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Case reports

Ruptured vertebrobasilar aneurysm associated with giant cell arteritis in a young boy

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Abstract

Intracranial aneurysms are rare in early childhood and there is little published information on their histology. We report a young boy who died of subarachnoid haemorrhage, 29 months after coiling of a giant vertebrobasilar aneurysm. Histology of the aneurysm revealed intramural inflammation with giant cells and fragmentation of the internal elastic lamina. The findings highlight the need for detailed examination in such cases, to elucidate the pathogenesis and pathology of cerebrovascular aneurysms in this age group.

Keywords: Giant aneurysm; Vertebrobasilar artery; Coil embolisation; Subarachnoid haemorrhage; Giant cell arteritis

1. Introduction

Intracranial aneurysms are rare during first few years of life. In this age group they are usually symptomatic and are more likely to involve the carotid artery, anterior communicating artery or vertebrobasilar system than are those in adults [1–4]. Limited information is available on the histological findings. A few papers have described loss of tunica media and an absent or discontinuous internal elastic lamina (for review, see Ref. [1]). In most cases, inflammation has not been present. We describe a young boy who had a giant intracranial aneurysm that was initially treated by coiling but subsequently recurred and caused fatal subarachnoid haemorrhage. Post-mortem examination revealed changes of giant cell arteritis within the ruptured aneurysm.

2. Case report

A Caucasian male aged 2.5 years was admitted to his local hospital after collapsing and becoming unrousable. He had recently complained that his face and mouth felt "funny" but had otherwise been well. There was no significant family history of neurological or cardiovascular disease. MR imaging revealed a giant left vertebrobasilar artery aneurysm with brain stem compression and hydrocephalus. He was transferred as an emergency to Frenchay Hospital where angiography confirmed the MR findings (Fig. 1A). Clinical examination revealed no evidence of disease outside of the CNS and routine biochemical and haematological investigations were normal. An external ventricular drain was inserted and the aneurysm subsequently treated by coiling with bare platinum Guglielmi coils (Fig. 1B). He made a good recovery but required insertion of a ventriculoperitoneal shunt for treatment of the hydrocephalus. Follow-up angiograms 12 and again 24 months after the coiling showed filling of a small remnant of the aneurysm neck (Fig. 1C) but no filling of the sac. The aortic arch and major arteries in the neck all appeared normal. Twenty nine months after the initial presentation, he was found unrousable in bed. A CT scan revealed subarachnoid haemorrhage and a swollen brain with small ventricles and loss of grey-white matter differentiation. He was again transferred to Frenchay Hospital. On arrival he was unresponsive and had fixed dilated pupils. He remained obtunded and died the next day. Con-

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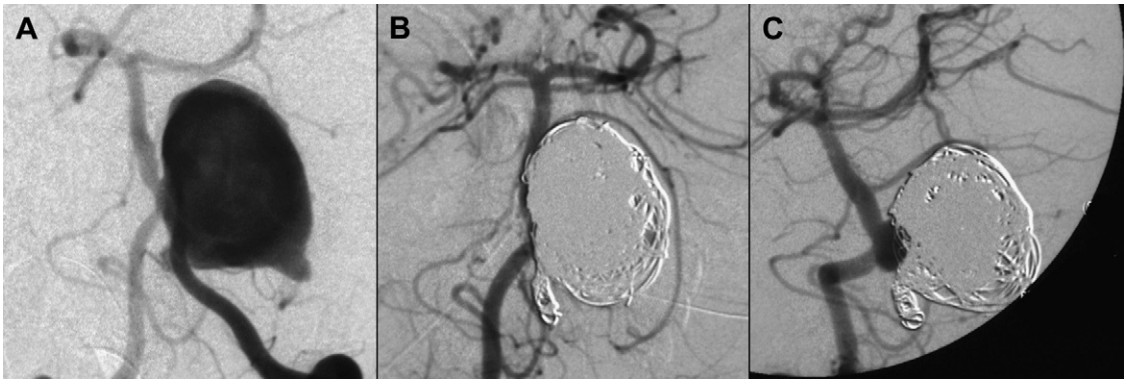

Fig. 1. Neuroradiological findings. (A) Left occipitomental oblique projection of a left vertebral digital subtraction angiogram (DSA) demonstrates a giant aneurysm arising from the distal left vertebral artery and extending up to and involving the vertebrobasilar junction. A daughter lobule arises from the sac inferiorly on the left. (B) DSA occipitomental projection following injection of the right vertebral artery immediately after coil embolisation demonstrates satisfactory occlusion of the aneurysm. (C) DSA occipitomental oblique projection following right vertebral artery injection 12 months after coil embolisation demonstrates some coil compaction at the aneurysm neck with the development of a small neck remnant. However, the aneurysm remains largely occluded.

sent was given for a post-mortem examination limited to the head.

The brain was soft and swollen. A patent shunt catheter was present in the right parieto-occipital region. Both temporal lobes had undergone uncal herniation and the cerebellar tonsils had herniated and were necrotic. Subarachnoid blood was present over the cerebral convexities but was most abundant over the base of the brain, and ventral aspect of the brain stem (Fig. 2A). Embedded within the blood clot was a giant aneurysm arising from the junction of the vertebral and basilar arteries. The posterior part of the aneurysm comprised admixed coils and fibrous tissue and measured approximately 25 mm in diameter (Fig. 2B and C). Attached to this mass of coils and fibrous tissue was an anterior, coil-free extension of the aneurysm, measuring 18 mm from above down, 16 mm from side to side and 9 mm from before back. The aneurysm had a large anterior saccular element and smaller superior, lateral and inferior protrusions. The remainder of the vertebral and basilar arteries and the circle of Willis appeared normal, with no aneurysms or other malformations. The brain was fixed by suspension in formalin. Examination of coronal slices through the cerebrum revealed slit-like lateral and third ventricles. The cortical ribbon and deep grey matter structures were a dusky brown colour. Horizontal slices through the brain stem and cerebellum showed marked distortion and displacement of the pons, left middle cerebral peduncle and adjacent cerebellar tissue by the aneurysm. Blocks for histology were taken from the anterior sac of the aneurysm, basal vessels and multiple regions of cerebrum, cerebellum and brain stem.

Histology revealed widespread acute ischaemic neuronal damage. The pons and cerebellum had been distorted by the adjacent aneurysm and included several small foci of perivascular haemorrhage. Occasional haemosiderin-laden

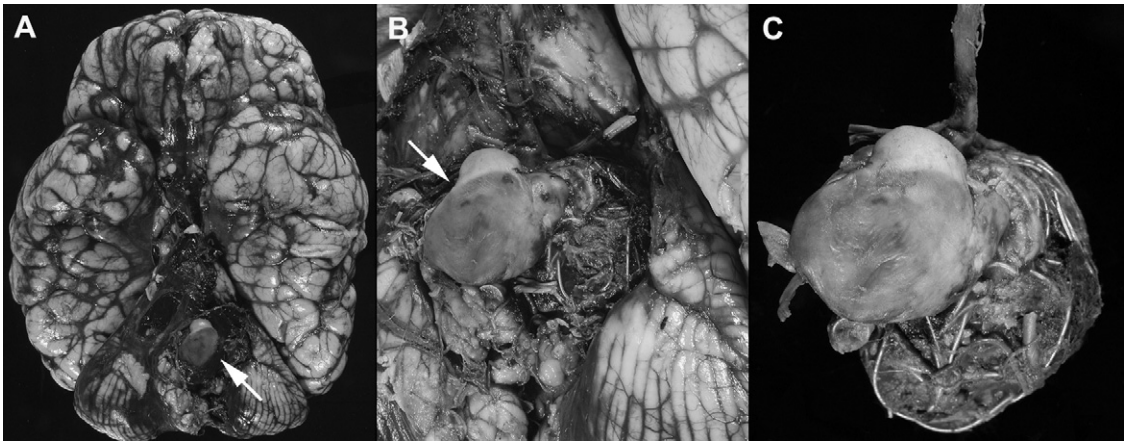

Fig. 2. Macroscopic findings post-mortem. (A) Basal brain structures are partly obscured by subarachnoid blood clot. An anterior extension of the giant vertebrobasilar aneurysm (arrow) is visible to the left of the midline, adjacent to the pons. (B) Removal of the blood clot shows the posterior part of the 'aneurysm' to comprise admixed coils and fibrous tissue. Attached to this is the anterior extension of the aneurysm (arrow), which did not contain coils. The necrotic cerebellar tonsillar tissue is visible below the aneurysm. (C) The two parts of the aneurysm and the attached basilar artery are clearly seen in this dissected specimen. The anterior extension of the aneurysm was processed for histology.