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Defects of Elastic Lamina in Middle Cerebral Artery

A Possible Cause of a Primary Intracerebral Hemorrhage in a Young Woman

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Summary. Circumscribed defects in the elastic lamina as a possible cause of intracerebral hemorrhage are reported. The abnormality was present in the right middle cerebral artery of a 24-year-old woman. The patient died of large intracerebral hemorrhage which extended into the lateral ventricle on the same side and to the subarachnoid space. In the absence of vascular malformation and secondary degenerative changes attributed to hypertension these defects of the internal elastic lamina associated with the rise of the blood pressure appear to lead ultimately to bleeding.

Key words: Intracerebral hemorrhage – Defects in the elastic lamina

Zusammenfassung. Es wird über umschriebenes Fehlen der Lamina elastica interna als eine mögliche Ursache der intrazerebralen Massenblutung berichtet. Die Veränderungen sind in der Wand der rechten cerebri media einer 24jährigen Frau beobachtet worden, die wegen einer ausgedehnten intrazerebralen Massenblutung starb. Die Massenblutung mit Einbruch in den Seitenventrikel und in den Subarachnoidalraum befand sich an der Seite der Elastika-Veränderungen. Diese Defekte der Lamina elastica interna im Zusammenhang mit der Blutdruckerhöhung scheinen schließlich zur Blutung geführt zu haben, da Gefäßmalformationen und hypertonische Gefäßerkrankungen ausgeschlossen werden konnten.

Schlüsselwörter: Intrazerebrale Massenblutung – Umschriebenes Fehlen der Lamina elastica interna

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Introduction

Primary intracerebral hemorrhage is rare under the age of 30 years (Stehbens 1972), and intracerebral hemorrhage due to rupture of an arteriosclerotic vessel or a berry aneurysm occurs mainly between 40 and 60 years of age (Sheffield et al. 1980). The purpose of this paper is to describe the case of a 24-year-old woman who died of massive intracerebral hemorrhage within 24 h. On microscopic sections there were focal absences in the internal elastic lamina confined to the middle cerebral artery on the side of bleeding. The walls of the other intracranial arteries sectioned serially were normal.

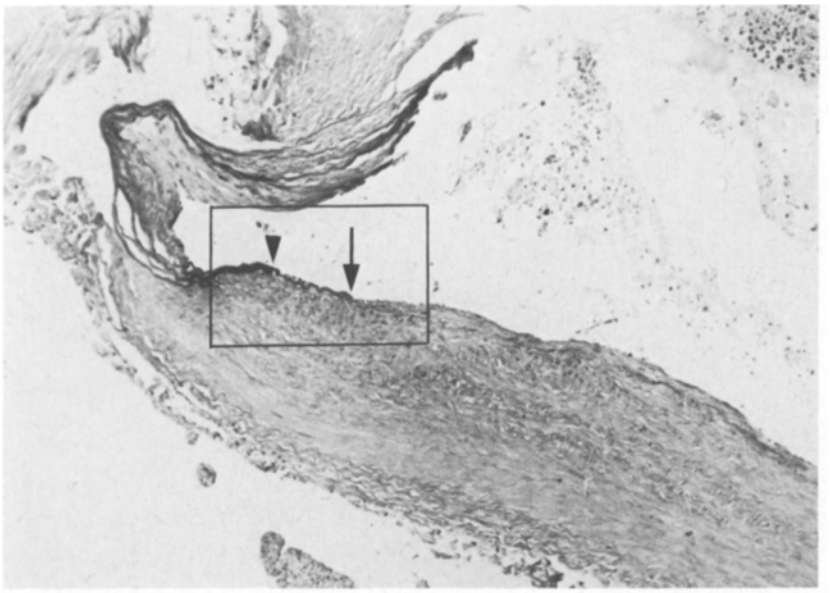
Case Report

A 24-year old, right handed woman was admitted to our department on June 17, 1981 because of sudden onset of right-sided headache and vomiting, followed by paralysis of the left arm and leg 2 h before admission. Although she became drowsy, there was no loss of consciousness. In June 1979 the patient has been hospitalized for toxemia of pregnancy, and since that time had been treated for hypertension. On admission her blood pressure was 280/130 mm Hg and pulse rate 80/min, and physical examination was normal. There was nuchal rigidity and severe left flaccid hemiparesis most marked in the arm with left central facial weakness. She was drowsy but responsive. Cisternal puncture revealed grossly bloody cerebrospinal fluid. Therapy with iv administered antihypertensive medication was started to maintain the blood pressure at 160/100 mm Hg. Despite treatment with mannitol and dexamethasone the patient became comatose and decerebrate and died 9 h following admission.

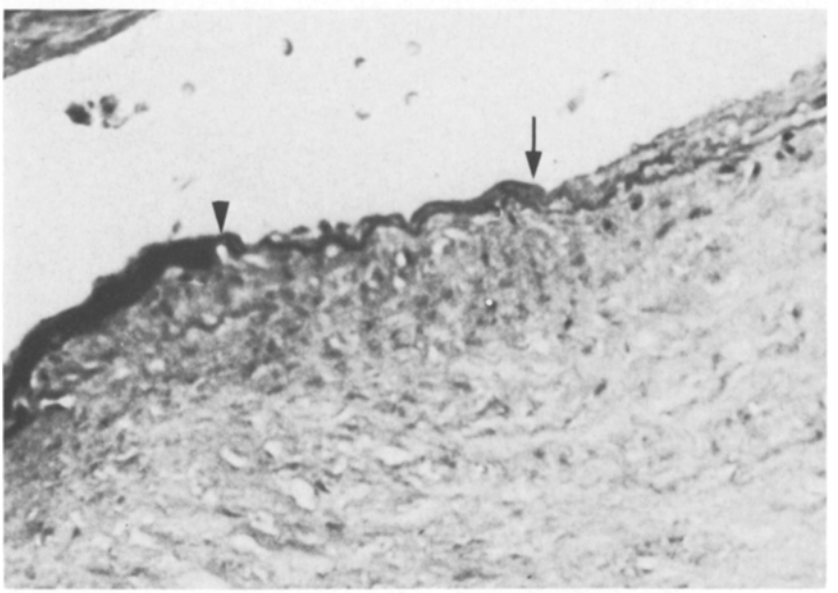
Pathological Findings

The brain showed edema externally with gyral flattening and bilateral, uncal, and tonsillar herniation. The right hemisphere was markedly swollen. There was subarachnoid hemorrhage over most of both hemispheres and in the interpeduncular space. Coronal sections revealed extensive destruction of the central white matter and basal ganglia of the right hemisphere secondary to an intracerebral hemorrhage which extended approximately 3 cm from the frontal pole to 4 cm from the occipital pole and had ruptured into the lateral ventricle. Small hemorrhages involved the tegmentum of the pons and midbrain.

Gross examination of the vessels of the circle of Willis and its branches revealed no aneurysm or other abnormalities. The microscopic sections from the proximal part of the right middle cerebral artery showed localised defects of the internal elastic lamina with slight bulging of the wall (Fig. 1). The lesion involved approximately a quarter of the circumference of entire vessel wall. At the origin of a little branch the vessel wall was extremely thin. In the distal segment of the right middle cerebral artery there was a break detected in the continuity of the elastic layer covered with a large intimal cushion (Fig. 2). The edges of the gap appeared to have been sharply cut. All the other larger cerebral vessels were sectioned and appeared healthy. On careful microscopic examination there was no evidence of vascular malformations including micro-angiomas or other secondary pathological alterations of the vessel walls in both hemispheres.

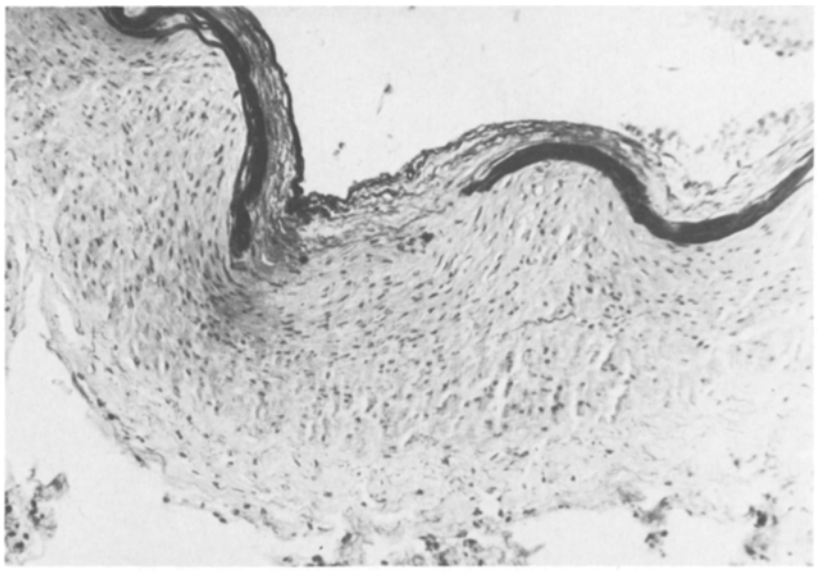


A



B

Fig. 1. A and B. The proximal part of the right middle cerebral artery: first the internal elastic lamina becomes abruptly thinner *(arrow-head)*, then disappears *(arrow)*. Elastic-van Gieson stain ×100. The area in the frame is shown at higher magnification in **B**



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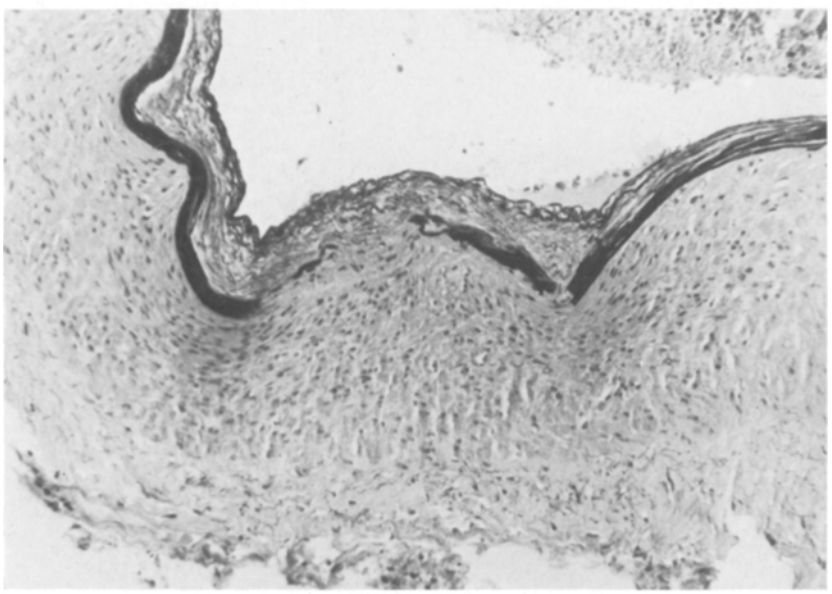

B

Fig. 2 A and B. Focal defects of the elastic layer in the distal segments of the right middle cerebral artery. **A** elastic-van Gieson, **B** orcein stain ×100

Discussion

In the great majority of cases intracerebral hemorrhages are the result of rupture of an arteriosclerotic vessel, being precipitated by elevated blood pressure. Other essential causes of intracerebral arterial rupture such as trauma, embolism, ruptured berry aneurysm or other vascular anomalies, blood dyscrasias are relatively rare. In spite of scrutinized pathological investigations one invariably encounters cases of unknown origin in any series of intracerebral hemorrhages. In our case there was no evidence of the above-mentioned pathogenetic factors. The pathological alterations considered characteristic of a minute aneurysm (Hassler 1961), and the so-called early aneurysmal changes (Stehbens 1963) were also absent.

Reuterwall (1923) first observed breaks in the continuity of the internal elastic lamina and described three types of tears: two with some type of intimal thickening and one with aneurysmal dilatation of the lumen. Hassler (1961) found such tears among the patients with aneurysm more frequently that in "normal" individuals and noted their increasing number with aging. Since Hassler's fundamental work there has been no mention in the literature about systematic investigations carried out on tears of the internal elastic layer, therefore their importance remains obscure.

Our case demonstrates that in the presence of defects of internal elastic layer the extreme rise of the blood pressure may cause the rupture of the arterial wall. It seems reasonable to assume that intracerebral bleeding of unknown origin in young patients can be a consequence of localised defects of the elastic lamina due to some developmental anomaly.

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