opisthotonus and brisk reflexes. A lumbar puncture revealed blood-stained CSF. Within 30 min he started gasping and displayed vertical nystagmus, poorly responsive pupils and reducing capillary return with central cyanosis. He required intubation and intermittent positive pressure ventilation. The infant was born at term with meconium-stained liquor. His mother had reported mild antepartum haemorrhage and reduced fetal movements at 33 weeks of pregnancy. His weight and head circumference at birth were 3.32 kg and 35 cm (36.5 cm on admission), respectively. He had two healthy siblings.

Cranial sonography followed by CT showed moderate hydrocephalus due to extensive subarachnoid and intraventricular haemorrhage, with a predominance of blood in the anterior interhemispheric fissure (Fig. 1). Within this, a rounded hypodense (relative to the haemorrhage) lesion was evident, highly suggestive of an aneurysm. This was confirmed by magnetic resonance imaging (MR) and digital subtraction angiography (DSA). The maximum intensity projection (MIP) of the 3D time-of-flight MR angiogram (Fig. 2) and DSA (Fig. 3) revealed a fusiform aneurysm 10 mm in diameter supplied by the A1 segment of the left anterior cerebral artery. The right A1 segment was completely aplastic, and both A2 segments arose from the anterior portion of the aneurysmal sac. There was moderately severe spasm of the left supraclinoid carotid artery and the anterior and middle cerebral arteries. Both carotid territories were anatomically otherwise normal.

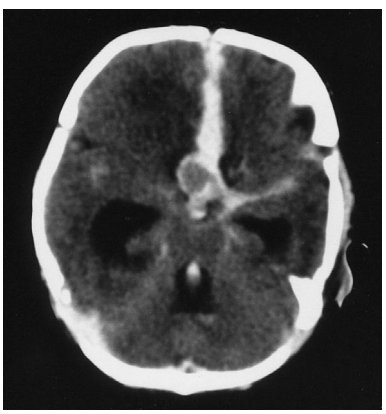

Fig. 1 CT scan shows extensive subarachnoid and intraventricular haemorrhage with a localised haematoma in the anterior interhemispheric fissure. The aneurysm is represented by an area of relative hypodensity within the haematoma

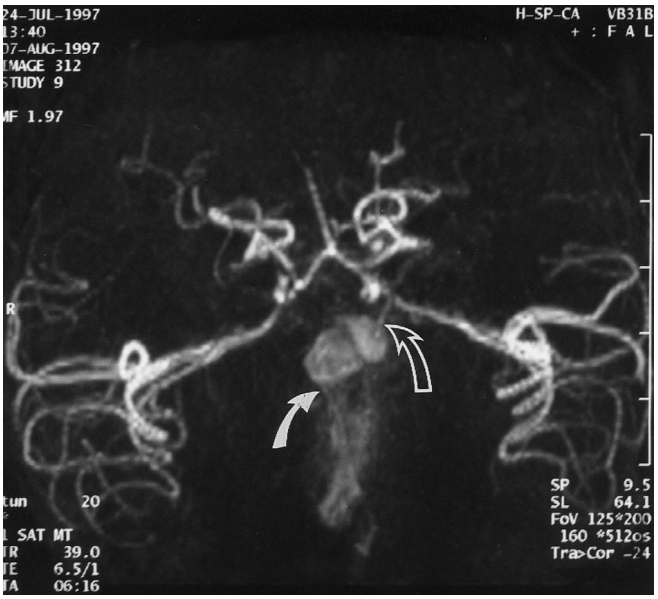

Fig. 2 MIP from an MR angiogram showing the A1 segment of the left anterior cerebral artery entering the aneurysm posterolaterally (*open arrow*) and both A2 segments arising from the aneurysm anteriorly (*solid arrow*). The right A1 segment is absent

A right frontal extracranial ventricular drain (EVD) was inserted to relieve the hydrocephalus demonstrated on CT. A sample of ventricular CSF taken during the procedure appeared xanthochromic, with a red cell count of 12,600/dl.

During the infant's stay in the intensive care unit there were persistent tongue movements, thought to be epileptic seizures. His EVD continuously drained blood-stained fluid, and regular transfusions were required. He was eventually weaned off the ventilator and was then extubated 7 days after the haemorrhage. He seemed to be fixing and focusing, with a reasonable sucking reflex, but dramatic headlag.

However, 3 days later, approximately 15 ml of blood leaked out of the EVD entry site, suggesting a rebleed. He remained haemodynamically stable, but demonstrated increased left lower limb tone. He was then taken to theatre for exploration.

Access was gained through a right frontal craniotomy, and a large subdural haematoma was immediately obvious. A massive brain infarct was apparent beneath that. Upon visualisation and exploration of the aneurysm and the area around it, the right internal carotid and anterior cerebral anteries were found to be in severe spasm. Attempts to clip the aneurysm and the feeding vessel were unsuccessful. During surgery, the child suffered a cardiac arrest and the operation was abandoned. A decision was made to opt for palliative care, and he died peacefully at home 2 weeks later.

Discussion

Large aneurysm series have found that paediatric aneurysms account for only 0.5–4.5% and that they are exceedingly rare in infants [5]. In children under 5 years of age there is a higher incidence in the first 2 years of life, with more than 50% occuring in the first year. A male predominance of 2:1 has been reported [1].

More infantile and early childhood aneurysms (40%) arise from the middle cerebral artery than from any other [7]. There is also more frequent involvement of the posterior circulation and of distal branches than in adult cases of cerebral aneurysms. Over three-quarters of cases in children under 5 years of age involve large (>10 mm) or giant lesions (>25 mm).

Microscopy studies have only been carried out in six neonatal cases. Five of these showed fragmentation or ab-

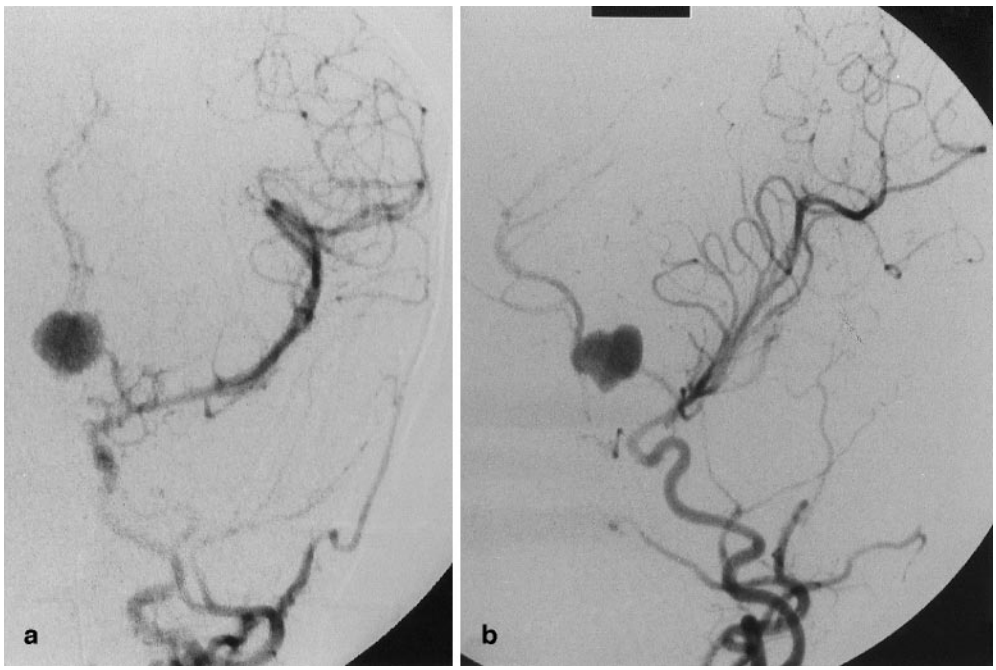

Fig. 3 **a** Frontal and **b** oblique DSA views of the left carotid territory confirm the presence of a large fusiform aneurysm of the left anterior cerebral artery giving rise to both A2 segments

sence of the internal elastic lamina and smooth muscle within the aneurysmal sac [1–3, 4, 9], a histological feature similar to that of adult saccular aneurysms [12]. The remaining case, a neonatal fusiform aneurysm, showed organised thrombus with pseudolime (a calcium-like substance) formation [10].

Anomalies of the circle of Willis have not been mentioned in association with other reported neonatal aneurysms, but our case displayed a dysplastic anterior circle of Willis with an aplastic right A1 segment, the aneurysm being supplied by the left A1, and both A2 segments arising from the aneurysm. Fusiform aneurysms are thought to occur in dysplastic vascular segments.

Hypoplasia of the A1 segment of the anterior cerebral artery is a common association with saccular aneurysms of the anterior communicating artery complex in adults. Such anatomical variations of the circle of Willis are thought to induce degenerative changes through haemodynamic stress [12].

The anterior communicating artery evolves embryologically from an arterial plexus. Incomplete resorption of the plexus may lead to mural weakness, precipitating aneurysm formation [14]. Nishio et al. [7] postulate that haemodynamic stress and embryonal development tend to influence the aetiology of aneurysms in infants.

Kuchelmeister et al. reported a possibly familial case, which was found to have features of fibromuscular dysplasia [3]. Other authors have considered their cases to be congenital, but this has been disputed by Stehbens, who found inco nclusive evidence after reviewing reports with a panel of experts. He claimed that even the occurrence of a few congenital aneurysms could not preclude an acquired, degenerative pathogenesis for the majority [11]. Neonatal aneurysms, particularly around the incisura, may also be related to birth trauma [8].

Neonatal aneurysms usually present with features attributable to subarachnoid haemorrhage: irritability, vomiting, lethargy, apnoea, cyanosis, opisthotonus, seizures and loss of consciousness. Mass effect however, has been reported in 18.1% of early childhood cases, as against 2.5% of adult cases. Ferrante et al. [1] concluded that children under 5 are less sensitive to posthaemorrhagic vasospasm, as only 3 of 40 cases demonstrated such a feature on angiography, whilst at least 30% of adults with ruptured aneurysms experience late arterial spasm [6]. However, our case had clear angiographic evidence of large vessel intracranial arterial spasm.

All neonates previously described with aneurysmal subarachnoid haemorrhage who have survived have been managed surgically, except the case reported by Piatt and Clunie: this patient was managed conservatively and the aneurysm was no longer detectable at 40 days of age. In Shimauchi et al.'s case [10], the parent artery of the fusiform artery was clipped. In the other cases, either the neck of the aneurysm was clipped or the aneurysm was excised.

Seven of those cases of ruptured neonatal aneurysm previously described were diagnosed post mortem. Among the six cases diagnosed during life, only one death was reported. All the four cases managed surgically survived. Ferrante et al.'s [1] review of 71 cases in children below the age of 5 showed an 11% mortality after first bleed and operative mortality of 9.5%. Thrush and Marano [13] reviewed 43 cases of infantile aneurysms, of which 18 were treated by surgery: 14 of the patients had a good surgical outcome whilst 2 died. Little is known about the long-term outcome of neonatal ruptured aneurysms.

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