Cerebral aneurysm in an infant with fibromuscular hyperplasia of the renal arteries

Case report

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The authors report a case of subarachnoid hemorrhage in an 11-month-old infant with tragic outcome. Radiological investigation showed an anterior communicating aneurysm, and postmortem examination confirmed the aneurysm to be a so-called "berry" aneurysm. There were also typical signs of fibromuscular hyperplasia of the renal arteries. The microscopic findings are discussed. In view of the rarity of both aneurysms and fibromuscular hyperplasia in such a small child, a possible association of these entities suggested by several earlier investigators is reviewed.

KEY WORDS • subarachnoid hemorrhage • fibromuscular hyperplasia • cerebral aneurysm • congenital vascular disease • infant

Intracranial arterial aneurysms are uncommon in children. In a total of 1175 cases of verified subarachnoid hemorrhage, Laitinen found only nine patients with saccular aneurysm who were younger than 15 years (1.3% of all saccular aneurysms). Only one patient was less than 5 years of age. Patel and Richardson collected 58 patients below the age of 19 years from among 3000 cases of intracranial aneurysms with subarachnoid hemorrhage. None of these patients was less than 7 years of age.

There are, however, several reports of intracranial aneurysms in infants and young children. Pool and Potts refer to seven cases in children of less than 1 year old. We have not been able to find any previous case of an intracranial aneurysm associated with fibromuscular hyperplasia of the renal arteries in an infant or child.

Case Report

This 11-month-old boy was the result of a normal pregnancy and parturition. There was no family history of hereditary vascular disease. The patient was treated in the hospital for a fracture of the clavicle and, in October, 1976, for a superficial skin infection with lymphangitis above the left ear. Two months after the skin infection, the patient appeared tired and vomited. Suddenly, several hours later, he had a tonic seizure, followed 5 minutes later by a clonic seizure. On admission to the hospital about 1 hour later, the patient was unconscious, with adequate reac-
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Fig. 1. Left: Computerized tomography scan, showing a small hematoma at the site of the aneurysm and rupture into the ventricular system. Right: Right carotid angiogram, anteroposterior view, showing the aneurysm on the anterior communicating artery.

tions to painful stimuli. The pupils were of equal size and reacted normally. His breathing was normal. On lumbar puncture, the cerebrospinal fluid was mixed with blood. During transfer to a neurosurgical clinic, the patient's condition worsened. He had more seizures and on admission he was deeply comatose (Botterell Grade 4) with Cheyne-Stokes respiration. Pulse varied between 80 and 160/min. The pupils were small with no reaction to light or corneal reflexes. Emergency computerized tomography and cerebral angiography showed an arterial aneurysm on the anterior communicating artery with a small intracerebral hematoma and hemorrhage into the ventricular system (Fig. 1).

The patient was considered to be unsuitable for surgery. Conservative treatment with artificial hyperventilation and osmotherapy was initiated. He died 2 days after the hemorrhage.

A complete postmortem examination was performed within 2 hours after death. The body weighed 7500 gm and was 71 cm in length. A large saccular aneurysm was found on the anterior communicating artery, with a hematoma extending to the left lateral ventricle; the ventricular system was filled with blood. The heart weighed 44 gm and was normal in size and configuration. No sign of aortic coarctation could be discovered. In both renal arteries, mainly at the opening from the aorta, the arterial wall was thickened and the lumen narrowed. No atherosclerotic changes were observed in the aorta or the large arteries. No aneurysm could be seen on the extracranial arteries.

Microscopic examination showed that the aneurysm wall was built up of connective tissue, most of which resembled intima and was remarkably rich in cells (Fig. 2). The latter also had polymorphous nuclei with some giant cells. No mitoses could be discovered. There was no smooth muscle or elastic tissue in the aneurysm wall. At the mouth of the aneurysm the intimal connec-
Fig. 2. Histological sections of the aneurysm. **Left:** Section of the aneurysm wall demonstrating the connective tissue resembling intima that was remarkably rich in cells. The nuclei are polymorphous and giant cells can be seen. Gomori's elastin & van Gieson, × 275. **Right:** Mouth of the aneurysm, lumen upward. The aneurysm is at the bottom and the parent artery at the top. The black-stained elastic lamella is split up into several thin lamellae varying in staining properties near the site of rupture. The edge of the tunica media is rounded. Gomori's elastin & van Gieson, × 90.

tive tissue was still thickened, the elastic lamellae split up before they ended abruptly, and the edge of the muscular layer was rounded. Only small media defects (less than 100 µ in diameter) occurred in other large cerebral arteries. The renal arteries had intimal protrusions that consisted of elastic lamellae interspersed with collagenous connective tissue and solitary smooth-muscle cells (Fig. 3).

**Discussion**

Earlier reports indicate a higher than average frequency of intracranial aneurysms in children with coarctation of the aorta and in children with polycystic kidneys. Other reports point to a higher frequency of aneurysms in children with Marfan's syndrome and Ehler-Danlos syndrome, in which there is an accompanying diffuse arterial disease. The histological picture of the aneurysm in the present case was of the so-called congenital type. There seemed to have been a defect of the media at the site of the aneurysm. The elastic lamella showed typical signs of hypertrophy and degeneration. The intimal proliferation was, however, remarkably cellular and polymorphous. Such intimal proliferation is generally not encountered in ordinary saccular aneurysms nor in the minute aneurysms that are sometimes found at autopsy in adults and are considered to be early stages of aneurysms. It may be questioned if the intimal proliferation represented a very early stage of an aneurysm in this case because the patient was only 11 months of age. It may also be questioned if the connective tissue of the intima of such a young child reacts in a different way so that a more cellular tissue is obtained. The etiology of the intimal proliferation is not known.

Fibromuscular hyperplasia (FMH) of the renal arteries is characterized histologically by irregularly spaced focal zones of fibrous and muscular hyperplasia of the intima and media, disruption of elastic lamella, and the occasional appearance of berry aneurysms at
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FIG. 3. Histological section of the origin of the left renal artery. The lumen is narrowed by a thickening of the intima that consists of elastic lamellae, smooth-muscle cells, and collagenous connective tissue, that is, fibromuscular dysplasia. Gomori's elastin & van Gieson, × 38.

points of disruption of the media. A relation between FMH and intracranial aneurysm has been suggested by several investigators.2,4,8,15 Palubinskas, et al.,8 found five cases of intracranial aneurysms in 70 patients with FMH of the renal arteries. Another four patients had intracranial aneurysms associated with signs of FMH of the carotid arteries.

The age of onset of FMH has been reported at between 1.5 and 79 years in both sexes, but the highest incidence is in middle age, which is similar to that of intracranial aneurysms.8 We believe that our case with an intracranial berry aneurysm and histological signs of FMH in the renal and cerebral arteries supports the theory of an association between these two entities. We further believe that the association can best be explained by the fact that FMH is a generalized arterial disease where an aneurysm can develop. The fact that FMH gives an arterial hypertension with a secondary effect of the formation of an aneurysm is not the main cause, even though it might be a contributing factor.

References


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