AN AUTOPSY CASE OF RHEUMATOID ARTHRITIS WITH AN INVOLVEMENT OF THE CARDIAC CONDUCTION SYSTEM

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A case of rheumatoid arthritis with an involvement of the cardiac conduction system was described. The patient was a 65-year-old man who had a 15-year history of classical rheumatoid arthritis combined with an advanced atrioventricular (AV) block resulting in Adams-Stokes seizure. Prior to the occurrence of the AV block, a complete right bundle branch block with a left axis deviation and a first-degree AV block were found on an electrocardiogram (ECG). The histological examination of the conduction system according to the serial sectioning method disclosed that the branching portion of the His bundle and anterior fascicle of the left bundle branch were replaced by the scarring tissue and that the penetrating portion of the His bundle, the AV node and the right bundle branch had rheumatoid granulomatous lesions. The extent and localization of the lesions in the conduction system were well correlated with the findings on the ECG.

ALTHOUGH chronic polyarthritis is the most prominent feature in rheumatoid arthritis, extraarticular manifestations including cardiac involvement are well recognized.^{1,2} Pericarditis is most common but the occurrence of a mild-degree heart block has also been documented.^{3,4} Usually the heart block does not develop into an advanced or complete AV block, probably because of the little organic impairment of the conduction system. Only 8 cases⁴⁻¹⁰ in which the conduction system was partially or entirely examined have been reported so far and the pathology of the conduction system associated with rheumatoid heart disease is still poorly understood.

Recently we have had a case of classical rheumatoid arthritis combined with an advanced AV

block. In the present paper, we describe the detailed histopathology of the conduction system involved by rheumatoid arthritis.

CASE REPORT

Clinical History

A 51-year-old man with a 7-year history of classical rheumatoid arthritis was admitted to Naruwa Hospital, Kanazawa, on November 1, 1973 for further treatment. On admission, there were deformities of multiple joints, including the fingers, knees, elbows and ankles. No cardiac abnormalities were found. On November 9, immediately after the synovectomy of the left knee joint, he developed rheumatoid pericarditis which was diagnosed by chest pain, rapid pulse rate with irregular rhythm, a grade 2/6 ejection systolic murmur, atrial fibrillation and triphasic rub by phonocardiography. Abnormal laboratory data was as follows: white blood cell count, 9,500/mm³ with neutrophilia; C-reactive protein, 5(+); rheumatoid factor, 3(+). This symptom

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Fig.1. Electrocardiogram showing a complete right bundle branch block with a left axis deviation and a first-degree AV block.

was subsided 10 days later and his electrocardiogram (ECG) was completely normalized. After that episode, his ECG remained normal until December, 1976 when an ECG showed a complete right bundle branch block with a left axis deviation and a first-degree AV block (Fig. 1). One month later, he had Adams-Stokes seizure due to an advanced AV block which necessitated the use of a cardiac pacemaker (Fig. 2). By this procedure, his cardiac rhythm was well controlled. On May 25, 1978, however, he developed syncope associated with anisocoria and abnormal involuntary movement of the left hand. Computerized tomographic scan revealed the low density area at the left cerebellar hemisphere, suggesting cerebellar infarction. Later

on, he gradually deteriorated and died on June 21, 1981 of cachexia and bronchopneumonia. He received no adrenocortical hormones throughout the course.

Pathological Findings

At autopsy, the body was emaciated and there was a decubitus ulcer over the right greater trochanter. There were deformities in the multiple joints of the fingers, elbows, ankles and knees. The histologic examination showed that the pannus with mild lymphocytic infiltration covered on the surface of the destroyed articular cartilage. A lipoid nodule¹¹ was found at the right elbow.

The heart weighed 350g and the complete pericardiac fibrous adhesion was noted. The left ventricle was slightly hypertrophied. The cardiac valves showed no abnormalities, except for mild thickening of the anterior leaflet of the mitral valve at the base. No appreciable pathological changes were observed in the endocardium, the myocardium and the coronary arteries.

The sinus node and the AV conduction system were serially sectioned according to the method of Davies.¹² Every tenth and eleventh sections were stained with hematoxylin and eosin, and with Weigert-van Gieson stains, respectively. Also, some of them were stained with Congo red in order to show the presence of amyloid. On the histological examination, the sinus node was unremarkable. The AV node was partially replaced by fibrous granulation tissue with local infiltration of small round cells (Fig. 3). There were rheumatoid nodules in the central fibrous body. The penetrating portion of the His bundle was disrupted by the rheumatoid nodules, chronic inflammatory cell infiltration and fibrosis (Fig. 4). There was severe fibrosis in the branching portion of the His bundle. The anterior fascicle of the left bundle branch was completely interrupted by the scarring tissue in the summit of the interventricular septum (Fig. 5). The posterior fascicle of the left bundle branch was almost intact. The rheumatoid nodules were also

Fig.2. Strip of standard lead III of the electrocardiogram recorded during an Adams-Stokes seizure. Advanced AV block is present.

Fig.3. Atrioventricular node (AV) showing partial replacement by fibrous granulation tissue and chronic inflammatory cell infiltration (HE stain, x36). CFB = central fibrous body

Fig.4. The penetrating portion of the His bundle. Note severe destruction of conduction cells by rheumatoid nodule (RN), inflammatory cell infiltration and fibrosis (HE stain, x36).

observed in the summit of the interventricular septum. The right bundle branch was markedly infiltrated by small round cells in the first portion in the summit of the interventricular septum (Fig. 6). Non-specific inflammation was observed in the aortic and mitral valves, especially at the base. There was no histological evidence suggesting of arteritis, myocarditis, or amyloido-

sis.

In the other visceral organs no pathological changes were found except for bronchopneumonia.

DISCUSSION

Clinically the present patient developed a

Fig.5. Scarring tissue in the summit of the interventricular septum leading to complete interruption of the anterior fascicle of left bundle branch (LBBa). The branching portion of the His bundle (His) shows severe fibrosis (Weigert-van Gieson stain, $\times 36$).

Fig.6. The first portion of the right bundle branch (RBB) disrupted by small round cells and rheumatoid nodule (RN) in the summit of the interventricular septum (HE stain, $\times 36$).

complete right bundle branch block with a left axis deviation and a first-degree AV block and one month later he had an Adams-Stokes seizure due to an advanced AV block. At autopsy, fibrosis was found in the branching portion of the His bundle and the anterior fascicle of the left bundle branch was completely interrupted by the scarr-

ing tissue. The right bundle branch and the penetrating portion of the His bundle were destroyed by rheumatoid nodules, chronic inflammatory cell infiltration and fibrosis. The AV node was slightly involved by granulomatous lesion and chronic inflammatory cell infiltration.

There is a good correlation between the find-

TABLE 1 SUMMARY OF THE REPORTED CASES WITH PATHOLOGICAL STUDIES ON THE CONDUCTION SYSTEM

Authors	Age	Sex	Duration of arthritis	Type of heart block	Pathology of the heart	Systemic studies of conduction system	Pathology of the AV conduction system
Carpenter et al. (1967)	65	F	15 years	First degree & LBBB	Rheumatoid nodules in epicardium, all valves, myocardium and inter-ventricular septum. Pericarditis	Partial	Rheumatoid nodules interrupting proximal portion of left bundle branch
Kirk & Cosh (1969)	69	F	7 years	First degree	Rheumatoid nodules in pericardium and between the roots of both great vessels	Yes	Rheumatoid nodules in AV node, bundle of His and its proximal divisions
Harris (1970)	69	F	7 years	Complete	Rheumatoid nodules in aortic and mitral valves	Yes	Rheumatoid nodules in all parts of AV conduction system
Gallagher & Gresham (1973)	65	F	8 years	Complete	Infected rheumatoid nodules in aortic root and summit of inter-ventricular septum	Partial	Infected rheumatoid nodules involving in AV node?
Thery et al. (1974)	50	M	Several years	Complete	Rheumatoid nodules in base of mitral and aortic valves, and in aortic root	Yes	Chronic inflammation from rheumatoid nodules involving all parts of AV conduction system
Thery et al. (1974)	36	F	More than 10 years	Complete	Secondary amyloidosis. No rheumatoid nodules	Yes	Amyloid deposition in central fibrous body and blood vessels destroying AV conduction system down to its bifurcation
Lev et al. (1975)	52	M	5-6 years	Complete	Scars in the summit of interventricular septum. Thickening and narrowing of arteries. No active inflammation	Yes	Scarring tissue pressing branching bundle, bifurcation and beginning of bundle branch
Takagi (1978)	36	F	?	Second degree (2:1)	Polyarteritis nodosa-like arteritis	Yes	Arteritis and its associated granulation destroying AV node and bundle of His

F = female, M = male

ings on the ECG and the histopathology of the conduction system. The lesions in the right bundle branch, anterior fascicle of the left bundle branch and the branching portion of the His bundle may account for the ECG changes of the right bundle branch block with a left axis deviation and the first-degree AV block. Because of mild involvement of the AV node, the advanced AV block is attributed mainly to destruction of the penetrating portion of the His bundle.

A mild-degree heart block, in rheumatoid arthritis, is not uncommon^{3,13}. However, an advanced or a complete AV block is rare and pathological studies on the conduction system of 8 cases⁴⁻¹⁰ have been reported. As indicated in Table I, the conduction systems of 6 cases were involved by rheumatoid granuloma⁴⁻⁸ or scarring tissue,⁹ one case showed rheumatoid arteritis¹⁰ and remaining one case secondary amyloidosis⁸. In the present case it is clearly shown that the AV conduction system was involved by rheumatoid nodules and scarring tissue. According to the previous reports^{5,7,9,14,15} on patients with heart blocks, rheumatoid nodules appear most commonly in the summit of the interventricular septum. Because of their location, they may involve in the branching portion of the His bundle and right and left bundle branches, producing the bundle branch block, the axis deviation and the mild-degree AV block which are occasionally associated with rheumatoid heart disease.^{3,13} A complete AV block appears to be caused by the involvement in almost all portions of the AV conduction system by rheumatoid nodules or scarring tissue, as shown by Harris⁶, Thery et al⁸ and Lev et al⁹, and by present report, as well. Although the previous reports^{6,8,9} did not mention the main sites of lesions responsible for the complete AV block, the present case suggests a possibility that the involvement in the penetrating portion of the His bundle induces the advanced or complete AV block.

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