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Dissecting Aneurysm of Aorta Complicating Coarctation of the Aorta

Case report and brief literature review

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The incidence of dissecting thoracic aorta aneurysm complicating coarctation of the aorta ranges from 1.7 to 23% in large series of coarctation of the aorta reported. In a review of 95 cases of dissection of the aorta, we found only one case with associated coarctation of the aorta. The presence of this anomaly, although rare, should be suspected and searched for in younger patients with dissection, in view of the surgical implications at the time of correction.

Rupture of the aorta, which in most cases is associated with aortic dissection, has been observed in 19 to 23% of patients with coarctation of the aorta in two large autopsy series (1,2). In a recent report of 234 patients with coarctation, the incidence of dissection was 1.7% (3). When we reviewed 95 cases of dissecting aneurysm of the aorta at Henry Ford Hospital from 1964 to 1978, we found only one case associated with coarctation of the aorta in a 16-year-old boy. Although rare, this anomaly should be considered and searched for in patients under 40 both before and at the time of emergency repair of a dissecting thoracic aorta aneurysm.

Case Report

A 16-year-old Caucasian boy was admitted to a local community hospital with sudden onset of throat and anterior neck pain associated with profuse perspiration and transient syncope. He had no known history of any previous illness except for a recent upper respiratory infection. A physical examination five years earlier was reported as normal. The same evening, he became hypotensive and was found to have a paradoxical pulse. A pericardiocentesis was performed with aspiration of 130 ml of blood with slight increase of the systolic pressure to 94 mm Hg. A second pericardiocentesis six hours later yielded only 50 ml of blood, and the boy was then transferred to Henry Ford Hospital with the tentative diagnosis of hemorrhagic pericarditis. When he was brought in, he was unconscious, cyanotic, without blood pressure or pulse, oliguric, although the pupils still reacted to light. Cardiopulmonary resuscitative maneuvers were started, followed by a left anterior thoracotomy with evacuation of blood from the pericardial sac and manual cardiac massage. When it became evident that the patient had experienced an acute dissection of the ascending aorta, he was taken immediately to the operating room where a median sternotomy was performed. Under cardiopulmonary bypass a 35 mm dacron graft was sutured intraluminally in the ascending aorta after the aortic cusps were suspended. Thirty minutes after surgery, his left ventricle gradually became distended, effective cardiac activity could not be maintained with the usual supportive measures, and the patient died in the operating room.

At autopsy, a dissecting aneurysm of the ascending aorta was found proximal to the origin of the great neck vessels. It was six cm long and was covered by an intact intraluminal dacron graft. A coarctation of the aorta was found immediately distal to the ligamentum arteriosum (Fig. 1). The lumen was about one third the diameter of the aortic lumen proximal and distal to it. The aorta distal to the coarctation, especially the abdominal segment, appeared hypoplastic. There was no blood in the pericardial cavity. All the cardiac chambers were dilated along with generalized myocardial hypertrophy; the heart weighed 700 gm. There was fibroelastosis of the endocardium of the left ventricle. The cardiac valves and coronary arteries were normal.

Microscopic examination of the ascending aorta confirmed the dissection, which occurred predominantly in the middle and outer one third of the wall. Extensive, diffuse patchy loss of elastic fibers in the wall of the ascending aorta was present at all levels but most marked in the outer one half of the wall (Fig. 2). In some areas of elastic fiber loss, there were accumulations of alcian blue positive ground substance. This medial mucoid degeneration of

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the aorta was minimal in the abdominal aorta but especially prominent in the ascending aorta.

Discussion

From an extensive analysis of the literature of coarctation of the aorta, Abbott (1) found that 38 of 200 patients (19%) died of rupture of the aorta. In this group, 22 of 33 cases involving the ascending aorta were associated with aortic dissection. Reifenstein et al (2) found rupture of the aorta in 24 of 104 (23%) reported cases of adult coarctation of the aorta. In 19 the rupture involved the ascending aorta with the usual symptoms of dissecting aneurysm, while in the other five patients the rupture occurred distal to the coarctation. Loss of elastic tissue in the media was the predominant finding.

Hirst et al (4) compiled clinical and pathologic findings in 505 cases of dissecting aneurysm in English literature from 1933 to 1954 and found 11 cases (2%) with some degree of coarctation. Seven cases occurring in individuals below the age of 40 comprised 9% of the 74 dissections encountered in this age group. Two had associated bicuspid aortic valve and two occurred during pregnancy.

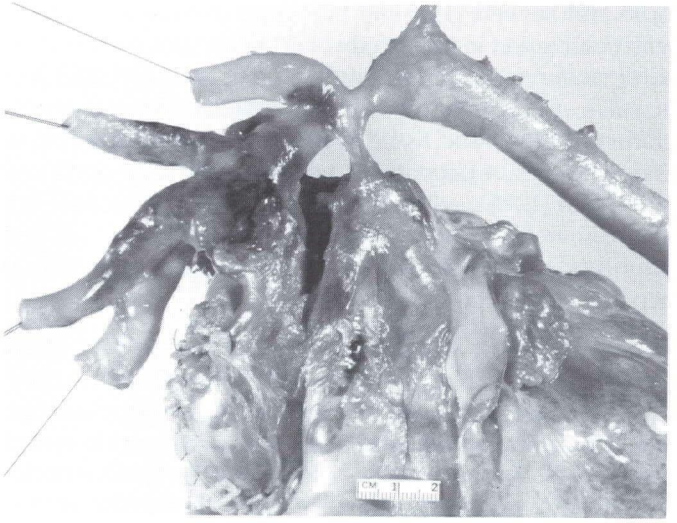

Fig. 1
Gross specimen showing the coarctation of the aorta immediately distal to the ligamentum arteriosum. Some evidence of recent surgery can be noted.

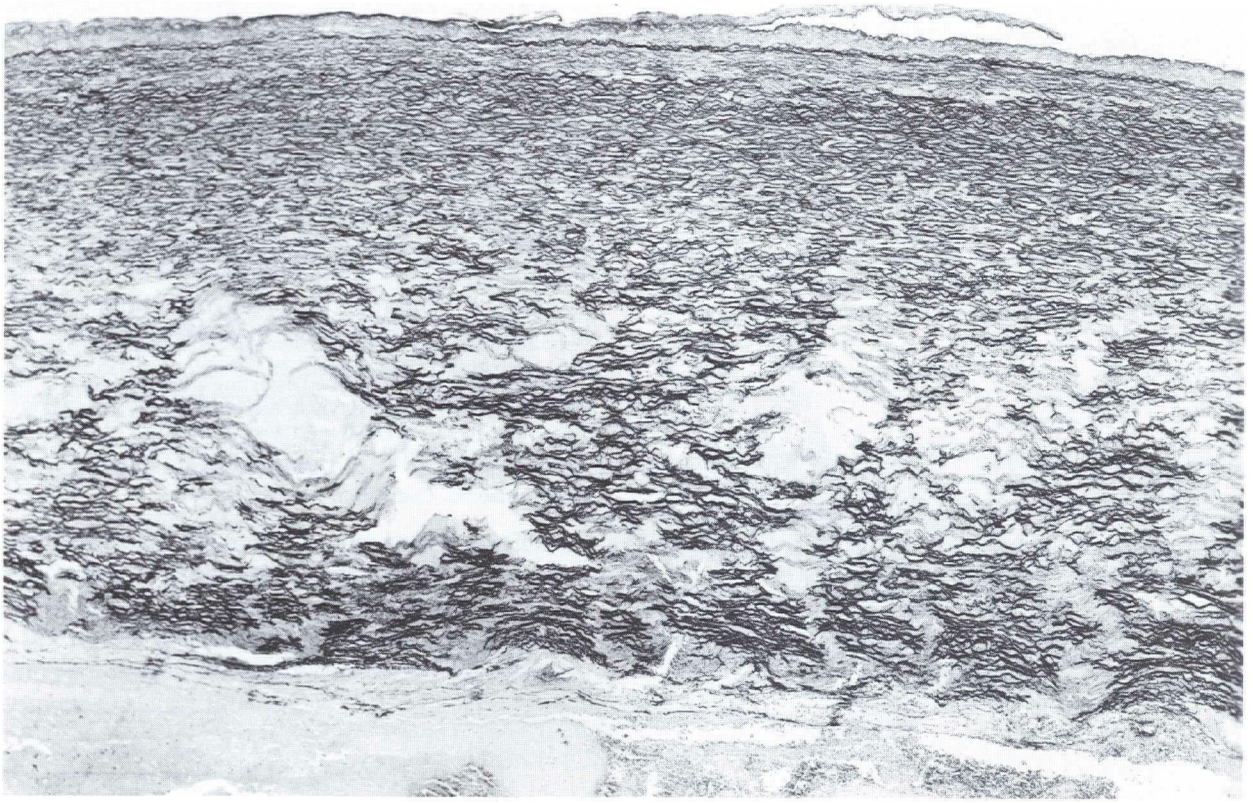

Fig. 2
Microscopic section of wall of the ascending aorta demonstrating fragmentation of the elastic fibers with mucoid cystic spaces. Elastic stain.

Gore and Seiwert (5) found five cases of dissection linked with coarctation in their study of 85 fatal cases of dissecting aortic aneurysm. One patient had associated bicuspid aortic valve, and a second had arachnodactyly. Medial degeneration of the elastic tissue predominated. Liberthson et al (3) reviewed 234 patients with coarctation of the aorta and found four (1.7%) with complicating aortic dissection; but they did not say how many of these patients had had previous repair of the coarctation and/or residual hypertension.

Recently, Fukuda and Edwards (6), in their report of six cases of aortic valvular stenosis with dissecting aneurysm, described an 11-year-old boy who had undergone resection of a coarctation of the aorta at the age of 10 months and at autopsy was found to have unicuspid, congenital aortic stenosis and an inadequate channel at the site of the previous coarctation. Dissecting aneurysm of the ascending aorta with rupture in the pericardial sac was the cause of death. No mention was made whether this patient exhibited residual postoperative hypertension as a possible risk factor for the dissection. Isolated examples of dissection distal to the coarctation have also been reported.

Among the hemodynamic forces that adversely affect the aortic wall media and therefore predispose to dissection are: hypertension; dilatation of the aorta following Laplace's law, by which wall tension increases proportionally with arterial radius; valvular aortic stenosis; supravalvular aortic stenosis; congenital bicuspid aortic valve; and coarctation of the aorta. As several authors have pointed out (5), the mechanical factors secondary to these conditions, superimposed upon a possible metabolic defect or connective tissue disorder of the aortic media, will help to initiate intramural hemorrhage and intimal tear. In cases with aortic valve disease, cystic medial necrosis seems to be more than a coincidence (8), and the poststenotic dilatation of the aorta may play a role, adding hemodynamic stresses to its wall.

Summary

Although uncommon, coarctation of the aorta or congenital aortic valve anomalies should be suspected and searched for in younger patients with dissecting aneurysm of the aorta in view of the surgical implications at the time of correction. When coarctation is present, at the time of cardiopulmonary bypass a second arterial cannula will need to be inserted in the axillary artery to provide adequate brain perfusion in addition to the usual femoral artery cannulation. Dacron patch repair of the coarctation could also be carried out during the same procedure. Stricter management with afterload reduction and proper monitoring of systemic pressures on the upper extremities would be mandatory in these patients.

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