Dissecting Aneurysm of the Aorta Secondary to Tuberculous Aortitis

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TUBERCULOUS aortitis is rare, only 29 cases having previously been reported. All but 1 of these have been associated with aneurysm, but in none has dissection been present. We believe the following to be the first reported case of dissecting aneurysm secondary to tuberculous aortitis.

CASE REPORT

G. P., a 55-year-old man was admitted to the hospital on March 28, 1953, with abdominal pain of 1 week's duration, cough, and dyspnea. The pain radiated to the chest, back, and interscapular region. There was a history of genitourinary and skeletal tuberculosis dating back to 1919, and the left epididymis and left kidney had been removed. Hypertension had been present for 6 or 7 years. Since April 1952 he had been treated for recurrent tachycardia and syncope. The past history was otherwise not significant, and the family history was negative.

On physical examination, the blood pressure was 230/130, the heart was enlarged, and there were signs of mild congestive heart failure. Grade III hypertensive retinopathy and optic atrophy of the right eye were present.

There was a normal hemogram, the blood urea nitrogen was normal, and the serologic test for syphilis was negative. An electrocardiogram showed left ventricular hypertrophy and digitalis effect.

Roentgenologic examination of the chest revealed cardiac enlargement, chiefly left ventricular, a tortuous elongated aorta, and old healed calcific tuberculosis in both upper lobes (fig. 1). An intravenous urogram disclosed a normal right pelvocalyceal system and ureter; the left was not visualized. X-rays of the spine revealed evidence of both healed and active tuberculosis involving the first 4 lumbar vertebrae.

Several days after admission hoarseness developed, and prominence of the left hilum was seen in the chest films (fig. 1A). Planigraphic examination of the left hilum demonstrated that the enlargement was due to vascular shadows and not to a tumor mass. Bronchoscopy showed that the hoarseness was due to paralysis of the left vocal cord, but no endobronchial lesion was seen. There was some decreased mobility of the left main bronchus. Chest fluoroscopy on April 22, approximately 3 weeks after admission, revealed deviation of the esophagus to the left and narrowing of its lower 2 centimeters. One week later paralysis of the left leaf of the diaphragm was noted. The esophageal changes were thought to represent esophagitis rather than neoplasm from the roentgenologic standpoint. On May 11, the left leaf of the diaphragm was still paralyzed (fig. 2). Tortuosity of the aorta was again observed but it was not noted that an increase in size and irregularity had occurred.

Diagnoses of hypertensive cardiovascular disease and healed pulmonary and bone tuberculosis were made. The cause of the esophageal abnormality and the paralysis of the vocal cord and diaphragm was not determined during this hospital admission.

The patient was readmitted to the hospital in November 1953 in congestive heart failure. Roentgenograms at this time showed an increase in the width of the aortic knob and increased width and tortuosity of the descending aorta (fig. 3). The left leaf of the diaphragm had returned to its normal position and its excursion was normal. There was marked distortion of the esophagus due to extrinsic pressure. A diagnosis of dissecting aneurysm of the aorta was made. The previously unexplained vocal cord and phrenic paralysis was attributed to pressure on the phrenic and recurrent laryngeal nerves by the aneurysm.

The patient was again hospitalized in July 1955 for congestive heart failure. The blood pressure was 210/150. Except for an apparent increase in heart size, there were no significant differences from the previous physical findings. Roentgenograms of the chest revealed further increase in the width of the thoracic aorta.

The patient was admitted for the last time in September 1955 in acute congestive heart failure. There was no chest pain. Despite therapy, he died 1 hour later.

At autopsy there was a dissecting aortic aneurysm extending from just beyond the origin of the left subelavian artery to a level just above the origin of the renal arteries. The proximal portion was sacu-
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Fig. 1. Initial roentgenogram of the chest showing cardiac enlargement, tortuous elongated aorta, and old healed calcific tuberculosis in both upper lobes. A. Posteroanterior projection. B. Left lateral projection.

Fig. 2. Chest roentgenogram showing increased width of aorta and paralysis of left leaf of diaphragm. A. Posteroanterior projection. B. Left lateral projection.

lar, measuring 6.5 by 6 by 3 cm.; the distal portion was cylindrical, 17 cm. in length (fig. 4A). The celiac and superior mesenteric arteries were not involved in the process, but both common iliac arteries were markedly dilated and had aneurysmal bulges containing mural thrombi. In the opened, dissected segment communication with the true aortic lumen could be seen at the junction of the saccular and cylindrical portions. At the distal end of the dissection, recommunication with the true aortic lumen had occurred (fig. 4B). Within the false lumen were several organizing thrombi. Marked atherosclerosis, with numerous areas of ulceration, was present throughout the aorta.

Microscopic examination of the aortic wall revealed 2 distinct processes: severe ulcerative atherosclerosis, and caseating granulomatous involvement of the adventitia (fig. 5A). In areas not involved by dissection there was abnormal intima of varying thickness, but the fibroelastic media was intact, and the adventitia was normal. Toward the area of dissection there was gradual transition in the adventitia to a longitudinally oriented mass of caseating granulomatous tissue. It was in this layer that dissection had taken place, so that the false lumen was lined by caseation tissue. At the proximal and distal ends of the dissection, the granuloma had extended into and destroyed the fibroelastic media. At these 2 points communication between the true and false aortic lumen had taken place through an intimal tear. The granuloma was consistent in appearance with tuberculosis (fig. 5B), and acid fast
FIG. 3. Chest roentgenogram showing further increase in width and tortuosity of aorta and marked distortion of esophagus by extrinsic pressure. Diaphragm has returned to normal level. *A.* Posteroanterior projection. *B.* Lateral projection.

FIG. 4. *A.* Autopsy specimen showing extensive involvement of aorta by dissecting aneurysm. *B.* Dissected segment open showing points of communication between true and false aortic lumen *(C)* and large laminated thrombus in saccular portion *(Thr).*
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FIG. 5. A. Low-power photomicrograph (×12) showing involvement of adventitia (Adv) by caseating granulomatous tissue. At upper right, adventitia is split by lumen of dissecting aneurysm (Fl). The media (Med) is intact at right but toward the left becomes involved by the granuloma, which extends to and breaks through the atheromatous intimal layer (Int). Large thrombus (Thr) lines true aortic lumen (T1). B. Higher power photomicrograph (×65) showing granuloma with many giant cells consistent with tuberculosis. Tubercle bacilli were demonstrated in other Kinyoun-stained sections.

bacilli were demonstrated in Kinyoun-stained sections. There was diffuse tuberculous involvement of the chest wall, lumbar vertebrae, lungs, adrenal glands, liver, spleen, prostate, and the stump of the left ureter, but no involvement of periaortic lymph nodes was found.

DISCUSSION
Tuberculosis of the smaller blood vessels, either by local extension or as part of a diffuse miliary process, is not uncommon. Tuberculous involvement of the large vessels, however, particularly the aorta, is rare. Only 29 cases of tuberculous aortitis have previously been reported. Twenty-one of these were included in a review by Gellerstedt and Säfvenberg in 1933 and since then 8 additional cases have been reported.2-9 All the other cases except that of Waser3 have been associated with aneurysm,
but ours is the first in which dissection of the aorta has been present. The aorta was most frequently involved by contiguity, usually by spread from adjacent lymph nodes. In 5 cases, including the present one, the infection was apparently blood borne.

A specific etiology for aortic dissection is rarely recognized and the majority of dissections are classified as "idiopathic." Dissecting aneurysm, as it presents itself clinically, probably results from 2 distinct processes. Splitting of the aortic wall is usually attributed to disease of the media, so-called medionecrosis cystica, and rupture of diseased medial nutrient vessels is believed to be responsible for the formation of a hematoma within the dissected segment. Atherosclerotic degeneration of the intima then permits an intimal tear, with resulting communication between the dissection and the aortic lumen. The present case differs in that the dissection occurred in the adventitia instead of the media. The adventitia was widely involved by a specific granulomatous process and at only 2 points did the necrosis extend to involve the media. Here the process merged with the atheromatous intima and the intimal tear occurred, establishing communication with the aortic lumen.

Whether aortic dissection is of known etiology or "idiopathic," certain predisposing factors are recognized. These are coarctation and other anomalies of the aorta, pregnancy, Marfan's syndrome, and hypertension. Hypertension is nearly always present at the time of dissection. Even in those patients whose blood pressure is not elevated, some evidence of hypertensive vascular disease is usually found either in the history, the physical findings, or the findings at autopsy. In some cases, however, the hypertension may be the result of the dissecting aneurysm rather than a predisposing cause, by a mechanism similar to that in coarctation of the aorta, with proximal hypertension and distal hypotension.

The clinical picture of dissecting aneurysm has been described elsewhere and will not be discussed in detail here. Several features, however, deserve re-emphasis. The presenting complaint most frequently encountered is severe pain in the chest or abdomen, radiating often to the back, the interscapular region, the neck, and the legs. Although this patient's initial pain had the characteristic distribution it was mild and did not persist. It should be recognized that dissection may be gradual, and the classical acute or subacute clinical picture of pain, shock, collapse, and death in a matter of hours or days may not be present. Extensive dissection has been observed at autopsy from the aortic arch to the bifurcation of the aorta without any history of symptoms referable to the lesion.

Of particular clinical interest in the present case was the occurrence of hoarseness due to involvement of the recurrent laryngeal nerve, which has previously been reported, and the transient phrenic paralysis to which we can find no previous reference.

The present case is an example of so-called "healed" dissecting aneurysm, resulting from distal re-entry from the dissection into the true aortic lumen. This is reported to occur in approximately 25 per cent of aortic dissections. The patient may die later as a result of rupture of the aneurysm but more commonly death results from some other related or unrelated cause. Recent attempts at surgical therapy of dissecting aneurysm have been patterned after this natural "healing." They have been directed at re-establishment of aortic flow by the production of a fenestration between the dissection and the true aortic lumen. The ultimate success of this approach remains to be demonstrated.

Summary

Tuberculous aortitis is rare, only 29 cases having previously been reported. The first known case of dissecting aneurysm of the aorta secondary to tuberculous aortitis is described. The site of dissection was in the adventitia instead of the usual location in the media. The possibility of gradual aortic dissection without the classical acute picture is emphasized.

Summary in Interlingua

Aortitis tuberculotic es rar. Solmente 29 previe casos se trova in le litteratura. Es hie describite le prime cognoscite caso de aneuryismos disseccante del aorta, occurrente secundari
a aortitis tuberculotic. Le sito del dissection eseva le adventitia in loco del usual sito in le media. Es signalate le possibilitate de un dissection aortic gradual sin le acute tableau classic.

REFERENCES


Other schools have earned a reputation in physiology and comparative anatomy, and those branches of medicine which are termed theoretic, but the enduring fame of the Dublin contributions to science arises from their essential practicality and truthfulness. They are records of unbiased observation made by men originally well educated, and brought up in a practical school. — William Stokes His Life and Work (1804–1878) by his son William Stokes. London, T. Fisher Unwin, MDCCCXCIII, p. 162.
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