Idiopathic Acute Myocarditis with Complete Atrioventricular Block in a Baby

Clinicopathological Study of the Atrioventricular Conduction System

Takako FUJWARA, M.D., Yuichi AKIYAMA, M.D.,***
Hiroyasu NARITA, M.D.,*** Tadashi UEDA, M.D.,
Tadashi HAYASHIDERA, M.D., Haruki MIKAWA, M.D.,
Hisayoshi FUJWARA, M.D.,* and Yoshihiro HAMASHIMA, M.D.**

SUMMARY

A 17-month-old Japanese girl with an idiopathic acute myocarditis had symptoms of vomiting, slight fever, and liver enlargement, but no edema. Clinical diagnosis of acute myocarditis was not made until she had Stokes-Adams syndrome and electrocardiogram revealed complete atrioventricular block on the day of death. At autopsy, idiopathic acute myocarditis was detected diffusely in the right and left ventricles. Vomiting and liver enlargement were due to congestive heart failure.

Serial sections of the atrioventricular conduction system revealed diffuse and severe acute inflammatory changes in the right bundle and the left bundle branches, especially in the terminal portions. Acute inflammation was focally noted in the atrioventricular node and the His bundle. The complete atrioventricular block probably followed the severe acute inflammation of the bundle branches.

Our case suggests that idiopathic acute myocarditis may be underdiagnosed in babies, as there is no way to determine whether there is dyspnea and palpitation on exertion, and idiopathic fibrosis of conduction system with or without conduction disturbances in children and adults may be sequelae of healed myocarditis in babies.

Additional Indexing Words:
Pathology of conduction system Bundle branch Cardiac fibrosis
pacing is usually carried out. Clinicopathological studies of the conduction system in cases of acute myocarditis have been done in patients with diphtheria,\textsuperscript{1)} acute rheumatic fever,\textsuperscript{2)} and Kawasaki disease\textsuperscript{3),4)} (mucocutaneous lymph node syndrome). Such studies are rare in cases of idiopathic acute myocarditis.\textsuperscript{5)–9)} As far as we are aware, study on the atrioventricular conduction system has not been reported in a baby with idiopathic acute myocarditis.

We report herein the association of complete atrioventricular block in a 17-month-old Japanese girl with idiopathic acute myocarditis, as supported by histological evidence related to atrioventricular block.

**Case Report**

A 17-month-old Japanese girl had been completely healthy until 4 days prior to her death, when she suddenly developed an anorexia and vomiting. She was

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{Fig1.png}
\caption{Electrocardiogram (lead II) on the day of death. Note the complete atrioventricular block with sinus tachycardia. \(\ddagger\) = P wave.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{Fig2.png}
\caption{Idiopathic acute myocarditis in the left ventricle. Note severe and diffuse mononuclear cell infiltration, edema and necrosis of the muscle fibers.}
\end{figure}
given 5% glucose parenterally and a diagnosis of cyclic vomiting was made. The
liver was palpable 3 cm below the right costal margin. But the cardiac arrhythmia
was not detected at the hospital to which she had been admitted. On the pre-
vious day, transient cyanosis was noted. On the day of death, there was cyanosis,
transient unconsciousness and tonic convulsion with the duration of about 30 sec
and she was admitted to Hikone City Hospital. She had slight fever and the lymph
nodes were not remarkable. The lungs had no rales. The heart rate was irregular
at 40–60 beats/min. The abdomen was distended and the liver was palpable 6 cm
below the right costal margin. There was no peripheral edema. An electrocar-
diogram revealed complete atrioventricular block and sinus tachycardia (Fig. 1).
Hemoglobin level was 11.8 Gm/100 ml and hematocrit value was 39%. The white
blood cell counts were 22,500/cumm with 43% polymorphonuclear leukocytes, 52%
lymphocytes, 5% monocytes and 1/100 erythrocytes. The erythrocyte sedimenta-
tion rate was 1 mm/hr. SGOT level was 220 IU; SGPT, 340 IU; lactic dehydro-
genase, 1700 IU; alkaline phosphatase, 21 IU; and leucine aminopeptidase, 389

Fig. 3. Atrioventricular node and His bundle.
a: Atrioventricular node (→). CFB=central fibrous body (hematoxy-
lin-eosin stain, ×40).
b: High magnification of the box area in a (hematoxylin-eosin, ×200).
c: The His bundle (hematoxylin-eosin stain, ×40).
d: High magnification of the box area in c (hematoxylin-eosin, ×200).
Note edema, necrosis and mononuclear cell infiltration in b and d. In the
atrioventricular node and His bundle, such inflammatory changes were focal.
IU. Serum electrolyte values were normal. Urinalysis revealed moderate proteinuria. Three hours after admission, she died suddenly following Stokes-Adams syndrome. Cardiac pacing was not done. Autopsy was done 1.5 hours after death.

General: Congestion was noted in the lungs (l; 60 Gm, r; 95 Gm), the liver (345 Gm) and the gastrointestinal tract. Pleural effusion (r; 70 ml, l; 70 ml), pericardial effusion (20 ml) and ascites were detected. Acute inflammatory changes were evident in the lungs and liver, in which there was evident diffuse mononuclear cell infiltration. Intensive microscopic examination of kidneys, spleen, lungs and heart revealed no microthrombi in the microvessels.

Heart: The heart weighed 65 Gm. The right atrium and the left ventricle were moderately dilated. Microscopic examination of the myocardium revealed severe and diffuse acute interstitial myocarditis, comprising chiefly mononuclear cell infiltration, edema, degeneration and necrosis of the myocardial cells, in both atria and both ventricles (Fig. 2). Acute endocarditis was focally noted.

Microscopic examination of the atrioventricular conduction system

Method: Tissue blocks containing the atrioventricular conduction system from the heart were removed and serially sectioned at a thickness of 5 μ according to the method of Lev et al.11) Every fifth section was stained with hematoxylin-eosin. All intervening sections were processed as deemed necessary for additional hematoxylin-eosin or other special stains. A total of 572 sections were studied.

Findings: Acute inflammatory changes comprising edema, infiltration chiefly of mononuclear cells, degeneration and necrosis of the conduction cells were focally noted in the atrioventricular node and His bundle (penetrating portion and branching portion) (Fig. 3). In the left and right bundle branches, severe acute inflammation, comprising infiltration chiefly of mononuclear cells, edema and necrosis were diffusely observed (Fig. 4). The most extensive changes were noted in the distal portion of both bundles. Such acute inflammation was continuous from the work-
ing muscle of the septum to the bundle branches. Inflammatory changes were nil in the mitral and tricuspid valves. There was no evidence of Aschoff bodies, fibrosis or granulomatous changes.

**DISCUSSION**

As far as we are aware, the present case is the first report on the findings of the atrioventricular conduction system in babies with idiopathic acute myocarditis. It is most difficult to differentiate acute myocarditis from common infectious disease with fever and/or vomiting in babies, as there is no way to determine whether there is dyspnea and palpitation on exertion. In the present case, the patient was vomiting and the liver was enlarged as a sequelae of congestive heart failure. But a diagnosis of acute myocarditis was not clinically made, until Stokes-Adams syndrome occurred on the day of death and electrocardiogram revealed complete atrioventricular block. The result suggests that many babies with an idiopathic acute myocarditis go over looked.

It is well known that idiopathic acute myocarditis frequently involves the atrioventricular conduction system and various conduction disturbances are seen in the electrocardiograms. In our patient, idiopathic acute myocarditis had invaded the atrioventricular conduction system. The most extensive changes were noted in the right and left bundle branches, especially in the peripheral portions. Complete atrioventricular block probably followed the severe acute inflammation of the bundle branches.

It has been detailed that idiopathic fibrosis of conduction system with or without conduction disturbances may result from a healed myocarditis.7) But the acute phase is actually unknown in most patients. Our data suggest that the transient acute myocarditis seen in babies may be one of the causes of idiopathic fibrosis of the conduction system in children and adults.

**REFERENCES**