CLINICOPATHOLOGIC CORRELATIONS

Dissecting Aneurysm of Aorta Complicating Aortic Valvular Stenosis

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SUMMARY Six cases of classical dissecting aneurysm of the aorta complicating aortic valvular disease in which aortic stenosis was dominant are described. In three of the cases the aortic complication occurred after the natural aortic valve had been replaced by a prosthetic valve. The longest postoperative period was four months. The factor common to all reported cases in our series was cystic medial necrosis of the aorta.

The authors agree with the concept of McKusick and associates that aortic dissecting aneurysm complicating aortic valvular disease is an association other than chance and is related to cystic medial necrosis of the aorta. The latter, in turn, is a consequence of the altered hemodynamics of aortic valvular disease, particularly stenosis.

Case 1

A 64-year-old man had been observed for two years with a clinical diagnosis of aortic stenosis and congestive cardiac failure. The blood pressure was 160/50. At the final admission, the patient was admitted with "a strange feeling" in the throat. Profound shock developed, followed by death.

The autopsy revealed a bicuspid aortic valve with a moderate degree of calcific aortic stenosis and a classical dissecting aneurysm of the aorta. The latter had ruptured into the pericardial sac (fig. 1). The heart was enlarged, the heart and ascending aorta weighing 750 grams.

Case 2

A hypertensive, 52-year-old man with a history of a cardiac murmur since young adulthood was admitted to the hospital because of sudden onset of severe substernal pain which radiated to the neck and epigastrium. The blood pressure was 170/100. A grade IV/VI systolic ejection murmur was heard over the aortic area and radiated to the carotid arteries. A 1/VI early diastolic blowing murmur was heard over the apex. The electrocardiogram showed signs of left ventricular hypertrophy.

About six hours after admission, there was accentuation of the precordial pain. This was shortly followed by hypotension, and loss of the left carotid, brachial and right femoral pulses. Cyanosis of the face and neck appeared and the central venous pressure was found to be elevated at 28 cm/H2O. A roentgenogram taken during this episode showed a markedly widened mediastinum. Death occurred about two hours after the onset of this episode in spite of attempts at resuscitation.

The essential autopsy findings were those of classical dissecting aneurysm originating in the ascending aorta (fig. 2), extending the full length of the aorta and terminating in the right common iliac artery. The ascending aorta ruptured, leading to massive hemopericardium. The aortic valve was congenitally bicuspid and stenotic on the basis of heavy calcification. The heart weighed 670 grams.
Case 3

The patient was a boy who, at the age of three months, had been found to have severe coarctation of the aorta (flush blood pressures: right brachial, 160 mm Hg; right leg, 50). A grade III/VI systolic murmur of equal intensity was heard over the left sternal border and back. No diastolic murmur was present. The coarctation of the aorta was resected at the age of ten months. Hypertension without demonstrable cause persisted postoperatively (brachial, 168/105; femoral, 148/115).

At the age of eight years, there was moderate widening of the ascending aorta and minor narrowing of the aorta at the level of resection of the coarctation (fig. 3a). One year later there was noticeable increase in caliber of the aorta over that seen earlier (fig. 3b and c).

When examined at the age of 11 years, there were electrocardiographic signs of left ventricular hypertrophy. A grade II/VI systolic ejection murmur was heard best along the right upper sternal border and a grade III/VI diastolic blowing murmur was heard along the left sternal border radiating to the apex. Two months following the last examination and during a flu-like illness, the boy collapsed and died.

The autopsy showed an adequate channel at the site of previous resection of the coarctation. The aortic valve showed a unicuspid type of congenital stenosis. The aorta was the site of a classical dissecting aneurysm beginning in the ascending aorta (fig. 4). Rupture into the pericardial sac was the immediate cause of death. The heart weighed 450 grams.

Cases with Prosthetic Aortic Valve

Three of the cases (cases 4-6) with dissecting aneurysm of the aorta complicating aortic valvular stenosis developed the aortic complication after the natural aortic valve had been removed and replaced by a prosthesis. In one, the dissecting aneurysm developed during the operation. Of the other two, aortic dissecting aneurysm developed 22 days after the operation in one and four months postoperatively in the other.

Case 4

A 66-year-old woman presented with clinical signs of aortic stenosis. A grade III/VI systolic ejection murmur was heard along the left sternal border, radiating over the carotid arteries. No diastolic murmur was heard. The blood pressure was 130/100 mm Hg.

At operation, a heavily calcified tricuspid stenotic aortic valve was replaced with a Lillehei-Kaster prosthetic valve.

Figure 1
Case 1. a. Congenital bicuspid aortic valve viewed from above. The posterior (P.) of the two cusps is heavily calcified and was rigid. b. Ascending aorta. Laceration of dissecting aneurysm.

Figure 2
Case 2. Ascending aorta and aortic arch. There is a circumferential laceration of the ascending aorta associated with an intramural hematoma. The aortic valve, which was stenotic, is mostly out of view.
Various resuscitative measures were necessary to obtain a spontaneous cardiac action. After this was accomplished, the line of the aortotomy showed dehiscence. Attempts to repair the aortic wall were unsuccessful and the patient died on the operating table. The autopsy showed classical dissecting aneurysm limited to the ascending aorta. The heart weighed 495 grams.

Case 5

A 57-year-old man presented with signs of aortic stenosis. A grade IV/VI systolic ejection murmur was heard best at the aortic area radiating over the carotid arteries. There was a grade III/VI diastolic decrescendo murmur along the left sternal border radiating to the apex. Invasive studies showed a systolic gradient of 74 mm Hg between the left ventricle and aorta and grade 3 aortic regurgitation. The blood pressure was 114/84.

The aortic valve was replaced with a Lillehei-Kaster prosthetic valve. Recovery from the operation was uneventful. Twenty-two days following operation, sudden onset of abdominal pain was experienced.

A grade III/VI systolic ejection-type murmur was heard over the aortic area. The brachial blood pressure was 220/72. An abdominal bruit was present. Neither femoral pulse could be palpated and both lower extremities were cool below the knee. Compared with earlier roentgenograms,
FIGURE 5 Case 5. Thoracic roentgenograms in frontal view. a. One week following replacement of the aortic valve. The mediastinum is within normal limits. b. Twenty-five days after replacement of the aortic valve. Mediastinal widening is consistent with widening of ascending aorta.

FIGURE 6 Case 5. a. Above the old aortotomy site is a laceration of the ascending aorta which led into a classical dissecting aneurysm. The aortic valvular prosthesis, which had been inserted 22 days earlier, has been removed prior to preparation of the photograph. b. Descending aorta. The wall of the false passage (F.P.) has been opened and retracted. The true lumen is designated by the letter A. c. Photomicrograph of the aorta showing a characteristic intramural hematoma lying, at this level, between the media (M.) and the adventitia (Ad.). (Elastic tissue stain; ×8.3.)
there was widening of the upper mediastinum consistent with widening of the ascending aorta (fig. 5). Aortography showed signs of a dissecting aneurysm originating in the ascending aorta and extending to the left common iliac artery. The patient was treated with antihypertensive drugs and a stable state was achieved, the brachial blood pressure being maintained at a level of about 110/70.

Three days later, signs of intestinal obstruction developed for which the abdomen was explored surgically. This revealed absence of pulsation in the superior mesenteric artery and evidence of necrosis of the intestine involving the ileum and the colon to its midtransverse portion. These segments were resected. While the immediate postoperative state was stable, sudden death occurred on the day following the abdominal operation.

The autopsy showed a hemopericardium resulting from intrapericardial rupture of a classical dissecting aneurysm of the aorta. The internal tear was in the ascending aorta about 1 cm distal to the healed aortotomy incision (fig. 6). The dissecting aneurysm involved the entire length of the aorta. Its most distal position was the left internal iliac artery. The heart weighed 600 grams.

Case 6

In a 55-year-old man with a heavily calcified stenotic and regurgitant aortic valve, the preoperative blood pressure was 140/80. At the time of operation, the ascending aorta was grossly dilated. After replacement of the aortic valve with a Starr-Edwards prosthesis, closure of the aortotomy incision presented difficulty because of friability of tissue.

Reoperation five hours postoperatively was necessitated by hemorrhage which was found to be originating from a tiny defect in the right atrial appendage and oozing at the aortotomy site. The latter was controlled by packing with oxycel gauze. The postoperative course was then uneventful.

Four months and 13 days after the operation, substernal pain radiating to the back developed suddenly. Bilateral brachial blood pressures were 160/100. Neither femoral pulse was palpable and a roentgenogram showed mediastinal widening (fig. 7).

Operation was then performed. A laceration was found in the interior of the ascending aorta lying about 1.0 cm above the ostium of the right coronary artery and associated with an intramural hematoma of the aorta. The valvular prosthesis was not remarkable.

The segment of the ascending aorta containing the laceration was excised and replaced with a dacron graft. In this process, the passage containing the intramural hematoma was oversewn. At the completion of the procedure, the patient could not develop adequate cardiac function and died on the operating table.

The essential autopsy findings were those of classical dissecting aneurysm of the aorta (fig. 8) with the intramural dissection extending into each common iliac artery. The heart weighed 500 grams.

Cystic Medial Necrosis

In each case sections were taken from various segments of the aorta with particular interest in the question of cystic medial necrosis. In most cases, tissue was available from the following zones: the right and left sides of the ascending portion, the arch and the descending aorta.

Each case showed some degree of cystic medial necrosis. This process tended, with minor exceptions, to be essentially

![Figure 7 Case 6. Roentgenogram of thorax four months and 13 days following replacement of the aortic valve. There is prominent widening of the mediastinum.](http://circ.ahajournals.org/)

![Figure 8 Case 6. Descending thoracic aorta showing the interior of the false passage characteristic of dissecting aneurysm. Fragments of the lacerated media are evident.](http://circ.ahajournals.org/)
the same among the various zones sampled. Using the criteria of Carlson and associates, we found grades 1 and/or 2 in four cases (cases 1, 2, 4 and 5; fig. 9a and b), grade 3 in one case (case 3; fig. 9c) and grade 4 in one (case 6; fig. 9d).

Comment

In 1957, McKusick, Logue and Bahnson called attention to the phenomenon of dissecting aneurysm of the aorta occurring in subjects with aortic stenosis (dominant) and insufficiency. In their report of four cases, they referred to earlier reports of similar cases. These authors suggested that the association was more than coincidence. They found cystic medial necrosis of the aorta in each of their cases and postulated that this change was secondary to the hemodynamics of aortic valvular disease. In our six cases, while aortic insufficiency was commonly present to some degree, the aortic stenosis was dominant and cystic medial necrosis of the aorta occurred in each case.

That the aorta is subject to laceration and secondary dissecting aneurysm in sites beyond obstructing lesions is not peculiar to aortic valvular stenosis. Cases of the aortic complication have occurred distal to supravalvular stenosis and distal to coarctation of the aorta.

It is interesting that in three of our six cases dissecting aneurysm of the aorta occurred after replacement of the aortic valve, one at the time of operation, one 22 days later and the third four months postoperatively.

Others have reported the phenomenon of aortic dissecting aneurysm occurring some time after replacement of the aor-

![Figure 9](http://circ.ahajournals.org/)

**Figure 9** Photomicrographs of aortas showing varying degrees of cystic medial necrosis. a. Case 5. Grade 1 change. (H & E; ×73.) b. Case 2. A pool of mucinous material separates elements of the lumen. The change was classified as grade 2. (H & E; ×90.) c. Case 3. The entire thickness of the aortic wall, including the hematoma (H.) within it, is shown. The media (above) shows numerous elements of disruption in continuity on the basis of grade 3 cystic medial necrosis. (Elastic tissue stain; ×14.) d. Case 6. Cystic medial necrosis, grade 4. There is disruption in continuity of the media with retraction of the edges. (Elastic tissue stain; ×40.)
tic valve. To our knowledge, the longest reported postoperative period was six years, as observed by Derkac and associates. The phenomenon common to our cases and in those reported was cystic medial necrosis of the aorta. In each of our cases this process was somewhat more advanced than is classically seen in cases of aortic dissecting aneurysm complicating hypertension. On the other hand, the process in general was less extensive than in arachnodactyly.

Hypertension, while present in some of our cases, was not uniformly present and none of the patients showed the physical stigma of arachnodactyly. These points support the concept of McKusick and associates that the hemodynamics of aortic valvular disease underlie the development of aortic cystic medial necrosis.

We view the internal tear seen in classical dissecting aneurysm as the primary process which, in turn, is followed by intramural dissection of blood. If the internal tear is not followed by an intramural extension of blood, either external rupture of the aorta may occur, as in the case of Flanders and associates, or a simple saccular aneurysm may develop at the level of the internal tear. Such a process was described in a case of aortic stenosis by Heath and associates and another by Baker and Kidd.

The occurrence of dissecting aneurysm at a time remote from the replacement of the aortic valve supports the concept that cystic medial necrosis is the fundamental basis for aortic dissecting aneurysm complicating aortic stenosis.

References

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