Primary Dissecting Aneurysm of Coronary Artery

A Cause of Acute Myocardial Ischemia

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SUMMARY

Primary dissecting aneurysm of a coronary artery is a rare form of acute coronary arterial obstruction.

A review of 24 cases, including two observed by the authors, indicates that this condition tends to occur (1) in the young adult, (2) in the female more commonly than the male, and (3) relatively commonly in the postpartum state.

The clinical manifestations take the form either of sudden death or acute myocardial infarction. In the latter situation, there is the potential for overcoming the coronary arterial obstruction by a surgical bypass procedure.

Additional Indexing Words:
Acute myocardial infarction  Sudden death  Nonatheromatous coronary disease

Among the variety of conditions other than atherosclerosis which are responsible for myocardial ischemia is primary dissecting aneurysm of a coronary artery. This entity, which is localized to the coronary arterial system, leads to profound changes including sudden death or acute myocardial infarction. As the coronary arterial system, except for the localized obstructive lesion, is generally normal, this disease has the potential for early application of surgical bypass therapy.

Our experience with two cases prompts us to place these cases on record and to review the essential features of the 22 other cases which we have been able to find in the literature in English.

Report of Cases

Case 1

A 42-year-old white woman had been in good health until the day of her death. There had been no history of cardiovascular disease and she had been taking no medication. The patient experienced the acute onset of severe pain in the chest and left arm shortly after preparing breakfast at home and died before arrival of an ambulance.

At autopsy, pertinent findings were limited to the heart. The heart weighed 275 g and showed no gross evidence of myocardial or valvular disease. The coronary arteries showed only very minimal atherosclerosis. The anterior descending coronary artery, beginning 4 cm from its origin and extending to the apex of the heart, was surrounded by a hematoma. The latter measured about 7 mm in diameter. Cross sections of the artery showed the lumen narrowed to the point of occlusion by the hematoma. No aortic disease was observed.

Histologic examination revealed a dissecting aneurysm of the anterior descending coronary artery characterized by the presence of fresh blood within a newly created channel in the media. There were corresponding severe degrees of luminal narrowing (fig. 1a). Serial sections
Case 1. (a) Photomicrograph of anterior descending coronary artery showing an intramural hematoma (H.) which causes distortion and compression of the lumen (L.) of the vessel. Elastic tissue stain; reduced slightly from × 20. (b) Anterior descending coronary artery at

Circulation, Volume XLI, February 1972
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were taken of the entire involved segment. Six centimeters from the origin of the artery, there was a tear of the intima and underlying media (fig. 1b). Through this tear, the arterial lumen communicated with the intramedial hematoma (fig. 1c). The latter extended both proximally and distally from the level of the tear. In one of the histologic sections, at a level about 1 mm proximal to the tear, there was an inflammatory infiltrate consisting primarily of neutrophils within the intima and media (fig. 1d). The media showed poor staining of its elements, suggesting a necrotizing process. No other vessels in the body showed inflammatory disease and myocardial infarction was not identified histologically.

Case 2

A 32-year-old white woman developed substernal pain that extended to the left shoulder and arm 6 days after uneventful delivery of a full-term infant. An electrocardiogram revealed the possibility of right atrial hypertrophy but was otherwise unremarkable. The patient’s condition stabilized, and she was discharged 2 days later. Twelve days following discharge from the hospital, she was found dead at home.

At autopsy, the abnormal findings were limited to the heart. The heart weighed 300 g. The anterior descending coronary artery and its branches were extremely tortuous and protruded some distance above the surface of the heart. Overlying this artery, the epicardium was hemorrhagic, containing multiple ecchymoses and petechial hemorrhages. The myocardium revealed signs of a recent anterolateral myocardial infarct that also involved the anterior half of the ventricular septum. The myocardium was softened in this area and contained large areas of interstitial hemorrhage that measured up to 2 cm in diameter. The right and circumflex coronary arteries revealed focal areas of mild degrees of atherosclerosis but no ulceration or hemorrhage. Grossly, the aorta was normal.

Histologic examination of the anterior descending coronary artery revealed extensive hemorrhage within the media extending the entire length of the vessel. The lumen was distorted and narrowed by the hematoma (fig. 2a and b). This artery was not examined in serial sections but, at one of the levels studied, there was a zone in which medial tissue was replaced by fibrous tissue. This area was not related to the intramedial hematoma and was interpreted as a zone of healed necrosis of the media and intima. No inflammatory infiltrate was associated. The intramedial hematoma was in the process of being organized (fig. 2c and d).

The media of the main left coronary artery showed numerous small cystic spaces and swelling of the ground substance with fragmentation of the elastic fibrils compatible with idiopathic cystic medial necrosis (fig. 3a). The myocardium revealed numerous foci of recent infarction with signs of removal of necrotic muscle (fig. 3b). Examination of the proximal aorta revealed areas of cystic medial necrosis at the area adjacent to the left coronary ostium.

Discussion

Among dissecting aneurysms involving coronary arteries, the most common situation is that the coronary arterial lesion is secondary to aortic dissecting aneurysm. In contrast, the condition herein reported and which we have chosen to call primary coronary arterial dissecting aneurysm is restricted to the coronary arterial system. Like dissecting aneurysms in other arteries, it is characterized by the presence of a hematoma within the media. The hematoma, in turn, is responsible for luminal narrowing.

Primary dissecting aneurysm of a coronary artery yields a clinical picture of catastrophic acute coronary arterial disease.

A search of the literature in English revealed reports of 22 cases of primary dissecting aneurysm of a coronary artery. These, in addition to our two cases, allow us to review the features of 24 cases. In contrast to classical atherosclerotic disease, two features of dissecting aneurysm of a coronary artery stand out; these are age and sex. The 24 cases are

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the level of a break in the intima and media (between arrows) allowing the lumen of the artery (L.) to communicate with the intramedial hematoma (H.). Elastie tissue stain; reduced from × 175. (c) Low-power photomicrograph of anterior descending coronary artery at the level shown in b. The tear of the intima and media (between arrows) allows the lumen to communicate with the medial hematoma (H.). The lumen (L.) of the artery is narrowed and distorted by the intramedial hematoma (H.). Elastie tissue stain; reduced from × 38. (d) Anterior descending coronary artery approximately 1 mm proximal to the level shown in b and c. There is necrosis of the media (M.) associated with leukocytic infiltration of the intima and media. L. = lumen. H & E; reduced from × 170.

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Case 2. (a) Photomicrograph of anterior descending coronary artery. Within the media is a hematoma (H.) which distorts and narrows the lumen (L.). A zone of the media (between arrows) shows replacement of medial elements by young connective tissue. Elastic tissue stain; × 14. (b) The zone shown between arrows in a emphasizes the segment of the arterial wall in which the media has become replaced by fibrous tissue (between arrows). H & E; × 75. (c) Photomicrograph of anterior descending coronary artery showing intramural hematoma (H.) causing major distortion and narrowing of the lumen (L.). A zone comparable to that shown in the rectangle is illustrated in greater magnification in d. H & E; × 60. (d) A zone comparable to that shown within the rectangle in c reveals part of the intramural hematoma (H.) which is undergoing organization. M. = media. L. = lumen. H & E; × 75.
summarized in table 1 and figure 4. From these, it is apparent that the ages ranged from 21 to 62 years, only five of the patients being 45 years of age or older. Somewhat more than half of the patients (13 patients) were 40 years of age or younger.

The sex of the patients is even more contrasting, since 20 of the 24 patients were female. An additional factor of interest was the postpartum state which was observed in eight patients. In seven instances, the postpartum state was 7 weeks or less in duration (range, 2 to 6 weeks). In the eighth patient, the postpartum state was 50 days.

In each of 22 cases, only one coronary artery was involved with the following distribution: anterior descending, 17 cases; right, three cases; left circumflex, one case; and posterior descending coronary artery, one case.

In two cases, involvement was more extensive. In Boscetti and Levine's case the dissecting aneurysm, primary in the main left coronary artery, extended into the anterior descending and left circumflex branches. In only one case, that of Benson, independent dissecting aneurysms were present both in the anterior descending and right coronary arteries.

In each of the cases, the clinical picture was that of acute myocardial ischemia. In 18 cases sudden death occurred without pathologic evidence of acute myocardial infarction. In the remaining six cases, there was a clinical picture of acute myocardial infarction. Classically, among the cases of sudden death, the arterial lesion was without reaction indicating a short interval between the appearance of the arterial lesion and death. In case 2 of Brody and associates, which was an example of sudden death, the histologic evidence suggested the arterial lesion to be several days old.

Each of the six patients presenting with a clinical picture of acute myocardial infarction survived from several hours to about 2 weeks. At necropsy, five of these showed acute myocardial infarction in the distribution of the involved artery with the histologic ages of the

Figure 3

Case 2. Photomicrographs. (a) Media of left coronary artery. This layer shows clear spaces which are characteristic of cystic medial necrosis. H & E. × 70. (b) Anterior wall of the left ventricle. Beside a zone of intact myocardium (M.), is a zone of infarcted (I.) myocardium. This is related to a third zone in which only myocardial stroma (S.) is present representing removal of infarcted myocardial tissue. H & E; × 85.
Table 1

Summary of Essential Findings in 24 Cases of Primary Dissecting Aneurysm of a Coronary Artery

<table>
<thead>
<tr>
<th>Author</th>
<th>Sex</th>
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<th>Clinical manifestations</th>
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*Although the clinical picture was that of acute myocardial infarction, myocardial necrosis was not found at necropsy.

Abbreviations: RC = right coronary; AD = anterior descending; LC = left coronary; L circ = left circumflex branch; PD = posterior descending; SD = sudden death; MI = acute myocardial infarction.

infarct and the coronary arterial lesion coinciding. The sixth case, that of Wells,8 with a clinical picture of acute myocardial infarction failed to show necrosis of muscle. The patient had been admitted one evening and died during the early hours of the following morning. The arterial lesion appeared considerably older than the clinical history, as the hematoma was organized.

The etiology of primary dissecting aneurysm of a coronary artery is difficult or impossible to identify according to current data. Hypertension does not seem to be a factor, as this condition was usually absent or might be so assumed from the descriptions of the cases. Two other conditions come up for consideration, namely, cystic medial necrosis of the coronary arteries and localized coronary arteritis. Three cases, those of Boschetti and Levine3 and of Ashley,11 as well as our case 2, showed this change. In the one case of arachnodactyly (Marfan's syndrome) in this series (the case of McKeown7 involving a 21-year-old man), however, this lesion was not specifically described. The media was described as showing some degeneration with loss of muscle fibers and replacement fibrosis.

Inflammatory lesions of arteries are frequently described, usually in the form of periarterial leukocytic infiltration. Such a change may simply be a response to the intramural hematoma, rather than an underlying condition.

Acute necrotizing changes of the media of the involved coronary artery were observed in only two cases, that of Barrett15 and our case.
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Figure 4
Distribution by age and sex in 24 cases of primary dissecting aneurysm of a coronary artery.

1. Loss of medial substance was described by McKeown,7 as mentioned above, and medial degeneration was described by Benson.16 In our case 2, the scar replacing a segment of media may represent either a healed rupture from underlying cystic medial necrosis or healing of a segment of arteritis.

Atherosclerosis appears to be highly uncommon among cases of primary coronary arterial dissecting aneurysm as judged from published accounts and from our observations. An exceptional case is that of Lovitt and Corzine.2 These authors found mild atheromatous disease in a 39-year-old woman who died 2 weeks after delivery. They were of the opinion that the dissecting aneurysm began in an atherosclerotic plaque (an unusual event in dissecting aneurysm in any artery).

The question as to the source of the intramedial hematoma remains. Is it a primary medial hemorrhage or does it result from a tear of the intima and underlying media, a so-called internal tear? In only two of the 24 cases reviewed was an internal tear identified. These were the cases of Burton and Zawadzki10 and our case 1. The rarity of identification of an internal tear through which the intramedial hematoma could develop might suggest that, in many cases, the medial hematoma is a primary condition. We do not accept this as an explanation in view of the experience with our case 1. In this case, 150 serial sections were prepared of the zone with intramural hemorrhage, while an internal tear was found in only one of the sections (fig. 1b). This experience indicates that, unless meticulous care is taken, an internal tear may be overlooked as a source of primary dissecting aneurysm of a coronary artery. In our case 2, a scar was present in the media of the involved coronary artery. This lesion may have represented a zone of necrosis through which rupture could have occurred. The site of this lesion, however, does not correspond with the hematoma, suggesting that other necrotizing lesions could have been present allowing an internal tear. Since serial sections had not been done, one is left with conjecture.

From the therapeutic point of view, primary dissecting aneurysm of a coronary artery is a monumental challenge. In the majority of cases, the sudden unexpected death leaves the clinician without any potential. It will be recalled, however, that in six of the 24 cases reviewed (25%), the patient’s course was less precipitous. Yet death occurred from hours to 2 weeks after the onset of signs of acute myocardial ischemia.

The fact supports an attitude of urgency in diagnosis and treatment if a catastrophe is to be avoided. In this condition, the potential exists for emergency surgical bypass of the stenotic zone in the involved coronary artery. Application of such a procedure in this condition, in contrast to attempting it in the usual case of acute myocardial infarction, may be stimulated by the three cardinal features of primary dissecting aneurysm of a coronary artery which are (1) relative youth of patient, (2) frequent occurrence in the female, and (3) frequent presence of the postpartum state.

References

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_Circulation_. 1972;45:259-266
doi: 10.1161/01.CIR.45.2.259

_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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