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AORTIC VALVULAR INSUFFICIENCY ASSOCIATED WITH RHEUMATOID ARTHRITIS*

DORRANCE BOWERS, M.D., M.S.(Med.), F.R.C.P.[C.], *Kelowna, B.C.*

THE FOLLOWING CASE REPORT is considered of interest for three reasons: (1) it describes the occurrence of acute rheumatic fever, rheumatoid spondylitis, and rheumatoid arthritis in the same individual; (2) it demonstrates that aortic valvular insufficiency may be tolerated for 23 years; and (3) it illustrates the uncommon clinical syndrome of chronic rheumatoid disease, delayed conduction of the stimulus to cardiac contraction, and severe aortic valvular insufficiency in association with the pathological findings of aortitis and aortic valvular endocarditis.

Clinical Findings

In 1916, an 18-year-old, white, Canadian-born soldier developed fever in association with acutely painful swollen hot ankles. His illness was diagnosed as acute rheumatic fever. His symptoms cleared in two weeks without apparent residual effects. Following this, he had frequent sore throats attributed to recurrent tonsillitis. For this reason, his tonsils were removed in 1919. Through the 1920's, he had intermittent lumbar backache and stiffness. In 1925, he was said to have a sacroiliac subluxation. By 1930, he had a "poker spine". In 1935, an aortic diastolic murmur was first recorded.

In the years between 1935 and 1946, he had almost continuous inflammatory involvement of the peripheral joints. An attempt at gold therapy in 1938 was discontinued because of the development of a skin rash. In 1945, he went to bed, never to be up again. In bed, his peripheral joints became fused.

In 1947, he developed severe bilateral iritis. With the formation of secondary cataracts, he became totally blind.

Symptoms of left ventricular failure began in 1957. In the month before his death in 1958, the following observations were made: bilateral cataracts; total spinal fusion; slight mobility in the shoulders; "opera-glass" hands; subluxations of the toe joints; fusion of the remaining peripheral joints; shaking of his whole body and bed with each heart beat; aortic diastolic murmur grade iv (out of vi) in intensity; grade iii precordial systolic murmur; blood pressure 160/50 mm. Hg; Corrigan pulse; "pistol-shot" sounds over the femoral arteries; increasing pulmonary congestion; hæmoglobin level 9.6 g. % per 100 ml.; red cell count 3.5 million per c.mm.; hæmatocrit 33%; sedimentation rate 58 mm. in one hour (Wintrobe method, not corrected for anæmia); electrocardiographic pattern of left ventricular hypertrophy; PR interval 0.24 second (Fig. 1). Repeated serologic tests for syphilis at irregular intervals throughout his illness were negative.

Gross Pathological Findings

The locomotor and cardiorespiratory symptoms were of particular interest. (The eyes were not examined.)

*From the Knox Clinic and the Kelowna General Hospital.

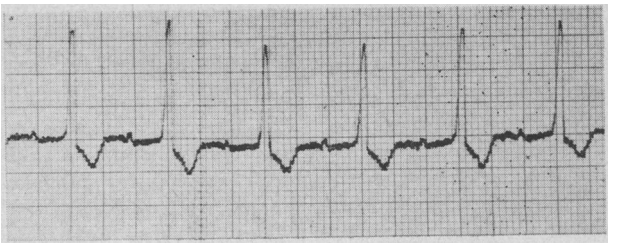

Fig. 1.—Electrocardiogram: standard lead I. The PR interval is 0.24 second in duration; the RST segment is depressed; the T wave is sharply inverted. These findings were interpreted to indicate delayed atrioventricular conduction and left ventricular hypertrophy. The patient was taking digitalis in small doses at the time the tracing was made.

The entire spinal column was fixed in a shallow curve (convex posteriorly). There was slight mobility at both shoulders; the joints of the fingers and toes were flail joints. The other peripheral joints were fused.

The heart was massively enlarged, weighing 1060 grams; this enlargement was due to hypertrophy of the left ventricular myocardium, the anterolateral wall measuring more than 2.5 cm. in thickness at some sites. The left ventricular cavity was monstrously dilated. The chambers and valves on the right side of the heart were normal. The left atrium was slightly dilated; the annulus of the mitral valve was also dilated, permitting mitral valvular insufficiency. The leaflets of the mitral valve were thin and felt normal in consistency; the chordæ tendinæ were normal. The annulus of the aortic valve and the ascending aorta were dilated. The commissures of the aortic valve were separated; the cusps of the aortic valve were thickened and contained palpable plaques of calcium at their bases; the free edges of the cusps were rolled and retracted.

There were large bilateral pleural effusions. The lungs were œdematous.

Histological Findings

Microscopic examination of the left ventricular myocardium showed no evidence of active or healed Aschoff nodules. Patchy areas of myocardium showed ischæmic changes. Focal collections of lymphocytes were present in the basal portions of the mitral valve leaflets.

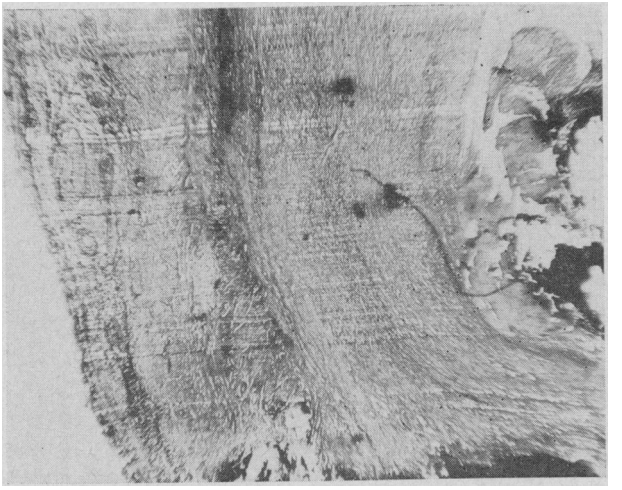

Fig. 2.—Photomicrograph of ascending aorta (Mallory's stain for elastic fibres). The right coronary cusp of the aortic valve appears on the right. Note the thickened rolled free edge of the cusp and the fragmented calcific deposits at the base of the cusp. The elastic fibres in the root of the aorta are few in number and disorganized in pattern.

Sections through the aortic valve and aortic valve ring showed the presence of interwoven bundles of hyalinized connective tissue in the free edge of the cusps. Towards the basal portions of the valve cusps, in addition to this hyaline degeneration, well-defined calcific deposits were present. In relation to these deposits, there were focal collections of lymphocytes and plasma cells showing no particular evidence of perivascular arrangement.

Elastic-tissue stains revealed a rather marked fragmentation and loss of elastic fibres at the base of the aortic valve and in the first portion of the ascending aorta (Fig. 2). The small vessels in the adventitia had thickened walls and narrowed lumina; this thickening appeared due to subintimal hyaline deposition rather than to endothelial proliferation.

DISCUSSION

The "rheumatic" diseases are not merely arthritides but systemic diseases. Cardiovascular involvement in the "rheumatic" patient, then, should not be considered a complication, but should be regarded as an integral part of the disease. For reasons that are presently obscure, clinically significant cardiovascular involvement in rheumatoid arthritis and spondylitis is uncommon. However, a small number of cases have been reported establishing a syndrome of severe aortic valvular insufficiency related to pathologically demonstrable aortitis and aortic valvular endocarditis in rheumatoid disease.

One of the earliest cases of this syndrome to be reported was presented at the weekly clinicopathological conferences[1] at the Massachusetts General Hospital in 1936. The patient was a young man (26 years old at the time of his death) with typical peripheral rheumatoid arthritis. He was anæmic and had an elevated sedimentation rate. His electrocardiogram showed left bundle branch block. He had advanced aortic valvular insufficiency and left ventricular failure. Postmortem examination revealed an active aortitis and aortic valvulitis.

By 1951, Walter Bauer and his associates[2] at the Massachusetts General Hospital felt that they had accumulated sufficient data "to conclude that aortitis and aortic endocarditis do occur as a manifestation of rheumatoid arthritis. The clinical and pathological features of this type of heart disease are sufficiently distinctive to exclude syphilis and rheumatic fever as etiological agents."

Recently, Schilder, Harvey and Hufnagel,[3] at Georgetown University, after evaluating 100 patients with aortic insufficiency for possible surgical treatment, collected a series of 6 cases (5 in the body of their report, one in an addendum) with certain common clinical features: (1) long-standing rheumatoid spondylitis; (2) elevation of the sedimentation rate; (3) anæmia; (4) severe aortic valvular insufficiency; (5) radiological and electrocardiographic evidence of left ventricular hypertrophy; (6) prolonged atrioventricular conduction; and (7) negative serologic tests for syphilis. In the two of their cases in which pathologic data were presented, there was aortic valvulitis. In the one case in which the root of the aorta was examined microscopically, "all the layers were greatly scarred and thickened by hyalinized fibrous tissue".

The total number of similar cases with both clinical and pathological documentation reported in the medical literature is small. This probably reflects the true rarity of this syndrome, for a large number of intensive investigations[4] of patients with rheumatoid disease have indicated a low incidence of clinically significant heart disease.

The patient described in this case report demonstrated all the clinical and pathological features of this syndrome of rheumatoid aortic valvulitis. His skeleton was almost totally fused. He was anæmic and had an elevated sedimentation rate. Repeated serological tests for syphilis were negative. He had the central and peripheral signs of severe aortic valvular insufficiency. It is interesting that, although this aortic insufficiency was tolerated for 23 years, it eventually resulted in his death. His electrocardiogram revealed first-degree heart block. The pathological findings indicated a long-standing aortic valvulitis and aortitis. Dilatation and hypertrophy of the left ventricle were marked. The aortic valve was incompetent because of separation of the commissures and retraction of the free edges of the valve cusps. Valaitis, Pilz and Montgomery[5] ascribe the deformities of the aorta and aortic valve to fibrosis of necrobiotic rheumatoid lesions. Microscopic examination showed that the inflammatory reaction was much less acute than that described in the early case[1] observed at the Massachusetts General Hospital (the total duration of that patient's illness was approximately seven years), but was similar to that observed in the second case reported by Schilder *et al.*[3] (the total duration of that patient's illness was 10 to 20 years). In these three cases (M.G.H., 1936;[1] Schilder *et al.*, 1956;[3] and Bowers, 1958), the absence of Aschoff bodies has been specifically emphasized.

The clinical picture and the pathological findings in the case reported in this paper lend confirmation to the conclusions clearly stated by Bauer and his associates[2] that aortitis and aortic endocarditis do occur as a manifestation of rheumatoid disease, and that this form of heart disease is a specific entity distinct from rheumatic and luetic heart disease.

SUMMARY

The case history of a patient with total skeletal fixation due to rheumatoid arthritis, total blindness due to bilateral iritis with secondary cataracts, and fatal aortic valvular insufficiency is presented in detail. It is suggested that the aortic valvular insufficiency was caused by a specific form of aortitis and aortic valvular endocarditis distinct from that occurring in rheumatic and luetic heart disease.

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BASILAR ARTERY OCCLUSION REPORT OF A CASE WITH TEN YEARS' SURVIVAL

HARRY P. ENGEL, M.D.,* *Edmonton, Alta.*

THROMBOSIS of the basilar artery is regarded as a rare and difficult clinical diagnosis, and although the majority of cases result in a fatal outcome, recoveries have been reported, as for example by Freeman and Ellis.[1]

The first recorded case of basilar artery occlusion was by Hayem[2] in 1868, and seven years later Leyden[3] published a report of two cases of basilar artery thrombosis from syphilitic endarteritis. In 1946, Kubik and Adams[4] made a survey of the incidence of basilar artery occlusion and found 13 cases in 4200 consecutive autopsies at the Massachusetts General Hospital, and a further 12 cases among 3200 consecutive autopsies at the Boston General Hospital, making a median average of 1:300 autopsies. Of these 25 cases no clinical data were available in 7, and of the remaining 18 the occlusion was found to be due to thrombosis in 11 and to embolism in 7. Survival following complete occlusion of the basilar artery either by disease[5, 6] or by surgical ligation[7] has been reported in the literature. Survival is attributed to the effectiveness of anastomotic connections between the cerebral arteries which permit bypass of the block in the basilar artery. Extensive communications between the middle and posterior cerebral artery and among the long cerebellar vessels have been described.[6-9]

SIGNS AND SYMPTOMS

The clinical signs and symptoms of basilar artery occlusion are typical, and diagnosis should be possible in the majority of cases. The principal features are the sudden onset with initial symptoms of headache, dizziness, disorientation and coma. Temporary improvement is common but in the majority of cases death occurs within 2 to 30 days. Other characteristic features which may be present in varying degrees are:

1. Dysarthria and unilateral paræsthesia.
2. Facial palsy.
3. Hemiplegia or quadriplegia with bilateral plantar extensor reflexes.
4. Pupillary abnormalities and disorders of ocular movements.
5. Normal cerebrospinal fluid.

This is a case report of a patient who survived for ten years following basilar artery occlusion.

The patient, a married white male aged 42 years, was admitted to Bridgeport Hospital, Bridgeport, Connecticut, on June 6, 1955. He gave a history of an episode of left-sided paralysis occurring ten years earlier and accompanied by diplopia, difficulty in swallowing and dysarthria. During this period he was treated at home by his family physician, and his condition improved rapidly so that by the seventh day he had no apparent residual disability. No definite diagnosis was made at that time. He remained in good health for the next ten years, except for regular episodes of impairment of consciousness occurring approximately every six months, for which he did not seek medical advice.

On his final admission he gave a history of left hemiplegia of three days' duration and sudden onset.

Physical Examination

The patient was well nourished and well developed, conscious and co-operative. There was complete paralysis of the left arm and left leg, and slight weakness of the left side of the mouth. The left pupil was larger than the right pupil and did not react to light. The left eye tended to deviate to the left and there was nystagmus of the left eye only on left lateral gaze. Movements of the left eye were possible in all directions but medial movement was weak. The right eye could be moved up and down normally but could not be moved either medially or laterally. Speech was dysarthric and co-ordination could not be tested. Over the next eight days his condition improved slightly with the return of some weak function in his left arm and leg and return of normal speech. On the ninth day the patient developed peripheral vascular collapse from which he slowly recovered, but without any alteration in his neurological status. After this he made a gradual but progressive recovery and on the 17th day after admission he was discharged to a nursing home; there was only moderate function in his left side but he was able to walk with the assistance of a "walker". Automatic bladder function was present. The condition was diagnosed at that time as an acute episode of multiple sclerosis.

Readmission

On August 15, 1955, the patient was readmitted with an acute recurrence. This time he was completely unable to move his body or his extremities or any muscles supplied by his cranial nerves and could make only inarticulate vocal sounds. He was capable of understanding to some degree when spoken to, but unable to carry out requested movements. Deviation of the head to the right was present; pupils

*Late Assistant Surgical Resident, Bridgeport Hospital, Bridgeport, Connecticut, U.S.A.
Present address: McEachern Cancer Research Laboratory and Department of Surgery, University of Alberta, Edmonton.