Healed Dissecting Aneurysm in Cystic Medial Necrosis of the Aorta

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Dissecting aneurysm of the aorta is a not uncommon disease that usually produces severe symptoms and often leads to an early death. This article reports an unusual case in which the patient was nearly asymptomatic and died of unrelated causes. Autopsy revealed a healed dissecting aneurysm. The literature on dissecting aneurysm, emphasizing its pathogenesis and its relation to cystic medial necrosis, is briefly reviewed.

Dissecting aneurysm of the aorta has long been recognized as a disease entity, and is ordinarily regarded as a rapidly fatal condition. A certain proportion of dissecting aneurysms redissect into the lumen of the aorta, forming the so-called “double-barreled” aorta. Occasional cases of dissecting aneurysm have survived for a considerable period after the acute episode, many of them being of the “double-barreled” variety. Most of the reported prolonged survivals suffered from congestive heart failure and eventually died as a result of the aneurysm. Few cases have been reported in which the dissecting aneurysm was essentially an incidental autopsy finding of relative clinical insignificance. The following case report is presented as an instance of chronic, nondisabling dissecting aneurysm associated with idiopathic cystic medial necrosis.

Case Report

Clinical Findings

The patient was a 43-year-old white male military officer. In 1950, a malignant melanoma was removed from his left shoulder. He had no further difficulties until he suffered transient episodes of vertigo and blurring of vision in February and June 1953. A physician at that time found no cause for the vertigo. In March 1954 a cardiac murmur was first noted. In August 1954 two more episodes of vertigo were followed by syncope. At this time examination showed atrial fibrillation and the patient was hospitalized. The rhythm reverted to normal after digitalization and the patient returned to duty. Fibrillation recurred in October 1954 and was again controlled with digitalis. He was hospitalized on September 5, 1955, for study of his cardiac condition.

The patient denied any history suggesting acute rheumatic fever. He had been in the military service continuously since the age of 20 and had enjoyed good health. He had no history of chest pain, dyspnea, or palpitation. The patient underwent a tonsillectomy in childhood and an appendectomy at the age of 21. At the time of the appendectomy physical examination of the heart was normal; blood pressure was recorded at 138/58. Subsequent determinations, however, revealed a normal pulse pressure. In the family history, the patient’s mother had died of heart disease at 59.

On admission to another Air Force hospital in September 1955, the blood pressure was 124/40. Grade III systolic and diastolic murmurs were heard over the aortic area, radiating to the neck, and the peripheral signs of aortic insufficiency were present. Chest x-ray showed minimal cardiac enlargement and widening and tortuosity of the aorta. An electrocardiogram showed normal sinus rhythm, left ventricular hypertrophy, and digitalis effect. Routine blood counts and urinalyses were normal. Serologic test for syphilis was negative. The basal metabolic rate was plus 1, and an electroencephalogram was normal. Shortly after admission a complete neurologic examination revealed a left pupil slightly larger than the right. The patient’s syncopal attacks were attributed to his heart disease, which was thought most probably to be rheumatic in origin.

While in the hospital the patient began complaining of headaches and of flashing lights in his field of vision. Over a period of a few days he became markedly lethargic. On October 22 papilledema was noted, as well as asymmetrical reflexes in the lower extremities, and a right Babinski sign. A ventriculogram showed a space-occupying lesion in the right cerebral hemisphere. On craniotomy a walnut-sized tumor was found under the right premotor cortex and it was enucleated. Pathologic diagnosis was metastatic tumor, probably malignant melanoma. The patient made a good recovery from the operation, complicated only by recurrent infection of the urinary tract. During hospitalization the blood pressure averaged 130/40.

On December 2 the patient was transferred to Parks Air Force Base Hospital. Admission blood
pressure was 120/40. The patient was oriented and alert, and showed a left hemiparesis. Cardiac findings were as previously described. Fine rales were noted at the left base posteriorly. Routine laboratory studies were negative except for evidence of a urinary tract infection. Chest x-ray showed for the first time a solitary, circumscribed lesion in the right lower lung field. The heart was minimally enlarged. An electrocardiogram showed atrial fibrillation and digitalis effect. Over a period of about 2 months the patient's neurologic status gradually improved on a regimen of physiotherapy, and he began to walk. He had no complaints referable to the cardiovascular system. Late in February, however, his general condition began to deteriorate. A new pulmonary lesion was noted, and hard nodes appeared in the neck. He died on February 23, following a generalized seizure.

Autopsy Findings

Evidence of the healed craniotomy was noted. Variably pigmented nodules of metastatic tumor were found widely scattered in the endocardium, epicardium, lungs, liver, adrenal glands, mediastinal, retroperitoneal, and cervical lymph nodes, and in the brain and meninges. The other notable findings were limited to the cardiovascular system.

The heart weighed 450 Gm. and was not dilated. It showed minimal hypertrophy of the left ventricle. The aortic valve ring measured 7.5 cm. in circumference. The cusps were not separated and had their usual thin, membranous appearance. The valve did not appear to be anatomically stenotic or incompetent. The ascending portion and arch of the aorta were involved in a fusiform dilatation (fig. 1). The adventitial surface of the aorta was smooth and not involved in adhesions or fibrosis. The caliber of the aorta increased rapidly above the valve, reaching its maximum of 8.5 cm. diameter about 4 cm. above it. The ascending aorta and arch had a double lumen beginning 8 cm. above the aortic valve and ending 1 cm. distal to the origin of the left subclavian artery (fig. 2). The proximal intimal tear, which measured 3.5 cm. in length, lay at about 45 degrees to the axis of the aorta. The 2 lumens were separated by a leathery septum approximately 2 mm. in thickness, both of whose surfaces were smooth and shiny (fig. 3). The smaller lumen, which measured 6.5 cm. in circumference at its origin and 4.0 cm. at its termination, lay superior and somewhat posterior to the larger lumen and gave origin to the innominate, the left common carotid, and the left subclavian

Fig. 1. External view of the aortic arch, showing the aneurysmal dilatation of the ascending portion.

Fig. 2. Diagram showing extent and relations of dissection, which is larger than, and lies inferior to, the original lumen.

Fig. 3. View of the aortic arch opened via the secondary channel, showing the intact original channel (arrows), which gives origin to the innominate, left common carotid, and subclavian arteries.
HEALED DISSECTING ANEURYSM

Fig. 4. Photomicrograph of media in area near dissection showing zone of muscular necrosis and adjacent early cystic change. (Hematoxylin and eosin, × 100.)

arteries. The septum underlying the origin of these arteries was intact. The intima of the aorta in the dilated portion showed slight longitudinal wrinkling and only an occasional slightly elevated plaque, as did the thoracic and abdominal aorta. There was no calcification, ulceration, or thrombosis. There were no congenital anomalies of the heart, and the remainder of the autopsy was essentially unremarkable.

Microscopic examination of the tumor masses showed an anaplastic tissue with dense focal pigmentation typical of malignant melanoma.

Microscopic sections from various portions of the aorta showed conspicuous changes in the media. Several sections showed long zones of muscular "necrosis" where the muscle fibers retained their outlines but had lost their nuclei and stained more deeply with eosin than the surrounding tissue (fig. 4). These necrotic bands varied in width up to one third of the thickness of the media and extended longitudinally for several centimeters. There were numerous, scattered, sharply delineated areas of myxomatous change in which no muscular or fibrous tissue was seen, but which consisted of fusiform or stellate cells irregularly arranged in a mucoid ground substance. These areas bore no notable relation to the zones of muscular degeneration and abruptly interrupted the regular architecture of the surrounding media, which showed no evidence of cellular infiltration or vascular proliferation. Some of these lesions had progressed to the point of cyst formation (fig. 5) and contained completely acellular mucoid material. An occasional cystic lesion showed evidence of organization with fibroblastic invasion, but this change was infrequent.

Sections stained with Verhoff's elastic stain showed large areas in which the elastic fibers were markedly thickened and fragmented and had lost their usual orderly arrangement (fig. 6). The areas of disruption of elastic tissue were always closely associated with the cystic lesions. The cystic lesions themselves were invariably completely devoid of elastic fibers. Sections from septum between the true and false channels showed it to be composed of a lamina of medial tissue of about half the normal thickness, covered on one side by an essentially
normal intima and on the other by much thinner condensation of fusiform cells, which in turn was lined by endothelium. The medial tissue of the septum showed relatively little necrosis and cyst formation. Similarly, sections from the wall of the dissection showed it to be composed of a thinner-than-normal media lined by a very thin intima-like tissue.

The various sections showed focal intimal changes suggesting only an early stage of atherosclerosis. The intima was focally thickened and contained small accumulations of foam cells and showed slight fibroblastic proliferation.

**DISCUSSION**

Cases of dissecting aneurysm have been recognized since the sixteenth century. Early descriptions of the disease were given by Vesalius and by Nichols. Rokitansky, in 1852, gave an accurate general description of the anatomic changes and a perceptive discussion of the pathogenesis. His views were remarkably modern, and he attributed dissecting aneurysm to a primary disease of the media, rather than to an intimal defect, expressing an attitude that is only recently becoming generally accepted. Rokitansky's conclusions are all the more astute for being based almost entirely on gross observations.

Most early reports of dissecting aneurysm dealt with patients of relatively advanced age; many authors, despite Rokitansky's early views to the contrary, regarded the condition as a disease of old age and associated it with senescence, arteriosclerosis, and hypertension. As recently as 1953, Jackson and Slavin suggest that a combination of arteriosclerosis and hypertension is the underlying cause in the majority of cases.

An early description of medial changes in dissecting aneurysm was given by Babes and Mironescu in 1910, but received little attention in the literature for several years. In 1928, Gsell described muscular necrosis in spontaneous rupture of the aorta. In 1929 and 1930, Erdheim's papers appeared, defining idiopathic cystic medial necrosis. His cases were characterized by changes limited to the media, including (1) focal necrosis with dropping out of nuclei, (2) mucoid change in the ground substance and cyst formation, (3) focal destruction of elastic fibers, and (4) repair by fibrosis without notable vascular proliferation. He emphasized the differences between the changes he observed and those of syphilitic aortitis. Soon a number of cases of dissecting aneurysm were reported with these changes. Shenan, in 1934, published a monumental review of dissecting aneurysm, analyzing 300 cases, 17 of them his own. Although he was apparently not aware of Erdheim's work, he emphasized the importance of medial changes, and concluded that "A factor common to all cases is degeneration of the elements of the media."

The descriptions of the microscopic changes in a number of his cases and several of his photomicrographs strongly suggest cystic medial necrosis. Shenan characterized 79 of his 300 collected cases as being "old" or "healed," but his criteria for this designation are not defined; many of the patients died of progressive dissection, and nearly all of cardiovascular disease. Gore in 1952 published an extensive report of the pathologic findings in 72 cases, all of which showed underlying changes in the media. Gore described 2 forms of medial degeneration: the elastic type, similar to that described by Erdheim, occurring chiefly in younger age groups, and the muscular type, occurring at older ages. He suggested that the primary change in dissecting aneurysm is intramedial dissection of hemorrhage from the vasa vasorum, which only secondarily ruptures through the intima into the lumen.

Later, Gore pointed out that 32 of his 72 cases occurred in patients less than 40 years of age, and emphasized the frequency of the condition in youth. A number of cases in children and adolescents have been reported. Many of them were associated with the skeletal changes of Marfan's syndrome.

In 1896, Marfan described a syndrome of musculoskeletal anomalies, including pigeon breast, abnormally long and slender extremities, and changes in the skull and palate. Since the original description a number of other stigmata have been added to the syndrome, including congenital anomalies of the eye and heart, and, more recently, fusiform aneurysm of the aorta and dissecting aneurysm. Cystic medial necrosis of the elastic type has been associated with Marfan's syn-
drome, and presumably accounts for the aneurysmal changes.\textsuperscript{28, 29} It has even been suggested\textsuperscript{13} that cystic medial necrosis may be regarded as a “form fruste” of arachnodactyly or Marfan’s syndrome. It is conceivable that there is a common underlying metabolic defect, which, in a mild form, is reflected anatomically only in the cardiovascular system, while in its severe form produces widespread changes in many organ systems.

Dissecting aneurysms occasionally communicate by 2 or more rents in the intima with the original lumen of the aorta, forming the so-called “double-barreled aorta.”\textsuperscript{5, 11} In these cases the secondary channel sometimes remains patent, and the new channel forms an endothelial lining.\textsuperscript{3, 5} It is from this group that the majority of long survivals have been reported.\textsuperscript{1-7}

Although a number of cases have been reported as examples of “healed” dissecting aneurysm, many of these represent only a relatively prolonged survival in the face of a progressive disease process. Only 4 of Gore’s 85 cases were healed.\textsuperscript{20} Most of the reported cases suffered an acute clinical episode of dissection, which was often associated with pain, and then survived for months or years, usually symptomatic, and often incapacitated. A notably frequent sequel to dissecting aneurysm is congestive heart failure, symptoms of which are noted in nearly all patients who survive. Many patients die in intractable failure,\textsuperscript{4} despite the absence at autopsy of obvious cause, such as valvular involvement or myocardial disease. Peery\textsuperscript{3} suggested that the loss of normal aortic elasticity following dissection results in circulatory failure by impairing the diastolic rebound mechanism, which normally makes an important contribution to circulatory efficiency.

It has been noted\textsuperscript{20} that although severe pain is a characteristic of dissecting aneurysm, approximately 50 per cent of reported cases are entirely painless throughout their course.

The case described above shows several unusual features and a number that are at variance with the traditional view of dissecting aneurysm as a degenerative disease of old age. The patient’s only definite clinical manifesta-

tions of vascular disease were the systolic and diastolic murmurs, the peripheral signs of aortic insufficiency, and the recurrent episodes of atrial fibrillation. The attacks of syncope may have been due either to the aneurysm or to the metastatic melanoma later found in the central nervous system. Excepting possibly the episodes of syncope, the patient never had symptoms of cardiovascular disease; notably, there were no signs or symptoms of congestive heart failure, although he was ambulatory until the preterminal stages of his neoplastic disease.

The clinical findings suggest a degree of aortic insufficiency despite the paucity of anatomic changes in the valve at autopsy. It must have been mild, however, in view of the borderline left ventricular hypertrophy. Several authors have noted and commented on the presence of clinical signs of aortic insufficiency in cases of dissecting aneurysm where autopsy fails to reveal significant anatomic changes.\textsuperscript{1, 4} It might be speculated that much of the elevation in pulse pressure is due to loss of elasticity of the aorta with resultant loss of the diastolic recoil phenomenon.

The patient was relatively young and active until incapacitated by his tumor. He was not hypertensive, and at autopsy showed only minimal evidence of arteriosclerosis. On the other hand, the characteristic changes of idiopathic cystic medial necrosis were conspicuous in all sections taken from the aorta. The medial change was chiefly of the type described by Erdheim, or of the “elastic type” in Gore’s classification, although considerable muscular necrosis was also present. The present case supports the increasingly accepted view that essentially all cases of dissecting aneurysm occur in aortas affected by cystic medial necrosis, and that dissection bears no relation to intimal disease and arteriosclerosis.

The dissection in the present case was completely healed, and the false channel was endothelialized. The dissection was associated with a fusiform dilatation of the involved portion of the aorta, a finding that is not uncommon. It is notable that the false channel was dilated, while the original aortic lumen retained its normal dimensions and was about three-fourths surrounded by the false lumen.
Summary

A case of healed dissecting aneurysm of the aorta is reported. The aneurysm produced few clinical manifestations and the patient died of unrelated causes.

The literature on dissecting aneurysm is briefly reviewed. Although the condition has long been recognized, it has only recently been related to the morphologically specific cystic medial necrosis, which is coming to be recognized as the most important underlying cause, rather than arteriosclerosis and hypertension. A number of cases of “healed” dissecting aneurysm have been reported, most of them surviving a limited period of time and succumbing to further dissection and rupture or to congestive heart failure.

Several unusual features of the present case are discussed.

Summario in Interlingua

Es reportate un caso de curate aneurysma dissecante del aorta. Le aneurysma produceva pauc manifestaciones clinica, e le patiente moriva ab altere causas.

Es presentate un breve revista del litteratura de aneurysma dissecante. Ben que le condition ha ab longo essite recognoscite, il es solo recentemente que illo essiva relateate con le morphologicamente specific necrosis cystica medial, le qual—plus tosto que arteriosclerosis e hypertension—se revela de plus in plus como le major causa primari. Le litteratura cognosce un numero de casos de “curate” aneurysma dissecante. In le majoritate de illos, le patientes superviveva un periodo limitate de tempore e succumbeva a dissection additional con ruptura o a congestive disfallimento cardiac.

Plure aspectos inusual del presente caso es discutite.

References

ORGANIZATION IN RESEARCH

"The catchword of our post-war times is organization. The individual freedom is our chief asset, the mainspring of the really new ideas, the guarantee of progress. Physiology does not go forward as an ordered line of battle on a continuous front, but must be carried on, as someone has aptly said, as a guerilla warfare against the unknown, conducted single-handed or by quite small units. There is no need for an extensive organization of research, but there is much need for voluntary cooperation on a limited scale between individuals and laboratories. There are many problems which can only be successfully attacked when experimental physiologists cooperate with histologists, with chemists or physicists or with clinicians, and some problems will require the combined efforts of several of these groups, but the affair is always one of local and voluntary cooperation and does not concern us here.

"While I have no faith in the organization on a large scale of research I think there is a wide and fruitful field for organization of what we might term the services behind the front."—A. Krogh. The Progress of Physiology, 1929.
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Circulation. 1957;15:540-546
doi: 10.1161/01.CIR.15.4.540
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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