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Abstract
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SOME PATHOLOGICAL ASPECTS OF DISSECTING ANEURYSM

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By M. J. MACLEAN

The term dissecting aneurysm implies the development of a circulatory pathway between the layers of the vessel wall. It can occur at all ages and in both sexes, being most frequent in men, in the fourth, fifth, and sixth decades, and in women in the eighth and ninth decades. It does not occur in normal arteries.

Aetiology

The aetiological factors are variable and largely hypothetical but the following theories appear to have substantiating evidence of their authenticity:

(1) **The mechanical theory.** Physical and mental strain causing a sudden increase in blood pressure is sufficient to produce the medial rupture leading to dissecting aneurysm only when the vessel wall is diseased. About 40 per cent. occur after mild exertion, although this as an aetiological factor is disputed by others. Hypertension which may be of the essential variety but is usually of renal origin is present in over half the cases. Mechanical injuries such as violence and traumatic accidents probably do not produce dissection.

(2) **Congenital deformities,** including coarctation, bicuspid aortic valves, and hypoplasia. 16 per cent. of cases of coarctation of the aorta develop dissecting aneurysm.

(3) **Pregnancy.** In 83 per cent. of one series of investigations, dissection occurred antepartum and thus cannot be attributed to the strains and blood pressure changes occurring during labour.

(4) **Diseased aortic wall.** Disease of the intima rarely causes dissecting aneurysm and those beginning at atheromatous plaques or ulcers are usually restricted in extent. An abnormal tunica media may arise through disease of the vasa vasorum or be essential in character. The degeneration, which may be local or diffuse, is a chromatotrophic or mucinous process probably involutional or senescent in nature and it may progress to cyst formation. There is no connection between syphilitic mesaortitis and dissecting aneurysm and the joint occurrence of the two conditions is merely coincidental. Since the reaction of human tissue to the *Treponema pallidum* is mainly one of fibrosis, the lesion in the aortic wall is localised rather than generalised.

Pathology

The primary rent in the aneurysm, usually a few centimetres in length, occurs in the aortic intima which is usually normal at the site of the tear. This would suggest that the tear is an effect rather than a cause of dissection. The most frequent sites of the tear are, firstly, the ascending aorta, accounting for 50 per cent.. of cases of dissection, secondly, the junction of ascending and transverse portions for 20 per cent., and thirdly, the junction of transverse and descending portions for 10 per cent., the dissection being in the descending thoracic or abdominal aorta. The length of the dissection

varies from a few centimetres to the entire length of the aorta, occasionally extending down as far as the popliteal arteries.

After the tear blood penetrates radially to a point between the outer and middle thirds of the aortic wall and a cleavage plane is created between the elastic laminae. Dissection usually continues in the direction of intraluminal flow but may be directed towards the heart, particularly in the first two groups, for 10 per cent. of the ruptures in those situations do so. Secondary rupture may occur along the aortic wall either back into the lumen or more frequently outwards, with fatal results.

The pathological lesion may be of several distinct types. Common to the great majority of instances is a process which is independent of the changes in the vasa vasorum or the intimal coat of the aorta and which is unaccompanied by cellular inflammatory changes. When reacting cells are present they are usually few and appear to be secondary to the degenerative process. This degeneration is much more marked than is the normal loss of smooth muscle with age. The outer third of the media contains the largest number of smooth muscle cells, which is worthy of note for it is in this outer third of the media that cystic areas, in which all the elements are degenerated, are so often found.

The various types of medial lesions that have been observed in this process, referred to as medionecrosis, are as follows:

(1) Primary degeneration of the elastic lamellae may occur in the form of a fatty metamorphosis, fragmentation, or necrosis, with some damage to the supporting collagen or muscle fibres, with or without mucoid accumulation, or

(2) Sometimes hyaline degeneration of the interlamellar connective tissue takes place, or

(3) Non-exudative necrosis of muscle cells proceeding from simple nuclear loss to extensive structureless homogenisation of the muscle cells and the adjacent collagen and elastic fibres, with or without mucoid accumulation, and

(4) Primary over-production of mucoid substance in the interlamellar ground substance with encroachment over the muscle, elastic and collagen fibres, with the ultimate development of cysts.

An actual quantitative reduction of the medial tissue elements linked with marked thinning of the vessel wall, may be associated with any of the preceding degenerative lesions, a process often found in Marfan's syndrome and thought to be an inherited mesodermal defect. In cases in which pronounced mucoid accumulation with cyst formation is observed these appear most often to involve the thoracic aorta, particularly the ascending portion of the arch.

Areas of destruction of medial tissue may be replaced by poorly vascularised fibrous scars or by regenerated muscle or elastic tissue. The removal of necrotic tissue is effected through a humoral route and some acellular zones of necrosis show no tendency at all towards replacement by a substitute tissue.

Pathogenesis

The mode of production or development of the disease can be found in the answers to three questions:

(1) What factors determine the rupture and the site of rupture of the tunica intima?

(2) What factors lead to the degeneration of the media?

(3) What factors determine the final course of the dissecting process, either out through the tunica adventitia or back through the tunica intima?

Dealing firstly with the mechanics of the initial intimal tear, the influence of an actively contracting hypertrophied left ventricle in producing a high systolic blood pressure is evident, and has to be accepted as an important factor in elongating and distending the aorta, especially in its ascending part. Moreover the direction of the blood stream is altered quite suddenly as it passes from ascending to transverse part, and again from transverse to descending portion, so that the wall of the vessel has to withstand a greater strain than elsewhere. This should be more marked on the greater curvature of the aorta than on its lesser curvature. The systolic propulsive force which is exerted chiefly longitudinally, parallel to the axis of the lumen, will have greater effect in elongating the vessel if any irregularities are present like atheromatous plaques on the inner surface which will increase frictional resistance.

Such irregularities are not as common in the ascending aorta as in the transverse and descending portions; but on the other hand primary rupture occurs much more commonly in the ascending aorta than at the junction with the transverse part or in any part of the extrapericardial aorta. This can be attributed to two reasons. Firstly, the longitudinal force is greatest in the ascending portion, as is borne out by the frequency of circumferential tears in this region, for the direction of the linear tear will be perpendicular to the direction of the preponderant stretch. Secondly, the abrupt diastolic recoil meeting the resistance of the closed aortic valves is of even greater importance as a factor in producing the primary tear. In diastole on closure of the valve, the longitudinal force is largely converted into a transverse one with consequent lateral stretching of the intrapericardial aorta. Additional factors are the resistance offered by the distended pulmonary artery and by the rigid attachment of the pericardium at its reflection, both of these tending to increase the strain which has to be borne by the intrapericardial aorta.

In consideration of the second question, the factors producing the degeneration of the media, we find that the cause of cystic medionecrosis is still unknown. Medionecrosis of the aorta has been produced experimentally in rabbits using the intravenous injection of diphtheria toxin. Intravenous tyramine injections also produced medial necrosis in some of the animals. Similarly necrosis has been produced in rats by feeding the sweet pea, the toxic component of which is Beta-amino proprionitrile. A medial degeneration has also been produced in guinea pigs and rabbits by deprivation of Vitamin E. Cystic medionecrosis may be found in patients dying from various causes and there is some association with age but none with sex or hypertension. Cystic necrosis may occur without dissection, but appears to be necessary for its development. It may result from exhaustion of the muscle in its efforts to prevent overdistension of the aorta, or an excess of adrenaline might cause spasm of the vasa vasorum, focal ischaemia and necrosis. Whatever the cause the media generally gives way before the intima.

The factors determining the final course of the dissecting process or secondary rupture are (1) weakness of the outer wall, (2) atheromatous plaques weakening the inner wall, and (3) the presence of normal anatomical structures such as aortic branches which tend to hinder continued dissection. Secondary rupture outwards may occur at any point along the aortic wall, but most frequently into the mediastinum. This occurs in 85 per cent. of cases, the other 15 per cent. dissecting back into the lumen.

The pathogenesis in subjects under the age of forty can generally be attributed to one of the congenital lesions already described. In 50 per cent. of these there is also an abnormality in the connective tissue, but it is unlikely that medionecrosis develops long before dissection occurs.

It is not known whether a true cause and effect relationship exists between pregnancy and dissecting aneurysm but, if it does, it may be due to the hormonal changes which may weaken the aortic wall, rendering it more susceptible to dissection.

Hypertension is usually present in those over forty suffering from dissecting aneurysm. Dissection is, however, rare in cases of malignant hypertension, its incidence being 0·2 per cent. (Beaven and Murphy, 1956). These authors report dissection in nine hypertensive patients being treated with methonium drugs, six of whom were in the malignant phase. Since the incidence is greater than would be expected by chance, some workers have suggested that the methonium compounds might play a role in the aetiology of dissecting aneurysm via the wide fluctuations in blood pressure which occur in therapy, these producing additional stress on the aortic wall. This observation is of value since physicians may mistake this complication for a myocardial infarction, withhold hypotensive therapy which may be of some value, and administer anticoagulants which further reduce the remote chance of survival.

Conclusion

In conclusion, the types of patients liable to develop dissecting aneurysm are:

1. Those with coarctation or extreme hypoplasia of the aorta.
2. A few patients with atherosclerosis of the aorta with ulceration of the intima which permits dissection to begin at this point.
3. Others who develop it through disease of the vasa vasorum of unknown aetiology with weakening of the media.
4. In the majority of cases no aetiological factor is found. They present no specific disease of the aorta, *in the absence of microstudies.*

In brief then, no single causative agent or morbid process can be demonstrated as consistently producing the vascular changes leading to dissection. Death in 70 per cent. of cases occurs from rupture externally, and there is little relation between the site of tear, the length of the dissection, and the time of survival.