Absence of Elastica from Inner Half of Aorta

Multiple Dissecting Aneurysms

By H. EDWARD MACMAHON, M.D., AND WILLIAM J. CHAMBERS, A.B.

SUMMARY

Multiple dissecting aneurysms were found in the course of an autopsy on a 52-year-old male patient who had expired suddenly as a result of a massive pericardial tamponade. Two of these aneurysms involved the aorta. The others involved muscular arteries, namely, the superior mesenteric and the splenic. In the case of the aorta, there was an absence of elastica from the entire inner half of the media, leaving the fabric of the outer half intact. The course of dissection in the case of each aneurysm lay along the line of demarcation separating the inner from the outer half of the media. This very unusual pattern, showing an absence of elastic lamina in the inner half of the media, differs remarkably from the changes that are commonly seen in dissecting aneurysms of this large elastic artery. Neither the etiology nor pathogenesis of this defect is known. The recognition that such a disease exists is only the first step toward its solution.

THE absence of elastic laminae in the inner half of the media of an aorta from a 52-year-old male hypertensive patient, the subject of several dissecting aneurysms, is the basis of this report. Dissecting aneurysms of the aorta are relatively common in man and have been found in animals.[1] They may be found in association with aneurysms in other parts of the body. They may appear at any age, but they are most common in the middle-aged hypertensive patient. They are quite distinct from arteriosclerotic, luetic, traumatic, and septic aneurysms.

While cases of dissecting aneurysms of the aorta may be found in which neither gross nor histological lesions in the wall are obvious, there is usually some underlying disease. The most common predisposing factor may be described grossly as a loss of the normal resilience of the wall and histologically as a combination of regressive changes characterized by a looseness in the texture, an accumulation of ground substance, often showing an unusual degree of metachromasia, a thinning or fraying of elastic lamina, atrophy of smooth muscle fibers, and at times foci of frank medial necrosis. Such changes are commonly grouped together and referred to as "arterio-medio-necrosis."[2, 3] Such a pattern was not present in the wall of the aorta under consideration. Instead, there was an absence of elastica in the inner half of the media, leaving the fabric of the outer half essentially unchanged.

Report of Case

The patient, a white maintenance worker, was suddenly seized while at rest with an excruciating pain over the precordium. This radiated to the back and down the left arm. Within a few hours it had shifted into the abdomen. The pain was accentuated on inspiration. A few minutes after the onset, the right leg became numb, but this was only temporary. The patient vomited repeatedly and experienced several dizzy spells. The blood pressure was 200/110, pulse 115. An ECG and X-ray of the chest were unremarkable. The BUN and SGOT were normal. The WBC count of 16,700 with 92% polymorphonuclear leukocytes (PMN) rose to 19,700 with 66% PMN. There was a past history of "dyspepsia" after spicy and fatty foods, but there was no record of hematemesis or melena. With increasing pain, weakness, sweating, and pallor the patient was transferred on the fifth day of illness to the New England Medical Center.

A physical examination on admission revealed an unusually muscular, well-nourished, well-hydrated, middle-aged male weighing 210

From the Tufts-New England Medical Center, Boston, Massachusetts 02111.

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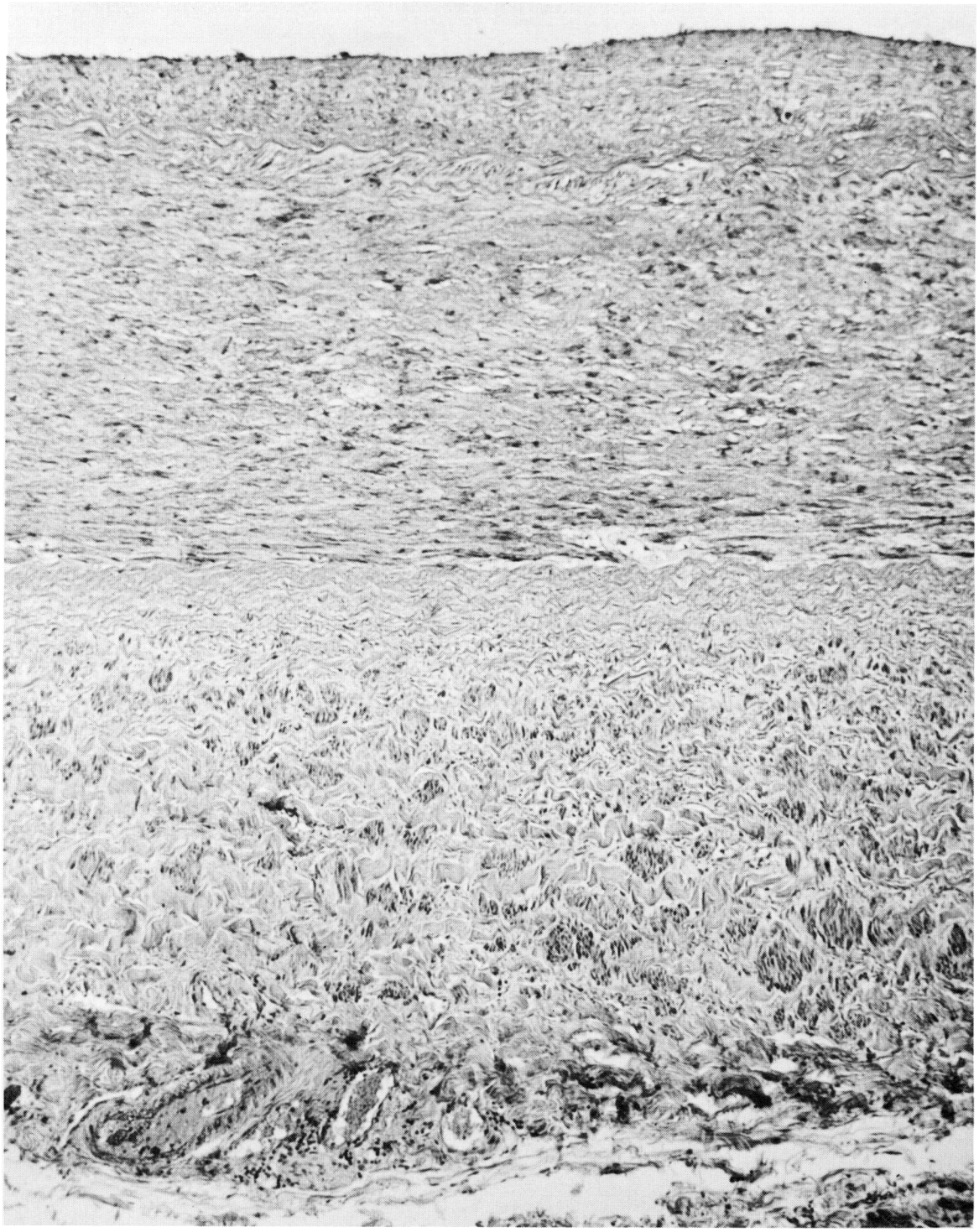

Figure 1

Wall of aorta (hematoxylin and eosin stain, × 100). The intima lies above, composed of loose fibrous tissue rich in ground substance. A clearly defined, solitary, internal elastic lamina separates the intima from the media. The media is about equally divided into an inner half,

pounds. The heart rate was rapid and irregularly irregular. The blood pressure was 160/100, pulse 100. There was extreme tenderness in the left upper quadrant, in the umbilical area, and in the left lower quadrant. There was no rebound tenderness. Breath sounds were distant and free of rales and rhonchi. Inspiration and expiration were equal. A complete blood count was within normal limits, as were the BUN, electrolytes, and urinalysis. Temperature was 103 F. Late on the day of admission, the patient experienced sudden cardiac arrest and despite resuscitative measures failed to respond. Clinical diagnoses included dissecting aneurysm of the aorta, atrial fibrillation with systemic embolism, acute pancreatitis, and perforated ulcer.

Permission for an autopsy was granted. There was no thrombosis, no embolism, no pancreatitis, and no ulcer. A dissecting aneurysm developing from a transverse tear, 3 cm above the sinuses of Valsalva, had ruptured into the pericardial cavity, leading to compression of the right atrium. There was upward extension of the dissection along the innominate artery as far as its bifurcation and forward extension as far as the left common carotid artery. There was a second and very much larger aneurysm, also arising from a transverse tear, 1 cm below the level of the ligamentum arteriosum. Several centimeters of intact aorta separated these two aneurysms. The larger had dissected downward along the entire length of the aorta and along the left iliac artery as far as the inguinal ligament. There was also an extension of this dissection along the course of the renal arteries. The aorta showed only minimal arteriosclerosis, and at the sites of transverse tears, there was none. More important was a loss of elasticity of the aorta. It had little or no resilience, and the wall could be stripped longitudinally as one might peel bark from a birch tree.

In addition to the aortic aneurysms, there were two others involving the splenic and superior mesenteric arteries. Each of these was small, bulbous, and measured 2 to 3 cm in length. The splenic aneurysm was older and partially calcified; the other was very recent.

Histologically, the aorta was of particular interest. The intima was slightly thickened, hypocellular, and rich in basophilic ground substance. Immediately adjacent was a coarse, interrupted, sharply defined, wirelike internal elastic lamina. An internal elastic lamina of this type is not found in a normal aorta. Beyond this lay the anomalous media. The inner and outer halves were strikingly different. The inner half was devoid of stainable elastic laminae. It was composed of degenerate collagen which in areas stained as collastin, an amorphous ground substance, and scattered atrophic smooth muscle fibers. In areas one could distinguish shadowy, unstained outlines of what suggested residual elastic laminae. The outer half of the media stood out in sharp contrast simply because it was essentially normal (fig. 1). The transverse tears which were responsible for the aneurysms crossed the intima, internal elastic lamina, and elastic-free inner half of the media. Each tear terminated abruptly at the level of the outer half of the media. The lines of dissection lay along the line of demarcation separating the inner and outer halves of the media (fig. 2). In the iliac artery and renal arteries, the dissection continued along the line of separation between media and adventitia.

The small aneurysm of the superior mesenteric artery, which is normally regarded as a "muscular artery," was associated with a small knifelike cut which crossed the intima, internal elastic lamina, and the entire thickness of the media. The adventitia supporting this vessel was rich in collagen and elastica. This remained intact. A small pool of freshly coagulated blood dissected the media away from the adventitia, leading to compression of the lumen. The media in this segment of the artery was distinctly abnormal. The width varied, and instead of the normal compact concentric layering of muscle, the wall was scarred and in places stained as collastin. Muscle fibers were reduced, and ground substance was increased. The picture was one of a degenerate vessel showing a fresh transverse tear and a localized intramural hematoma. In the case of the splenic artery, the histology was much more complex. The aneurysm, being much older, showed signs of both injury and repair. Although there was no atherosclerosis along the course of this vessel, there was an abundance of lipid-laden material, cholesterol crystals, calcium, and old fibrin in the area of hemorrhage. The lesion resembled an old atherosclerotic plaque complicated by recurrent hemorrhage. The lumen in this area was narrow but not totally obstructed.

It might be of interest to add that minor developmental anomalies were found in this patient. These included scattered cysts of one kidney, a solitary diverticulum of the bladder,

free of elastic lamina and composed of loose fibromuscular tissue embedded in basophilic ground substance, and an outer half composed of elastic laminae together with an abundance of collagen and smooth muscle. The adventitia at the base contains several small vessels.

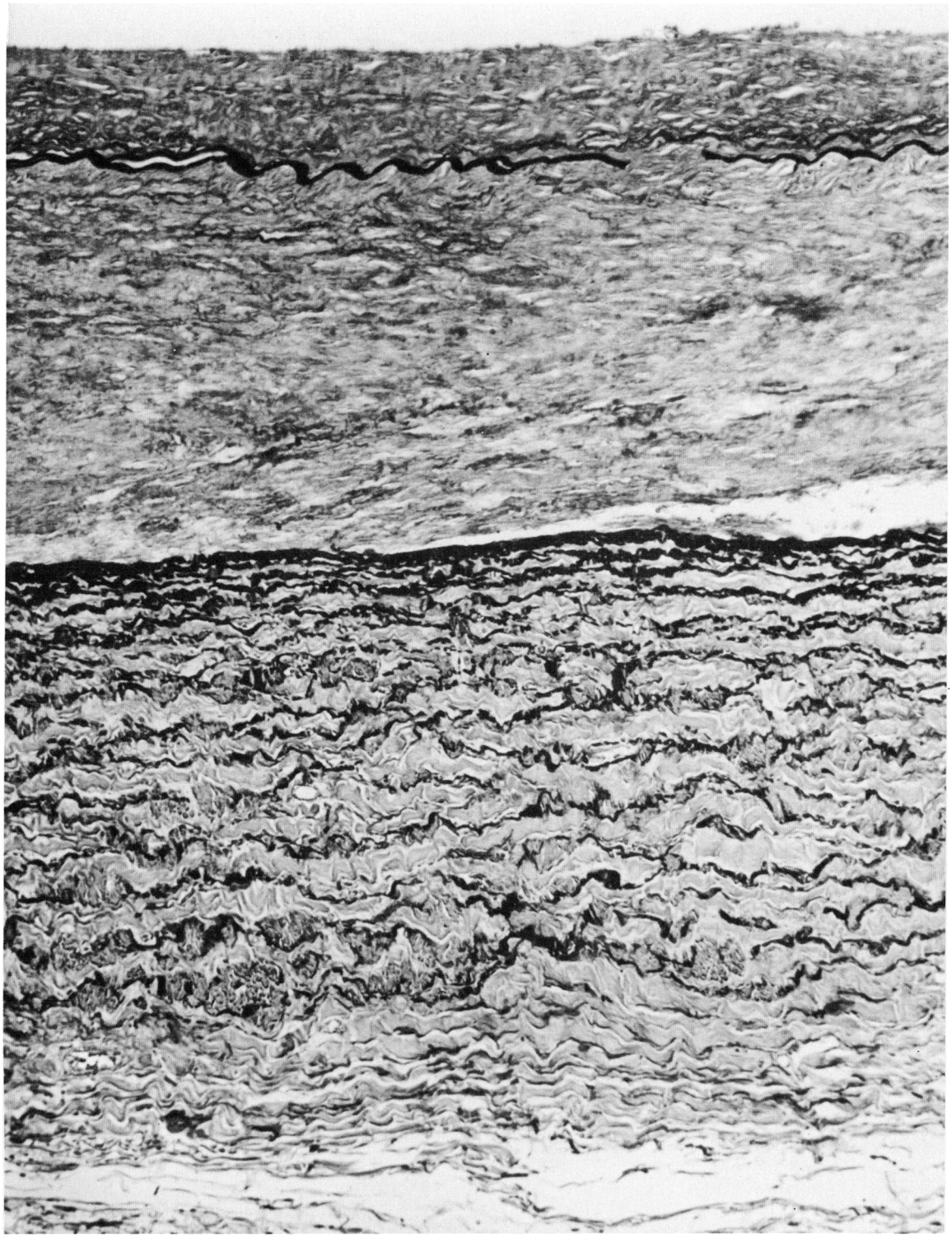

Figure 2

Wall of aorta (Verhoeff's elastic tissue stain, × 100), showing the line of dissection of the aneurysm at the right. This corresponds exactly with the line of demarcation between the

and an island of ectopic pancreas in the wall of the stomach.

Discussion

From the autopsy findings it was apparent that the large aortic aneurysm was the older, and by compressing the renal arteries, it could have raised the blood pressure which in turn could have been an important factor in causing the second aneurysm that was responsible for the death of the patient. In any case the autopsy findings offered a rational explanation for the clinical signs and symptoms and sufficient gross and histological evidence to account for the nature and extent of the two aneurysms. What the autopsy did not provide was a solution to the problem concerned with the etiology and pathogenesis of the disease of the walls of the arteries which was basically responsible for the aneurysms.

Most spontaneous dissecting aneurysms of the aorta are associated with recognizable degenerative changes in the media, and of these, "arterio-medio-necrosis" is most frequent. Such regressive changes may occur as a primary disease confined to the aorta. They may also represent a vascular manifestation of such inheritable diseases of connective tissue as Marfan's syndrome,[4] the Ehlers-Danlos syndrome,[5, 6] and the syndrome associated with homocystinuria.[7] They may also be found as a manifestation of experimental lathyrism,[8] and in this group, dissecting aneurysms of the aorta are not uncommon. Added to these are cases of dissecting aneurysm associated with the rheumatoid diseases of the aorta[9] and giant-cell aortitis.[10, 11] More recently, certain ganglionic and neuromuscular blocking agents used to lower blood pressure, such as methonium and pentolinium, have been implicated as possible etiological agents.[12]

In the case under consideration, the unusual pathological histology of the media of the aorta does not resemble the pattern of arterio-medio-necrosis, nor does it resemble in any way the rheumatoid diseases of the aorta or the granulomatous forms of aortitis. Furthermore, there was no evidence that a drug could have been responsible, and yet, in the medicated society of today, the possibility of chemicals playing some role in the initiation of this unusual disease of the elastica cannot be excluded.

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elastic-free and the elastic-rich layers of the media. The narrow intima is clearly separated from the media by a single coarse interrupted internal elastic lamina. The adventitia is composed of little collagen and delicate elastic lamina. This is seen at the base.

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