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Dissecting Intracranial Aneurysm

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Summary. A case of spontaneous dissecting aneurysm of the right internal carotid and middle cerebral arteries is presented in a 13-year-old boy. The pathogenetic factors incriminated in previously reported cases are reviewed and the pathological findings are discussed. The abnormalities of the internal elastic lamina as seen in this patient have been observed in numerous cases of intracranial dissecting aneurysms. It is concluded that these defects play an important role in the development of dissection.

Key words: Stroke – Intracranial dissecting aneurysm – Defective elastic lamina of cerebral arteries

Zusammenfassung. Es wird über einen Fall von einem 13jährigen Jungen mit Schlaganfall und spontanem dissezierendem Aneurysma berichtet. Die angenommenen pathogenetischen Faktoren und die pathologischen Befunde der in der Literatur bereits beschriebenen Fälle werden diskutiert. Die Veränderungen der Lamina elastica interna, die auch in dem vorliegenden Fall gefunden wurden, sind in vielen Fällen der intrakraniellen dissezierenden Aneurysmata beobachtet worden. Aufgrund dieser Beobachtung darf darauf geschlossen werden, daß Schädigung dieser Art eine wichtige Rolle in der Entstehung der Dissektion spielen kann.

Schlüsselwörter: Schlaganfall – Intrakranielles dissezierendes Aneurysma – Schädigung der Lamina elastica interna der Hirnarterien

Introduction

Intracranial dissecting aneurysms are usually characterized by the subintimal site of dissection. Approximately 40 cases of dissecting aneurysms of the cerebral arteries have been described in the literature since 1915, when Turnbull [24] published the first case. Clinically the manifestation of these lesions were strokes of

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sudden onset in young patients. Not all cases of intracranial dissection are fatal [6, 16, 27]. A number of pathogenetic mechanisms have been suggested in the literature, but no specific cause was identified in most reported cases [7, 14, 18, 19]. We present the case of 13-year-old boy with dissecting aneurysm of the right carotid and middle cerebral arteries.

Case Report

A 13-year-old boy was admitted to our Department on January 13, 1981. The patient had been in good health before admission, and his growth and development had been normal. On the day of admission he suddenly developed a severe right frontal headache and lost consciousness. Approximately 4–5 min later he regained consciousness, but was unable to move his left extremities.

Examinations

There were no abnormal physical signs on general examination. The blood pressure was 120/70 mm Hg and the radial pulse 80/min. He had no neck stiffness. The optic discs appeared normal with no evidence of papilledema. Left homonymous hemianopsia, a severe flaccid left hemiparesis, severely affecting the arm and left central facial weakness were observed. The tendon reflexes of the left extremities were exaggerated with an extensor plantar response. The abdominal reflexes were absent on the left side. He was conscious but drowsy.

Laboratory Data

The ESR was 10 mm in the first hour. Hemoglobin was 127 g/l, white blood cell count $10.0 \times 10^9$1 with a differential count of 0.84 segmented forms, 0.12 lymphocytes and 0.04 monocytes. The blood sugar was 10.4 mmol/l. Blood coagulation studies were normal along with all other laboratory findings. X-rays of chest and skull were normal. EEG showed high amplitude 2–5 c/s activity in the right frontoparietal region. The cerebrospinal fluid was clear, colorless, it contained 1 white blood cell mm^3. The protein content was 275 mg/l.

Right carotid angiography was performed on the day of admission, and the supraclinoidal portion of the internal carotid artery was narrow and its lumen was irregular. There was no filling of the right anterior cerebral artery. Two days later right carotid angiography was repeated, and showed almost total occlusion of the right middle cerebral artery and its branches. A left carotid angiogram on January 16 revealed normal internal carotid, middle cerebral and posterior cerebral arteries. Both anterior cerebral arteries were seen to fill from the left carotid injection. The anterior cerebral arteries and deep veins were shifted 1.5 cm from right to left.

Course

During the first week the clinical state of the patient did not change appreciably. He was treated with hyperosmotic agents and intravenous fluids. Seven days after admission he suddenly became comatose and decerebrate with irregular breathing and he developed a right third cranial nerve palsy. He was placed on a respirator, spontaneous respiration never recurred. He died 9 days after admission.

Necropsy

The autopsy revealed arteria renalis duplex on both sides and thrombi were observed in the periprostatic veins.

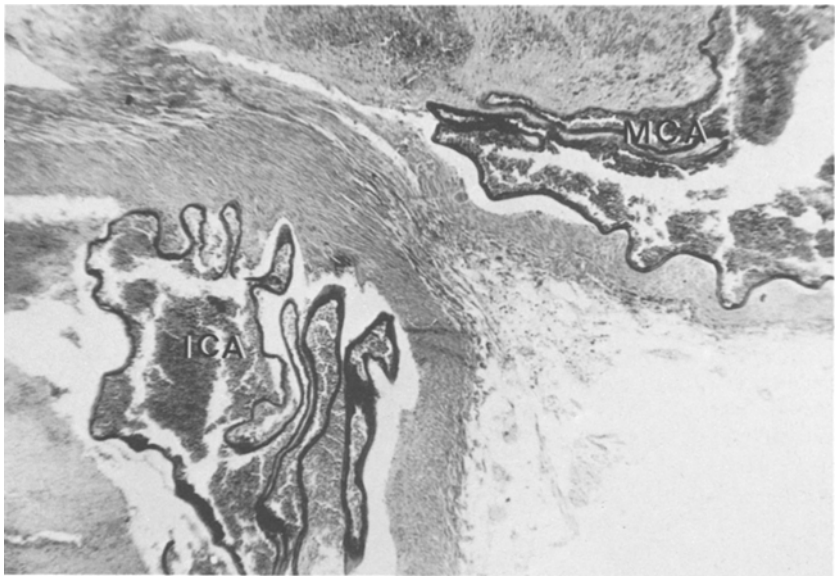

Fig. 1. Transverse section of the right internal carotid *(ICA)* and middle cerebral arteries *(MCA)* at the site of the bifurcation. The lumen of both arteries is narrowed to a convuluted slit on one side by an intramural thrombus. – Elastic-van Gieson (original magnification × 32)

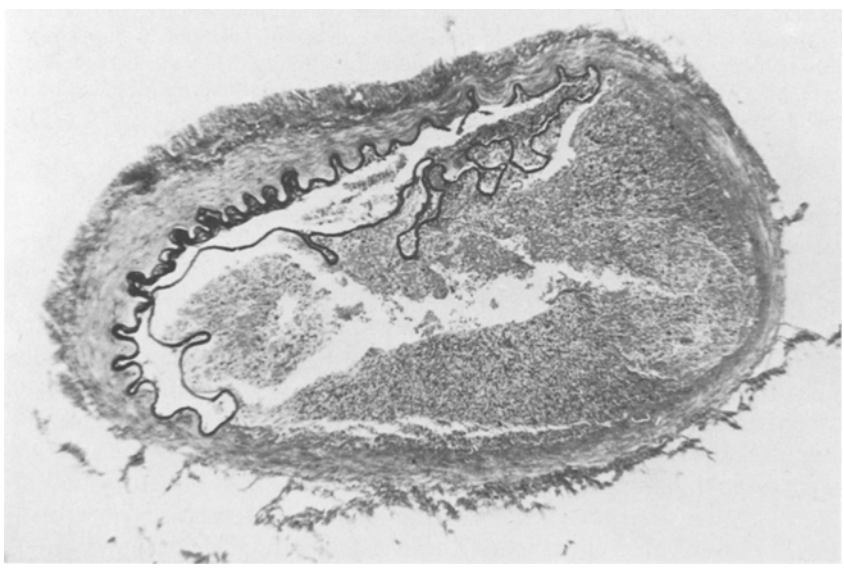

Fig. 2. Microscopic section of a distal branch of the right middle cerebral artery demonstrates the displacement of the elastic lamina by a large thrombus. – Elastic-van Gieson (original magnification × 32)

Neuropathological Findings

The brain weighed 1820 g and was markedly edematous, with transtentorial herniation on the right side and bilateral tonsillar herniation. The right hemisphere was markedly swollen with flattening of the convolutions and narrowing of the sulci, the right cerebral hemisphere was soft in consistency. The midline structures were displaced by 2 cm to the left. The parenchyma was dull and pale gray in contrast to the left hemisphere which was relatively normal in appearance. External examination of the vessels of the circle of Willis and its brances revealed no apparent abnormalities either in caliber or course. Sections through the right internal carotid and middle cerebral arteries showed the vessels to be filled with a thrombus.

Histological Examination

Brain. Microscopic sections from the right cerebral hemisphere showed changes usually seen in recent infarction within the distribution of the internal catorid artery. Numerous shrunken hyperchromatic nerve cells were present in the affected area and there was a slight polymorphonuclear leukocytic reaction in the softened tissue. The brain tissue was spongy due to edema.

Vessels. Examination of the right carotid and middle cerebral arteries showed acute hemorrhagic dissection of the intima from the media with early thrombus formation causing considerable narrowing of the lumen (Fig. 1). The internal carotid artery was completely occluded. The lesion extended from the supraclinoidal portion of the internal carotid artery to distal branches of the middle cerebral artery such as the posterior parietal branch (Fig. 2). The anterior cerebral artery was not involved in the dissection. A rupture was found in the internal elastic lamina at the origin of the middle cerebral artery (Fig. 3. a, b) and endothelial cells had proliferated and covered the edges of the ruptured lamina. At the site of the subintimal dissection there were large fibroelastic intimal cushions (Fig. 4) with splitting and focal absence of the internal elastic lamina. The vertebral, basilar and posterior cerebral arteries were normal, whereas splitting, fraying, disintegration and irregular thickening of the elastic lamina marked all the major vessels of the intracranial carotid system on both sides. These defects of the internal elastic lamina did not involve the entire circumference of the vessel wall, but were often continous with apparently normal portions of internal elastic lamina with in the same cross section. Intimal pads and thinning of the media were usually present at the arterial bifurcations. There was no evidence of inflammation within the vascular wall. Histochemical reactions failed to reveal abnormal accumulation of acid mucopolysaccharides as seen in cystic medial degeneration.

Discussion

Several theories concerning the pathogenesis of intracranial dissecting aneurysms have been discussed in the literature. Turnbull [24] and Szabó [23] reported subintimal dissection on the basis of syphilitic arteritis. In two other cases the dissection has been associated with nonspecific arteritis [5, 20]. Mucoid or cystic medial necrosis as underlying causes are mentioned by several authors [2, 25, 26]. A number of authors related the dissection to head injury [13,22]. Spudis [21] has suggested a causal relation between migraine and the dissecting aneurysm. Wolman [27] observed medial defects of the vessel wall in his three cases of cerebral dissecting aneurysm, and he labelled these as congenital in origin. Various abnormalities of the internal elastic lamina such as splitting, fraying, reduplication and irregular thickening have been described in numerous cases of cerebral dissecting aneurysms with or without congenital gap defects [2, 5, 11, 15, 17, 26].

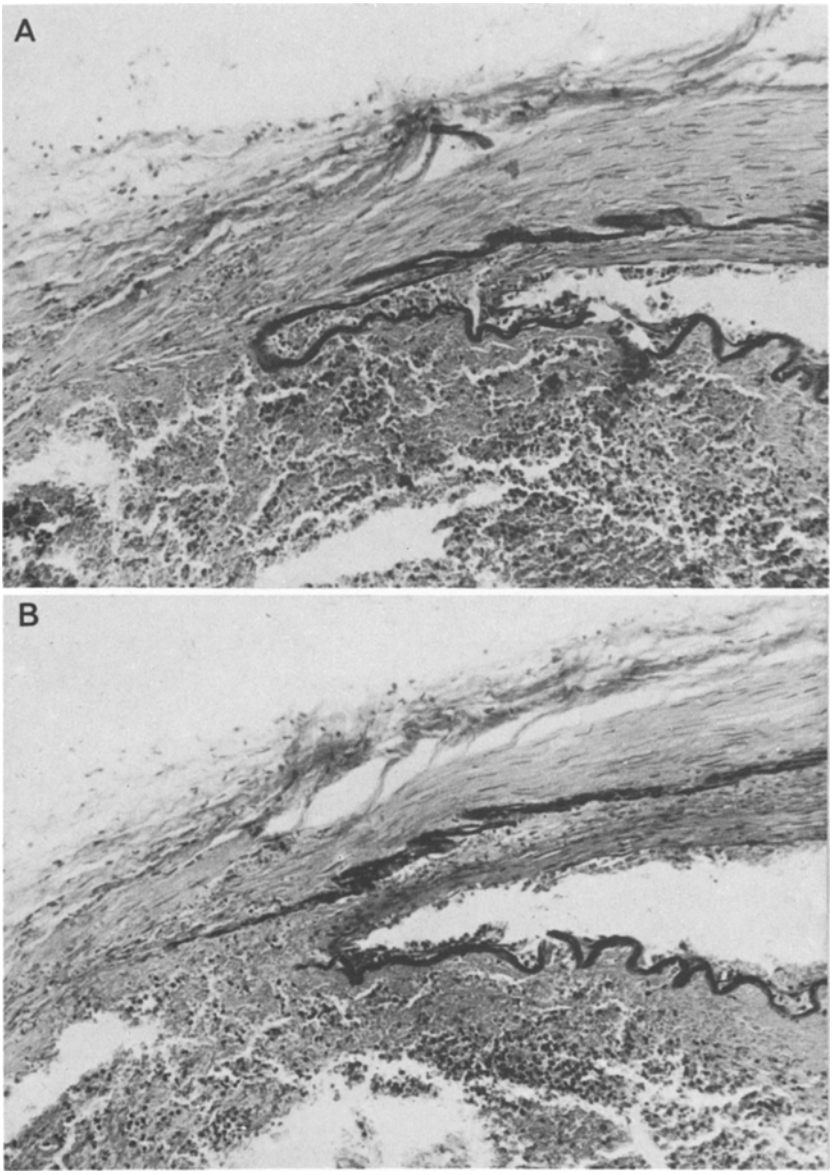

Fig. 3 A, B. The right middle cerebral artery *(A)* proximal to the rupture of the elastic lamina, *(B)* the site of the rupture. Elastic-van Gieson (original magnification × 100)

Adelman [1] found focal absences of the internal elastic lamina in a 14-year-old boy with bilateral dissecting aneurysms of the carotid and middle cerebral arteries. The association with Moya Moya syndrome and fibromuscular dysplasia of extracranial arteries has been observed [17].

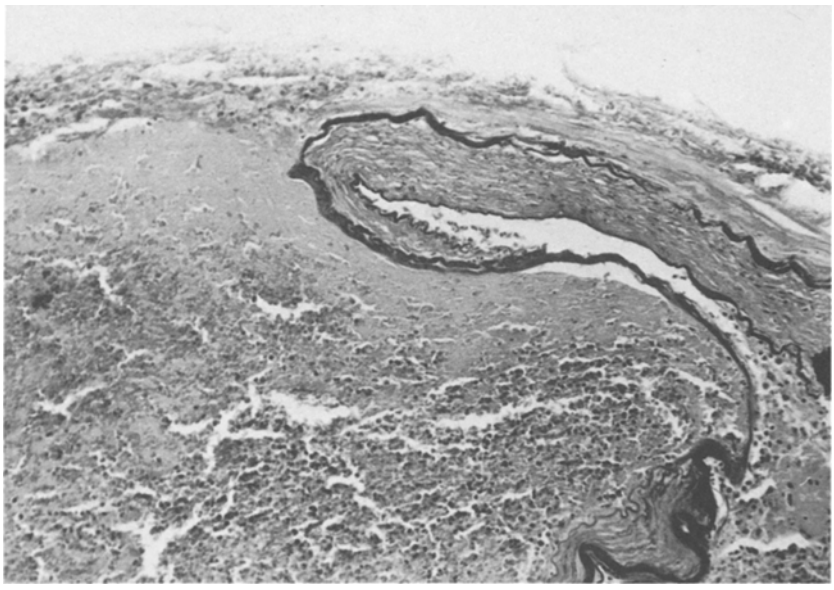

Fig. 4. The right middle cerebral artery. A fibroelestic intimal cushion covers the rupture of the internal elastic lamina. Elestic-van Gieson (original magnification × 100)

In most cases the dissection was subintimal and separated the internal elastic lamina from the media. The hematoma occurred within the media or adventitia in only a few cases of cerebral dissecting aneurysms [12, 28].

It is well known that the cerebral arteries differ from arteries of the same caliber elsewhere in that their walls are much thinner and have only one elastic layer [8, 22]. The majority of cerebral vessels, except for the initial segments of the carotid and vertebral arteries lack vasa vasorum [28]. Yonas [28] assumed that the rare dissection within the media or adventitia originated either from the vasa vasorum of these proximal vessel segments or from new vessels which form in the necrotic media. In other cases the vessel lumen is the only possible source of intramural hemorrhage. Wolman [27] postulated that defects of the internal elastic lamina adjacent to medial defect are responsible for the development of dissection in the cerebral arteries. Focal absence and other abnormalities of the internal elastic lamina have been reported in patients dying of subarachnoid hemorrhage [8, 9], in cases of basal arterial stenosis and thalamic and basal ganglionic telangiectasia [4, 10, 17] and in Marfan's syndrome [3]. Hassler [8] did not find such changes in 250 normal patients of all ages. These diseases are presumably different manifestations of the same underlying disorder. Media discontinuities or gaps and intimal pads are common at the bifurcation or branching of the cerebral vessels and their number increases with age [8, 22].

The changes of the internal elastic lamina as seen in the present case must be considered abnormal but are of unknown significance. Our observations show that young patients with clinical signs of stroke may actually have dissecting

aneurysm of cerebral arteries and pathological changes of the elastic lamina probably play an important role in the development of the aneurysm.

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