

## “Vaso-Muscular Mediadysplasia” of the Arteria vertebralis

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*Summary.* Report of a 48 year old woman with 15 years' history of arterial hypertension. Death due to dissecting aneurysm of the aorta with pericardial tamponade.

A peculiar abnormality in the architecture of the vascular wall of the vertebral arteries was observed: A vascular valve and transluminal bands in the right vertebral artery and tortuous arteriolar vessels as well as cavernomatous venous vessels in the outer part of the media of both vertebral arteries. The direction of muscle fibers in these sites was abnormal. These abnormalities are interpreted as a hamartomatous malformation, for which the term: vaso-muscular mediadysplasia is proposed.

So far little is known about the pathology of the vertebral arteries. Although differences of the caliber between the right and left arteries and abnormalities of the course of their extracranial part have been adequately described (Riechert, 1952; Bellingrath, 1954; Neimanis, 1956; Hutchinson and Yates, 1956; Krayenbühl and Yasargil, 1957; Jaquet, 1961; Kunert, 1961; Plötz, 1964, a. o.), detailed studies on their tissue level structure have been scarce (Benninghoff, 1930; Bellingrath, 1954). To the best of our knowledge abnormalities in the architecture of the vascular wall have not been described so far. It, therefore, seems to be of some interest to report a unique case with a peculiar form of “vaso-muscular media-dysplasia” of the vertebral artery.

### Case Report

A 48 year old woman was admitted to the emergency ward of the Department of Surgery (Heidelberg University) in acute distress. Apart from a 15 years' history of arterial hypertension (180/120 mm Hg) no further information on her medical history could be obtained. 11 h before admission she noticed excruciating pain in the dorso-lumbar region and a choking sensation in the throat. This was followed by acute paraplegia. Aortography revealed a dissecting aneurysm. Aortic fenestration and implantation of a dacron prothesis were performed unsuccessfully; 10 min after the operation the patient died from cardiac standstill.

### Autopsy Report (Nr. 922/72)

Concentric muscular hypertrophy of the left heart. Sclerotheromatosis with dilatation of the aorta. Sclerosis and ectasia of the coronary arteries. Moderate arteriosclerosis of the arteries of the cranial base. 1.2 cm above the aortic valves transverse intimal tear of the ascending aorta of 1.5 cm width. Dissecting aneurysm of the thoraco-abdominal aorta; dissecting circumferential hematoma reach-



Fig. 1. "Vascular valve" of the right vertebral artery

ing down to the bifurcation. Pericardial tamponade. Acute congestion of internal organs. Pulmonary edema. Cerebral edema.

#### *Vertebral Artery*

An extreme adhesion of the vertebral arteries to the surrounding connective tissue was noted, rendering dissection extremely difficult. Moderate atherosclerosis was found. The dissected right vertebral artery revealed further unusual changes. A *vascular valve* measuring  $0.6 \times 0.5$  cm was found attached to the dorsal wall at the level of the 5th cervical vertebra. While the valve's structure closely resembled that of a semilunar heart valve, its consistency differed markedly, the structure being more stiff and rigid.

In addition, in the right vertebral artery at the level of the so-called "Atlas-schleife" (atlas-loop), 8 yellowish-white bands up to a wool thread's thickness stretched across the vascular lumen.

Histological analysis revealed that the vascular valve consisted of a muscular ridge, an outfolding of the media, which was covered by an internal elastical membrane and an intima layer.

Similarly, histological sections of the *transluminal bands* described above again revealed a muscular core, which was again covered by an internal elastic membrane and an intima layer. Thus, the vascular valve as well as the transluminal bands

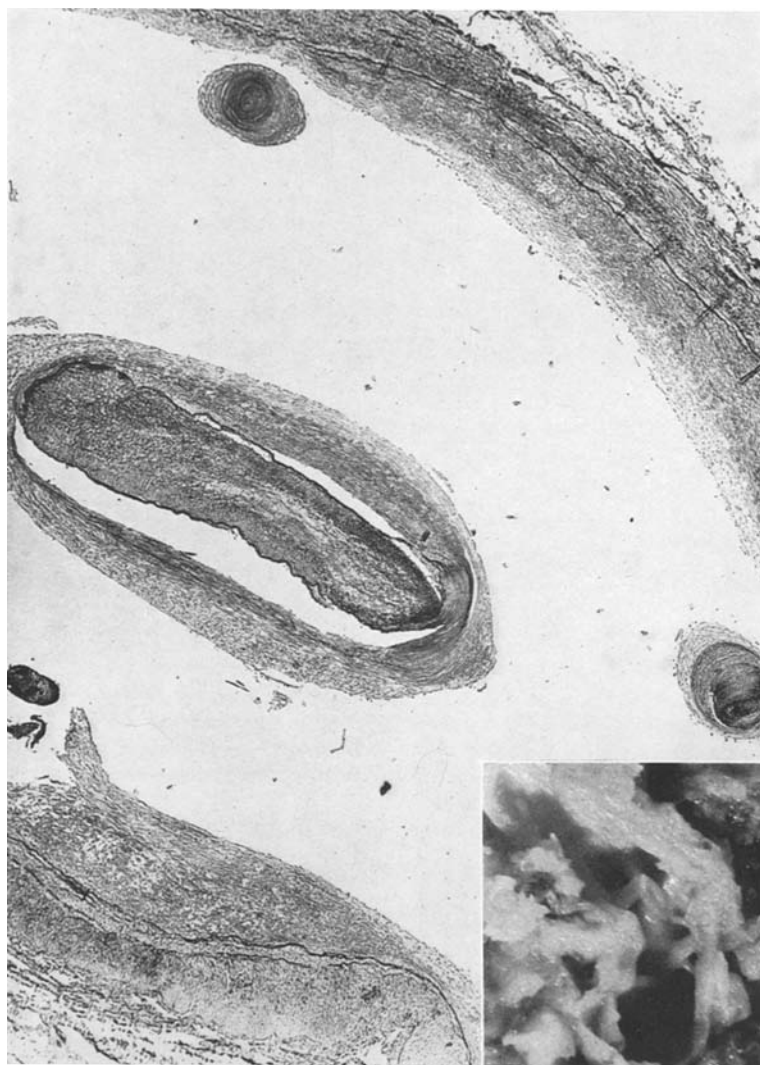
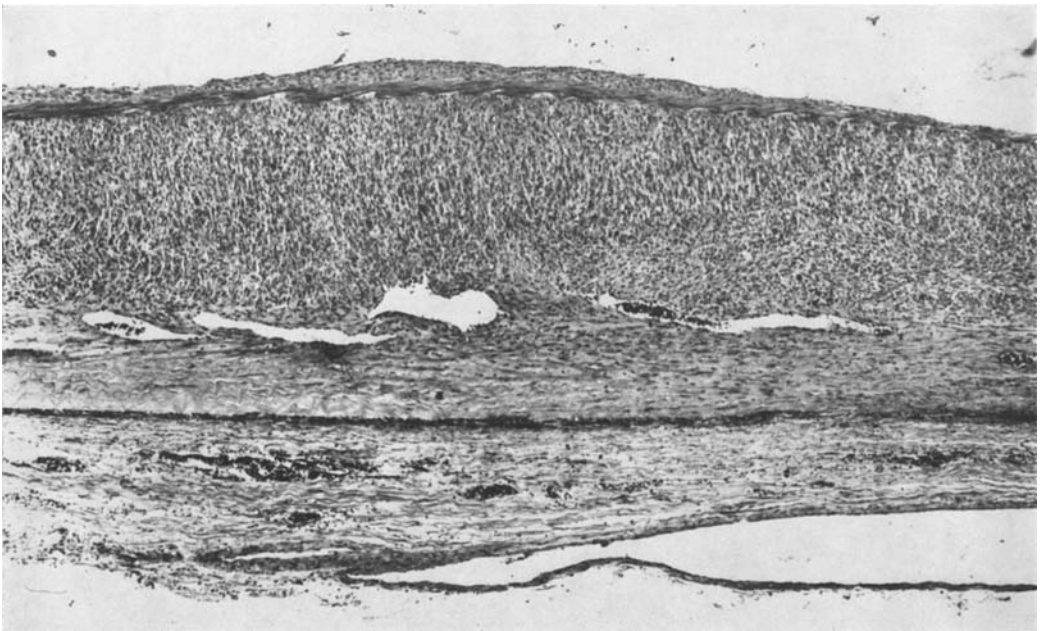


Fig. 2. Insert at the low right corner: Transluminal bands. Rest of the figure: Histological section of the band, showing the organoid structure of a vessel wall with media, internal elastic membrane and intima. Paraffin, van Gieson-Elastica, photomicrogramme, enlargement  $2.5 \times 10$

exhibited the same organoid structure as the vertebral artery, the only difference being that there was no adventitia.

Both of these structures might, therefore, be considered as a protrusion of the media into the lumen. Obviously, any outfolding of the media must be covered by an internal elastic membrane and an intima.

In addition, the media was found to be markedly thickened in several transverse sections at different levels. The direction of the muscle fibers was not circular as usual, but predominantly longitudinal. Within these layers of longitudinal muscle fibers, clefts had formed, which were lined by an endothelium and filled with erythrocytes, and which somewhat resembled a cavernoma. More-



a

Fig. 3. a In the outer part of the media large cavities, lined by a thin epithelium, which resembles somewhat cavernomatous structures. Note the abnormal longitudinal direction of the muscle fibers and the enlarged venous vessels in the adventitia. Paraffin, van Gieson-Elastica, photomicrogramme, enlargement  $4 \times 10$ . b Section through tortuous arterioles with media cells showing epitheloid transformation. Note the abnormal longitudinal direction of the muscle fibers. Paraffin, van Gieson-Elastica, photomicrogramme, enlargement  $4 \times 10$

over, clusters of tortuous arteriolar vessels with thin endothelium and a thick cellular media were encountered. The cells of the media of these arteriolar structures were of epitheloid character. Fibrous thickening of the adventitia was seen in both vertebral arteries; occasionally cavernomatous cavities were also observed. Neither myelinated nor unmyelinated nerve fibers were demonstrable by special stains in the sites with abnormal structure of the vascular wall.

Routine sections of the aorta and art. brachialis did not reveal corresponding abnormalities of their media.

### Discussion

Peculiar abnormalities of the structure of the vascular wall of the vertebral arteries were encountered in the case described.

There were two different types of malformations: On one hand, increased amounts of abnormally textured vascular wall tissue with an organoid structure were observed. The excess tissue had the macroscopic appearance of valves and bands. On the other hand, tortuous arteriolar vessels and venous cavities were demonstrable in the outer part of the media, in which the direction of muscular fibers was distinctly abnormal.

The latter finding might, perhaps, suggest the presence of true a-v anastomoses. This suspicion might even be supported by the epitheloid character of



Fig. 3b

the media muscle cells, resembling those seen in glomera digitalia (Hoyer, 1877; Grosser, 1902; Schorn, 1959) and other a-v anastomoses. The media cells in these locations are “epitheloid modifizierte Muskelzellen” (i.e. muscle cells with epitheloid transformation). (Lit. see Clara, 1956; Schorn, 1959; Hammersen and Gross, 1968.) However, two arguments can be adduced against the interpretation of the above findings being a-v anastomoses.

According to Staubesand (1968) the presence of anastomoses is not proved unless demonstrated in an uninterrupted sequence of sections.

In our case we were unable to show a-v connections by this technique.

In addition, with the usual special stains we were unable to demonstrate myelinated or unmyelinated nerve fibers, the presence of which is obligatory in a-v anastomoses according to Popoff (1934), Masson (1937) and others.

We, therefore, conclude that the epitheloid character of the medial muscle cells is insufficient evidence to interpret these changes as true a-v anastomoses.

The interpretation of the vascular valve and of the transluminal bands with their peculiar organoid structure presents some difficulties. One might consider the possibility that a contraction of these structures leads to a certain narrowing of the vascular lumen, although the functional meaning of this would remain unclear.

It is well known that the atlas-loop (Atlasschleife) of the vertebral artery is subjected to traction and shearing forces during rotational movements of the head. This is particularly true for the caudal part of the atlas-loop. It is

in this very site where transluminal trabecular bands were found in our case. One might speculate that these trabecular bands act as a guy line, resembling the so-called "moderator bands" of the right ventricle of the heart.

In spite of the above arguments it does not seem justified to attribute any functional significance to the vascular valve and to the dysplasia of arterial and venous vessels in the media of the vertebral arteries of our case.

We, therefore, conclude that the above abnormalities of the vascular wall architecture represent a hamartomatous malformation, so to speak a freak of nature. We, therefore, propose to call this finding: vaso-muscular mediadysplasia of the arteria vertebralis.

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