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Dissecting Coronary Artery Aneurysm: A Report of Two Cases

Dissecting aneurysm of a coronary artery is an unusual occurrence, with a review of the literature revealing only 32 cases. Because this process results in sudden and unexpected death, frequently in young individuals, such cases often fall within the jurisdiction of the medical examiner. This paper reports two cases of coronary artery dissection seen at the Office of the Chief Medical Examiner, State of West Virginia, during a recent six-week period. A review of the literature concerning this unusual entity is also presented.

Case Reports

Case 1

This 37-year-old married white female, gravida V, Para 5, abortus 0, four weeks postpartum, had been in apparent good health. After eating, she complained of chest pain with radiation down the left arm accompanied by dyspnea, a choking sensation, and vomiting. She was rushed to a hospital where she was dead on arrival.

The decedent had no history of previous illness, and her pregnancies and deliveries had been uncomplicated. During her recent pregnancy she had been hospitalized on two occasions because of false labor. Her third admission was due to premature rupture of membranes with leakage of amniotic fluid for several hours. After 24 h of observation, labor was induced with vaginal delivery of a full-term baby, followed by spontaneous delivery of the intact placenta. The mother and baby were discharged in satisfactory condition four days after delivery, to return for examination in six weeks. She was started on Modicon-28® contraceptive pills (0.5 mg norethindrone and 0.035 ethinyl estradiol), which she took for 2 to 3 weeks prior to her death.

Her only complaints during hospitalization for delivery were several episodes of "heartburn" prior to delivery, interpreted as not unusual for term pregnancy.

Pertinent autopsy findings were confined to the 375-g heart. A small isolated atheromatous plaque was present in the middle third of the anterior descending branch of the left coronary artery. The right coronary artery appeared to be thrombosed from a point 3 mm distal to the ostium to the origin of the posterior descending branch. The posterior wall of the left ventricle and the posterior interventricular septum showed an extensive area of dark discoloration. Microscopically the right coronary artery showed hemorrhagic intramural dissection with compression of the lumen and marked chronic inflammatory infiltrate in the adventitia (Figs. 1, 2, and 3). Verhoeff's elastic stain confirmed that the

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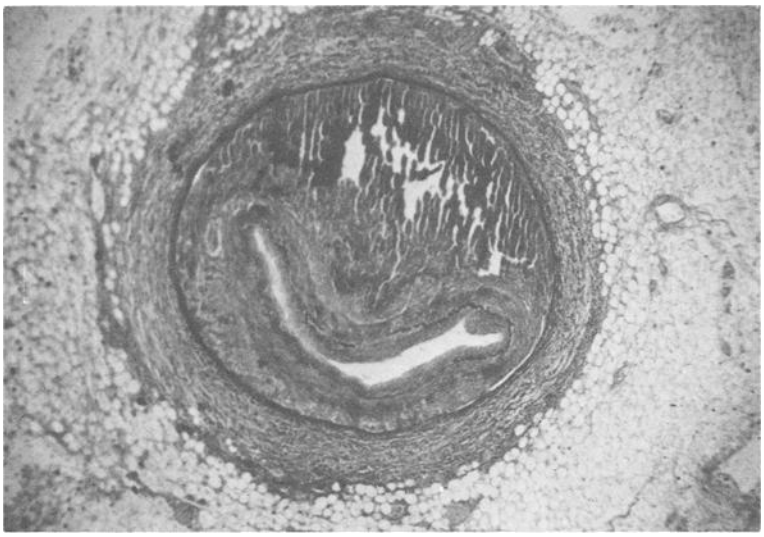

FIG. 1—*A cross section of the right coronary artery showing hemorrhagic intramural dissection with compression of the lumen (Verhoeff's elastic stain).*

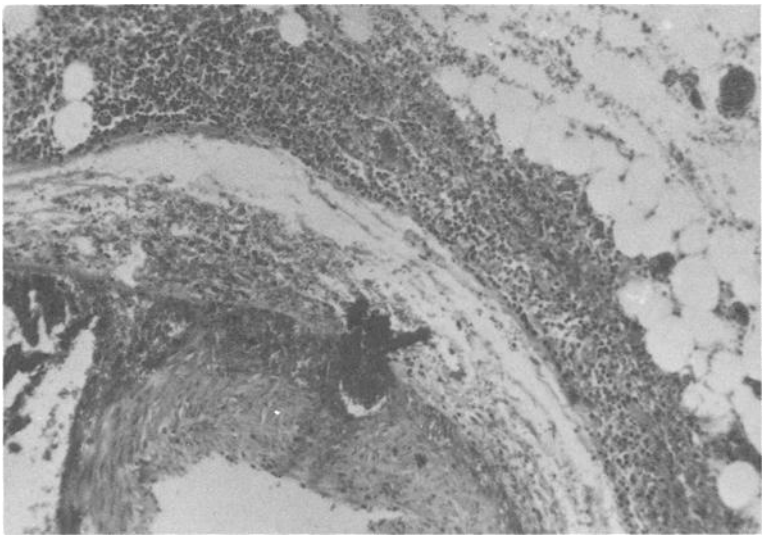

FIG. 2—*Chronic inflammatory infiltrate in the adventitia surrounding the artery (hematoxylin and eosin).*

hemorrhage was in the media adjacent to the external elastic lamina. The intima showed no atherosclerotic change and the vessel wall showed no rupture. The posterior wall of the left ventricle and the septum showed extensive replacement of the myocardium by loose fibrous tissue with areas of granulation and areas of recent necrosis.

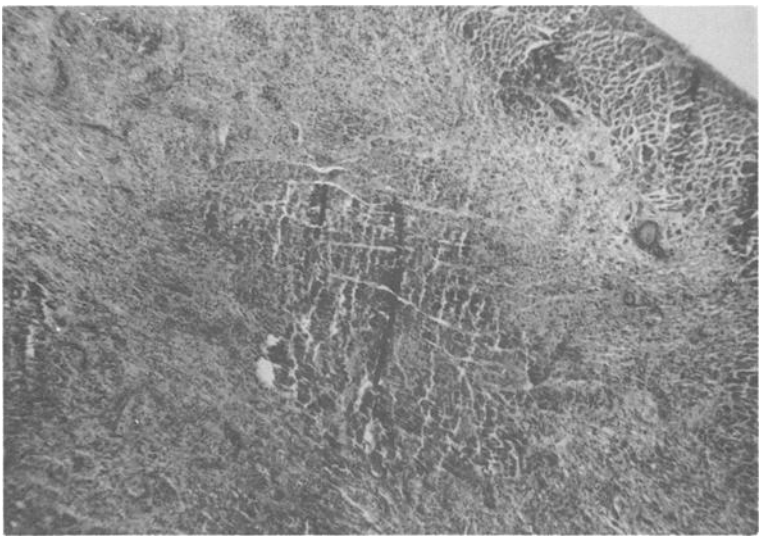

FIG. 3—*Extensive replacement of the myocardium by loose fibrous tissue with a central area of recent infarct (hematoxylin and eosin).*

Case 2

The decedent was a 49-year-old white female found dead in the bathroom of her living quarters. The decedent apparently lived alone and no retrospective medical history was obtainable except that she was hypertensive and had complained of numbness of the left upper extremity associated with nausea and vomiting on the day prior to death. This sudden and unexpected death in a 49-year-old female with a scanty past medical history and no attending physician placed the body within the medical examiner's jurisdiction.

The body was well developed and well nourished, measuring 173 cm (68 in.) in length and weighing 70 kg (154 lb). The pertinent autopsy findings were limited to the 340-g heart. The heart was right coronary dominant; the left coronary artery was slightly smaller than average diameter but, however, not hypoplastic. The coronary arteries showed no arteriosclerosis. The midportion of one of two equal-sized paired anterior descending arteries revealed a dissecting aneurysm 0.75 cm in length (Fig. 4). The position of the lesion was overlying the interventricular septum midway between the apex and base of the heart. The myocardium showed no fibrosis or recent ischemia.

Microscopically, focal areas of the left coronary artery adjacent to the dissection and the ascending aortic arch revealed minimal patchy areas suggestive of cystic medial degeneration (Fig. 5). The dissection of the coronary artery was recent and the surrounding pericoronary epicardial fat showed a preponderance of eosinophilic leukocytes. The left ventricle also revealed patchy, focal interstitial inflammatory cells composed of mononuclears and some polymorphonuclear and eosinophilic leukocytes (Fig. 6). Recent or old ischemic changes were not noted. The myocardial inflammatory change was considered as representative of focal interstitial myocarditis and not resulting from ischemia.

Discussion

Table 1 presents background data, retrieved from literature search, concerning 32 cases of coronary artery dissection. Twenty-three of those recorded were female. The average

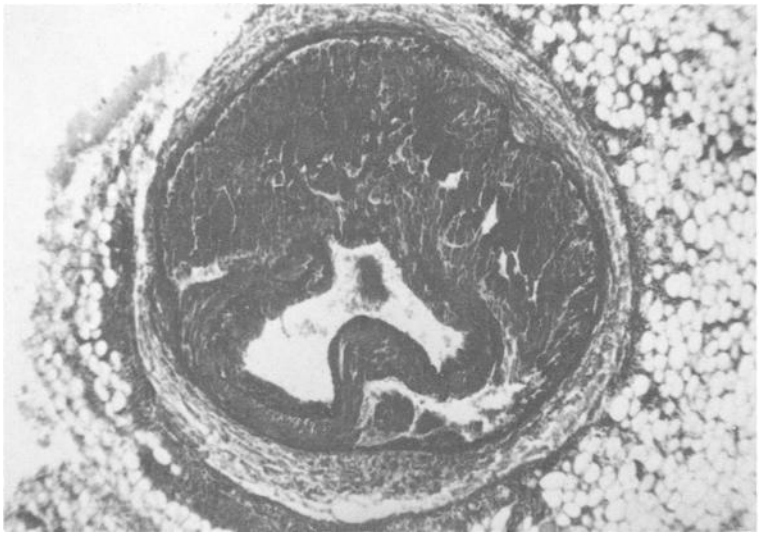

FIG. 4—*The left anterior descending coronary artery showing dissecting hemorrhage of the media (Verhoeff's elastic stain).*

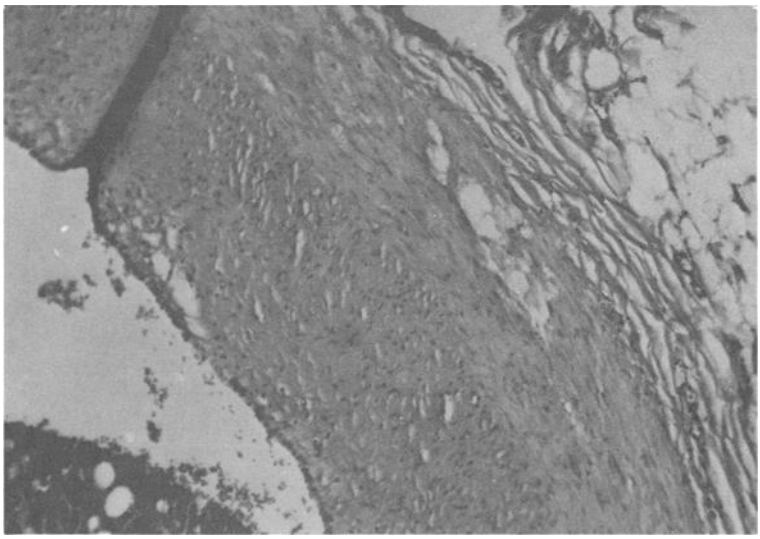

FIG. 5—*A focal area suggestive of cystic medial necrosis of the left coronary artery adjacent to the area of dissection (hematoxylin and eosin).*

age of the males was 45.7 years, with a range of 21 to 62 years. The average age for females was 39.1 years, with a range of 17 to 60 years. Thirteen of the 23 females (approximately one half) were age 40 years or less, while 3 of the 9 males (one third) were below the age of 40 years. Of the 9 reported male cases, 6 occurred in Europe; among the females, only 4 of 23 cases were reported from Europe.

Among the 23 females, 9 were postpartum, ranging from 14 to 80 days after parturition. In addition, one female was found to have cystic medial necrosis [7], while another

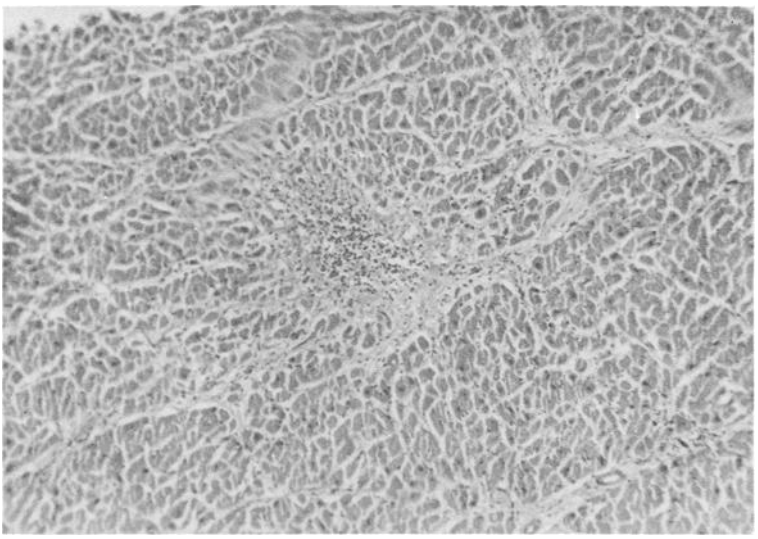

FIG. 6—*Focal interstitial myocarditis of the left ventricle (hematoxylin and eosin).*

had changes suggestive of cystic medial necrosis [*17*]. Among the males, one individual had a history of trauma to the chest antemortem [*4*], while two were subjected to severe physical exertion shortly prior to death [*3*]. One male had arachnodactyly, possibly representing a "forme fruste" of Marfan's syndrome [*11*]. One male, the only reported survivor, had severe arteriosclerotic cardiovascular disease [*25*]. Three individuals were known to have pulmonary disease [*5,8,18*]. Among the remaining males and females, no causative factors or relationships were suggested either from history or autopsy findings.

Among 26 cases for which such information was available, 11 were found to have histologic evidence of myocardial infarction. In 26 of the 32 cases, the dissection involved the left coronary artery; in 5 cases the right coronary was afflicted; and in 1 case, both the right and the left coronary arteries were involved. Quite frequently, death was sudden and unexpected, with the one reported survivor presenting with hemopericardium treated surgically [*25*].

The present cases include a postpartum female (four weeks) who showed histologic evidence of a myocardial infarction consistent with occurrence during the time of parturition. Additionally, areas of more recent myocardial necrosis were seen. The adventitia of the right coronary artery histologically showed chronic inflammatory infiltrate in addition to the recent hemorrhage. These findings strongly suggest an episode of dissection with resultant ischemia several weeks prior to death.

The second case showed recent dissection of the midportion of the anterior descending branch of the left coronary artery without evidence of infarction. In other sections of the same vessel, small areas of cystic change were noted in the media, suggestive of cystic medionecrosis. Sections of the aorta showed similar patchy foci suggestive of cystic medionecrosis.

As noted above, one literature case [*11*] was considered as possibly representing Marfan's syndrome. The pathologist should be alerted to the possibility of dissecting coronary aneurysm in such cases. The typical body habitus of Marfan's syndrome consists of tallness because of excessive length of long bones and spider-like fingers (arachnodactyly). An excessive growth of ribs may result in pectus carinatum or pectus excavatum; redundant ligaments, tendons, and joint capsules result in loose-jointedness. There are

TABLE 1—*Background data retrieved from literature search.*

Author	Age	Sex	Artery Involved	Postpartum State	Evidence of Myocardial Infarction
Pretty, 1931 [*1*]	42	f	r	...	...
Homma and Yenikomschian, 1933 [*2*]	17	f	l	...	?
Uehlinger, 1947 [*3*]	45	m	l	...	+
Uehlinger, 1947 [*3*]	46	m	l	...	...
Hedinger, 1948 [*4*]	53	m	l	...	+
Schmid, 1951 [*5*]	47	m	l	...	...
Lovitt and Corzine, 1952 [*6*]	39	f	l	14 days	...
Boschetti and Levine, 1958 [*7*]	45	f	l	...	+
Schornagel, 1958 [*8*]	39	m	l	...	...
Ahronheim and Wagman, 1959 [*9*]	41	f	r	...	...
Iglauer et al, 1959 [*10*]	38	f	r	...	+
McKeown, 1960 [*11*]	21	m	r	...	+
Wells, 1960 [*12*]	42	f	l	6 weeks	...
Leithoff, 1961 [*13*]	40	f	l	...	+
Edwards, 1961 [*14*]	40	m	l	...	?
Burton and Zawadzki, 1962 [*15*]	35	f	l	6 weeks	...
Nalbandian and Chason, 1965 [*16*]	38	f	l	...	...
Nalbandian and Chason, 1965 [*16*]	60	f	l	...	...
Ashley, 1965 [*17*]	34	f	l	...	...
Brody et al, 1965 [*18*]	40	f	l	2.5 weeks	+
Brody et al, 1965 [*18*]	36	f	l	19 days	...
Brody et al, 1965 [*18*]	41	f	l	80 days	...
Brody et al, 1965 [*18*]	49	f	l	...	+
Kurrein, 1965 [*19*]	39	f	l	...	?
Kurrein, 1965 [*19*]	51	f	l	...	?
Palomine, 1969 [*20*]	31	f	l	16 days	?
Barrett, 1969 [*21*]	42	f	l	...	?
Benson, 1970 [*22*]	62	m	r + l	...	+
DiMaio and DiMaio, 1971 [*23*]	27	f	l	2 weeks	...
Claudn et al, 1972 [*24*]	42	f	l	...	...
Claudn et al, 1972 [*24*]	32	f	l	20 days	+
Forker et al, 1973 [*25*]	56	m	r	...	+
Present Case 1	37	f	r	4 weeks	+
Present Case 2	49	f	l	...	...

abnormalities of the eye, especially ectopia lentis. Neither of our cases showed evidence of Marfan's syndrome.

The only condition frequently associated with dissection of the coronary artery in cases reported within the United States has been the postpartum state. The significance of the association remains obscure. It has been suggested that loosening of ground substance of connective tissue during pregnancy may contribute to dissection. This change in ground substance may also account for the association of pregnancy and dissecting aortic aneurysm reported by Schnitker and Bayer [*26*].

Summary

Two cases of sudden, unexpected death resulting from coronary artery dissection have been reported. Since sudden and unexpected death falls within medical examiners' jurisdiction, the systematic autopsy examination of such cases offers an opportunity to evaluate this entity as well as other rare causes of natural death. A review of the literature concerning the subject has been presented.

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