Healed Dissecting Aneurysm of the Aorta Erroneously Diagnosed Paramediastinal Effusion; Death Following Attempted Aspiration

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Approximately 1 per cent of dissecting aneurysms of the aorta develop re-entry tears and heal, forming the so-called "double-barreled" aorta which is compatible with a variable period of survival. Occasionally a case is found wherein the dissection has remained limited to the media, healing without evidence of intimal tears. We describe such a case, so rare as to constitute a medical curiosity.

Two unusual features have prompted us to report the following case of dissecting aneurysm of the aorta. The x-ray shadow of the aneurysm was interpreted as due to a paramediastinal effusion in a patient with known pulmonary tuberculosis. An attempted aspiration led to perforation through the dissection into the lumen of the aorta which resulted in a fatal cardiac tamponade. At necropsy the aorta exhibited a healed medial dissection with complete absence of an intimal tear.

Case Report

W. F., a 27-year-old white man, was known to have had pulmonary tuberculosis since 1938. The pulmonary lesions had been progressive despite intermittent bed rest. A right pneumothorax was performed in 1943 and a left pneumothorax in 1946. The last examination prior to the onset of the present illness was made on Feb. 25, 1948, following return to work. A complete physical examination at this time revealed no cardiac murmurs and a roentgenogram of the chest (fig. 1, A) disclosed bilateral pulmonary tuberculosis, a mediastinum of normal width, and no cardiac enlargement. Bilateral intrapleural pneumothoraces were maintained.

On April 27, 1948, the patient noted the onset of intermittent substernal pain without radiation and not related to exertion. On May 7, 1948, as he stepped down from a streetcar, he suddenly became short of breath and developed severe substernal pain radiating into the neck and the left arm. The pain persisted for twenty-four hours and then gradually subsided during the next four days. The shortness of breath was constant and moderately severe. X-ray examination of the chest on May 18, 1948 (fig. 1, B) revealed a convex bulge of the right border of the mediastinum at the level of the hilus of the right lung. This was thought to represent a loculated pleural effusion and the patient was permitted to continue in his employment as a piano player. However, because of constant shortness of breath, epigastric pain, and anorexia of four days’ duration, he was readmitted to the hospital on June 12, 1948. On the day before admission there was one episode of expulsion of dark brownish-red vomitus.

The temperature was 37.5° C., the pulse rate 110, the respiratory rate 30, and the blood pressure 170/70. Physical examination revealed a poorly nourished and chronically ill white man who was slightly cyanotic. There was a moderate funnel-shaped deformity of the chest. Bronchial breathing and increased dullness to percussion were noted over the apices of both lungs. Coarse râles were audible over the posterior base of the right lung. The heart was not clinically enlarged. A blowing, high-pitched diastolic murmur was heard over the third left parasternal area, transmitted well to the aortic auscultatory area. A blowing systolic murmur was heard best at the base and transmitted faintly to the apex. The second aortic sound was barely audible. The edge of the liver was barely palpable and slightly tender. There was no edema of the extremities or enlargement of lymph nodes. The reflexes were normal.

The hemogram revealed 10 grams of hemoglobin, and 3,500,000 erythrocytes per cu. mm. of blood. The urine was normal. The reaction to the Kline test for syphilis was negative. An x-ray examination of the chest revealed findings similar to those shown in the previous film. In addition there were an apical pneumothorax on the right, a small pleural...
effusion at the right base, and scattered mottling throughout both lungs with several foci of calcification within the right and left upper lobes.

The clinical diagnoses were: loculated pleural effusion at the hilus of the right lung and bilateral pulmonary tuberculosis. On the second hospital day, aspiration of the pleural effusion was attempted. An 18-gage needle was inserted into the right second intercostal space 2 cm. from the border of the sternum and directed in an inferior and medial direction. At a depth of approximately 6 cm. the syringe suddenly filled with arterial blood and the needle was withdrawn. Shortly afterward the patient became pale, apprehensive, and severely dyspneic. The blood pressure was unobtainable and death occurred ten minutes after the aspiration.

Gross Postmortem Findings. The pericardial sac was distended with approximately 800 cc. of bloody fluid and blood clots. A needle puncture tract over the anterior surface of the pericardium opened into a sac of the aortic wall. This sac formed at the level of the sinuses of Valsalva, beneath the superior reflection of the pericardium, and extended for a distance of 6 cm. along the ascending aorta. The sac measured 2 cm. in depth and extended around all but 2 cm. of the circumference of the base of the aorta. There was slight compression of the base of the pulmonary artery (fig. 2). The lower part of the external wall of the sac was covered on its outer surface by blood-stained epicardium and the upper portion by adventitia and fibrous tissue of the mediastinum. The sac was filled with blood, and when evacuated, presented a pale, yellowish-gray, finely irregular lining surface. The internal wall of the sac was composed of a thin lamina of aortic wall. The position of the puncture holes of the sac indicated that a needle had entered the pericardial sac, pierced the external sac wall, traversed this sac, and then punctured the internal wall to enter the lumen of the aorta. Gross examination of the remainder of the aorta was not remarkable except for scattered yellowish-gray intimal plaques particularly about the ostia of the intercostal vessels and between the renal arteries and iliac bifurcation. There was no evidence of a healed intimal tear.

The heart weighed 480 grams. The right and left ventricles measured 0.6 and 1.6 cm. in thickness respectively. The circumferences of the heart valve
FIG. 2.—The base of the heart showing the great vessels and the intramural sac of the aorta.
rings were within normal limits. The leaflets of the mitral valve were slightly thickened and somewhat nodular along the line of closure. There was slight fusion of the commissures of the aortic cusps. None of these valvular changes appeared to be functionally deforming.

The right and left lungs weighed 600 and 700 grams respectively. The bases were adherent to the thoracic wall by fibrous adhesions and the apices were smooth except for a single fibrous band attaching the right apex to the overlying cupola. Approximately 200 cc. of clear pale-yellow fluid were present in each pleural cavity.

There was recent erosion of the mucosa of the esophagus and severe chronic passive hyperemia of the gastric mucosa and of the liver. The ileum revealed two approximately round ulcers of the mucosa with thickened borders and dark reddish-gray granular bases. Similar mucosal ulcers were found in the colon. The remaining findings of the autopsy were not remarkable.

Microscopic Postmortem Findings. The sac had been formed by a separation of the media of the aorta so that the outer wall was composed of loose, young fibrous tissue containing a thin layer of elastic fibrils. The inner wall of the sac was composed of aortic media lined on one surface by intima and on the sac surface by young fibrous tissue covered by endothelium. The elastic fibrils of the inner wall of the sac composing the media of the aorta were irregularly disposed and revealed minute foci of necrosis. The lining surface of the sac was covered by endothelium beneath which were a few phagocytes containing brown pigment granules that were positive for iron stain. Several small, similar sacs, occasionally filled with blood, were present within the aortic media just proximal to the large sac. Microscopic sections from the thoracic and abdominal aorta revealed no changes other than those of moderate arteriosclerosis.

Microscopic examination of the lungs revealed fibrocaseous tuberculosis, focal pulmonary fibrosis, and chronic pulmonary emphysema. The esophageal ulcers were nonspecific and those in the ileum and colon were tuberculous.

The pathologic diagnosis was dissecting aneurysm of the aorta with intramural aortic sac. There had been perforation of this sac by a needle with hemopericardium and cardiac tamponade. Other pertinent diagnoses included hypertrophy and dilatation of the heart, nondeforming endocarditis of the mitral and aortic valves, fibrocaseous pulmonary tuberculosis, ulcerative tuberculous enterocolitis, and acute and chronic esophagitis.

Discussion

The differential diagnosis of dissecting aneurysm of the aorta has been discussed in numerous publications and does not merit mention here. A review of 698 cases of dissecting aneurysm of the aorta by one of us in a previous report disclosed no similar instance of aortic dissection that was clinically mistaken for a paramediastinal effusion. In retrospect the history was fairly typical, and the appearance of a well-defined aortic diastolic murmur only a few months after the patient’s heart had been found normal should have made one suspicious of the correct diagnosis. There was no evidence pointing to a recent inflammatory lesion of the aortic valve.

Healed dissecting aneurysm of the aorta has been clinically mistaken for tumor of the mediastinum. In the case reported by Patrick and Taylor the patient, a 32 year old white man, underwent an exploratory laparatomy two years after the onset of intermittent epigastric and low left-sided chest pain, a history suggesting gastric ulcer, which was not found at operation. He later developed progressive dysphagia and chest roentgenograms showed a large, nonpulsating mass which displaced the esophagus forward and to the left. Thoracotomy was undertaken and upon incision of the mass marked hemorrhage occurred. Death occurred one week later, and necropsy revealed a large dissection filled with laminated clot extending from the arch of the aorta to 4 cm. below the diaphragm where a small re-entry foramen was found. At the proximal end of the dissection there was a transverse scar of the intima that was thought to be a healed intimal tear.

Von Möllar described the first case to be reported of dissection of the aorta in the absence of rupture and of intimal tear. In Krukenberg’s patient, reported in 1920, the main aortic dissection communicated with the lumen of the aorta, but in both inferior thyroid arteries there were numerous raised cystlike nodules, within the media, filled with blood and very similar to the smaller lesions found in contiguity with the main dissection in our patient. The third such case reported was that of Whitman and Stein in 1924, as an incidental finding at necropsy. There was a healed, endotheialized dissection of the aorta extending from the root of the aorta to within 10 cm. of the iliac bifurcation. The sac surrounded the
aorta in its entirety with the exception of the posterior portion overlying the vertebral bodies and was filled with a clear, lymphlike fluid. There was no evidence of an intimal tear and the sac had evidently never communicated with the lumen of the aorta.

These cases support the tenet first propounded by Babes and Mironescu,2 and later clarified by Erdheim,3 that medial disease precedes the intimal tear, with secondary rupture of the intima and forceful extension of the dissection by the column of blood. In our patient the contents of the sac during life could not be ascertained because of the unique perforation, but there was certainly no evidence of ante-mortem clot. Here, too, the sac may have contained a transudate.

The clear-cut appearance of a classical aortic diastolic murmur likewise supports the now generally accepted thought that such a murmur is due to insufficiency created at the aortic orifice by distortion caused by the bulging dissection, as opposed to the views of Letulle6 and of Keefer and Resnik4 who held that the diastolic murmur arose in the eddies produced at the lip of the intimal tear by the inflow and outflow of blood.

In our patient there were signs of early congestive failure which most likely would have caused death at a later date. Gouley3 has shown that when healed dissecting aneurysm produces aortic insufficiency, death occurs in congestive failure indistinguishable from that due to intrinsic lesions of the aortic valve.

**Summary**

A case of chronic healed dissecting aneurysm of the aorta is presented in which the dissection was limited to the media; there was no intimal tear, no rupture, and no area of reentry. The dissection was mistaken for a para mediastinal effusion, and death was due to cardiac tamponade incident to exploratory needle aspiration. At necropsy the needle was found to have penetrated the superior pericardial reflection, the outer sac wall, the sac cavity, and the lumen of the aorta.

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