Is Cystic Medionecrosis the Cause of Dissecting Aortic Aneurysm?

OVER FOUR DECADES have elapsed since Erdheim described the aortic lesion currently referred to as idiopathic cystic medionecrosis, cystic medionecrosis, and often abbreviated to medionecrosis of the aorta. In two articles, each based on a single case, he described in meticulous detail the histologic lesions encountered not in dissecting aneurysm but in spontaneous rupture of aneurysm of the ascending aorta. He did not regard the lesions as unique, and gave credit to other authors who had previously described similar changes.

His first case report emphasized the accumulation of mucoid substance in cysts which interrupted the continuity of the muscle and elastic tissue of the media. He noted that the lesions were essentially noninflammatory and were not associated with alterations in the intima, adventitia, or vasa vasorum. In his second publication, he stressed the lack of a healing reaction, and the resultant impairment in strength which predisposed to aneurysmal dilatation and rupture.

In the intervening years, the terms employed by Erdheim have become accepted as typical of the histologic lesions encountered in dissecting aneurysm, and are considered by most authors of current textbooks of pathology as the chief predisposing cause for dissecting aneurysm of the aorta. Erdheim's designation or an abbreviation thereof frequently appears on examinations to test the knowledge of vascular disease of both students and physicians.

Despite the overwhelming acceptance of Erdheim's terms, there is considerable doubt that these designations accurately describe the lesion or that cystic medionecrosis is the most frequent lesion encountered in dissecting aneurysm. The term "cystic" is inappropriate, since the lesions do not form true cysts with a distinct lining, but rather represent structural faults or voids in the media which have become filled with the semifluid ground substance as a result of intramural tension. The term "medionecrosis" is not justified because, as Erdheim himself admitted, necrosis is seldom encountered in the lesions, but is inferred as a result of depletion of the cellular elements of the media.

While Erdheim employed the term idiopathic to indicate an ignorance of the etiologic factors in the medial lesion he described, recent studies have provided some insight into the pathogenetic mechanisms involved. Although the majority of human cases must still be categorized as idiopathic, future investigations may permit the identification of a precise etiology, an essential step in the prevention or inhibition of medial degeneration and its serious complication, dissecting aneurysm.

Some form of degeneration of the media is generally considered a basic requisite for the development of dissecting aneurysm, since this normally strong layer provides the chief support for the aortic wall. In most instances, medial lesions can be categorized as involving primarily either muscle or elastic tissue depending on which degeneration predominates. Future studies may reveal alterations in the ground substance or collagen as well.

Considering the close structural relationship of the muscle and elastic tissue of the media, their metabolic independence is unique. The muscle layers, composed of metabolically active cells, are dependent on a steady flow of nutriments and oxygen for survival. Intimal thickening, particularly that due to atherosclerosis, may interfere with diffusion and permeation into the inner media, while nutrition to the outer media may be compromised by sclerosis of its nutrient vessels, the vasa vasorum. Both intimal thickening and sclerosis of the vasa may be aggravated by longstanding hypertension. It is significant that the vasa vasorum are most abundant in the ascending aorta and arch, precisely the segments most susceptible to the development of dissecting aneurysm. It has been well documented that muscle lesions of the media tend to increase with age, especially after forty, and in patients with hypertension, thus correlating well with the known occurrence of dissecting aneurysm. Some large series of dissecting aneurysms subjected to careful histologic studies have revealed that muscle lesions are considerably more frequent than elastic tissue lesions.

Loss of muscle cells is usually focal, involving one stratum in the vessel wall. In such an acellular zone, the elastic laminae lose their normal sinuous pattern and appear straight and attenuated as though stretched by the loss of supporting muscle cells. Muscle lesions do not tend to accumulate mucopolysaccharide and are therefore readily distinguished from the mucoid lesions. Because muscle lesions occur approximately as frequently in the absence of aortic dissection as they do with it, some are reluctant to attribute

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etiolologic significance to this type of change. Although muscle lesions undoubtedly weaken the aortic wall, the degree to which these lesions impair the intramural cohesive strength is presently unknown.

In contrast to the muscle lesions, elastic tissue lesions are encountered in a younger age group, most often below 40, and particularly in Marfan's syndrome, suggesting a relationship to hereditary defects or perhaps a subtle biochemical defect. Apparently ischemia plays little or no part in the degeneration of elastic tissue, since this tissue persists for prolonged periods in the absence of a blood supply, a characteristic which has made feasible the use of human aortas for homografts. The ability of elastic tissue to stretch and retract without expenditure of energy or oxygen requirements makes it an ideal constituent for the proximal portion of the aorta which pulsates with cardiac systole and diastole. Elastic tissue defects are characterized by interruption of one or more laminae of the media leaving microscopic voids, often multiple, which become filled with ground substance. As a result of loss of elastic tissue into which they normally insert, muscle cells lose their normal parallel orientation. The result is precisely the type of lesion that Erdheim described, and which he apparently regarded as the result of a primary increase in ground substance. Such accumulations are currently considered as a compensatory or secondary phenomenon, i.e., an ineffectual attempt to restore the structural strength of the media.

There is little doubt that the disorganization of the media due to the loss of elastic tissue greatly reduces the cohesive and perhaps also the tensile strength of the media. It seems reasonable to suggest that the added stress resulting from loss of elastic support accelerates the deterioration of the muscle cells.

Only infrequently does dissecting aneurysm arise from inflammatory disease of the vessel wall or advanced atherosclerosis, conditions which also share the common denominator of medial structural weakening, but must be clearly distinguished from the more common histologic lesions which are found in medial dissections.

Although a review of large series of dissecting aneurysms suggested that the frequency of "cystic medionecrosis" ranged from 0–83%, it is likely that the wide discrepancy represents a difference in the threshold for the diagnosis among pathologists, many of whom accept a slight increase in ground substance in the absence of other structural alterations as diagnostic, particularly when the ground substance is bubbly or vacuolated. Such observations have usually been made in the absence of control studies which would have revealed that this feature is often encountered in otherwise normal aortas. Furthermore, the section of aorta is often taken at random along the course of dissection, under the false assumption that the medial lesion is necessarily diffuse. As Prokop et al. have shown, once dissection is initiated in a vessel it may extend distally into segments which are histologically normal, since propagation is dependent on the presence of a pulsating pressure, and particularly the steepness of the pulse wave. Obviously, efforts to find a predisposing lesion should be concentrated on the proximal aorta, particularly in the region of the intimal tear, where medial lesions are most likely to be demonstrable.

A critical appraisal of the literature and ample personal experience lead to the conclusion that medial elastic tissue lesions of the "mucoid cystic" type are not a common cause of dissecting aneurysm. Such lesions are found most consistently in dissecting aneurysms associated with Marfan’s syndrome, which comprise less than 3% of the total cases and in the rare aneurysms of the ascending aorta occurring in the absence of syphilis or atherosclerosis. It seems advisable to abandon the term idiopathic cystic medionecrosis in favor of terminology which designates the defective component(s) of the vessel wall, as either muscle or elastic tissue or both.

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