A Case of Primary Rhabdomyosarcoma, Replacing the Atrioventricular Node

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SUMMARY

A case of primary rhabdomyosarcoma is reported with various electrocardiographic findings during clinical course. At autopsy an atrioventricular node was found to be replaced by tumor tissue which was diagnosed histologically as rhabdomyosarcoma of the pleomorphic type.

Additional Indexing Words:
Rhabdomyosarcoma of pleomorphic type Atrioventricular node Arrhythmia

PRIMARY tumors of the heart are very rare, and the incidence of these tumors found on autopsy is said to be about 0.017%.1 About three-fourths of primary cardiac tumors are benign tumors, such as myxoma.2 According to Prichard,3 the incidence of the cardiac rhabdomyosarcoma is lower than that of angiosarcoma, and nearly the same or slightly higher than that of other fibrosarcoma, reticulum cell sarcoma, and lymphosarcoma. We experienced an extremely rare case of rhabdomyosarcoma, which replaced the atrioventricular (A-V) node completely, and presented various kinds of arrhythmias. This is the eldest case of rhabdomyosarcoma.

CASE REPORT

An 81-year-old Japanese male was in good health before the gradual onset of mild productive cough in the beginning of 1968. Somewhat yellow sputa were excreted but neither mixed blood nor showed rusty color. Essential hypertension and supraventricular extrasystole were noted. In February 1973, slight pleural effusion on the right side was discovered without any other abnormal shadow. Blood count, hemogram, and the routine chemical laboratory data were within normal limits except the slightly higher level of lactate dehydrogenase (LDH).

The pleural fluid drawn at that time was not grossly bloody, but contained a large number of tumor cells. The grade of malignancy of the tumor cells was class IV, i.e. strongly suggestive of malignancy.

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The patient was transferred to our clinic on March 28, 1973. In his family there had been no case of heart disease or sudden death. The patient had not experienced rheumatic fever, chorea, or trauma on the precordium. Pertinent findings on admission included typical barrel chest, marked kyphosis, diffuse moist rales over the bilateral precordium and sharply demarcated erythematous skin lesions on both of the upper and lower limbs, which were later disclosed as psoriasis vulgaris by the skin biopsy. Slight systolic hypertension (172/52 mmHg), mild tachypnea and slight stridor were noted, but any other abnormal findings were not found, such as cyanosis, clubbing of the fingers, pulsus paradoxus, Kussmaul’s sign, engorgement of the superficial veins, pretibial pitting edema, ascites, abnormal mass at the abdomen or the rectum, and superficial lymphadenopathy. Neurological findings were within normal limits.

There were no abnormal findings of the heart except a slightly accentuated aortic second sound. No murmur was heard throughout the clinical course. Chest X-ray films showed a moderate cardiomegaly with right pleural effusion and a slight lymphadenopathy of the right pulmonary hilus (Fig. 1). Tomography showed no abnormal findings except the above findings. Sinus arrhythmia with occasional supraventricular extrasystole was found on the electrocardiogram (ECG), which is shown in Fig. 2. Laboratory findings included the following data: mild normochromic and normocytic anemia; serum total protein, 5.52 Gm/100 ml; LDH, 544 units; alkaline phosphatase (AL-P), 13.4 K.A. units; normal transaminase level (SGOT, 19 units; SGPT, 10 units); erythrocyte sedimentation rate, 22 mm in 1 hour.

Clear fluid was aspirated 4 times from the right chest by thoracocentesis. Each histological examination of the fluid revealed highly malignant tumor cells of class IV. The chemical laboratory data of the pleural fluid showed the following

Fig. 1. Chest roentgenogram on admission. Note moderate cardiomegaly and pulmonary effusion in the right lung.
values: total protein, 3.97 Gm/100 ml; urea nitrogen, 12.8 mg/100 ml; AL-P, 3.2 K.A. units; and LDH, 1,545 units. An antineoplastic agent (Mitomycin C) was administered into the pleural space, but the extent of the effusion remained unchanged for a few days.

Nine days after the admission, the heart rate abruptly fixed at 48 per minute, and an ECG showed the pattern of complete A-V block with junctional escape rhythm (Fig. 2). But the patient showed no sign of Stokes-Adams syndrome. In the night of April 26, the combined symptoms of apnea, cyanosis, convulsion, and abrupt fall of the blood pressure appeared. Ventricular fibrillation was observed at that time, and cardiopulmonary resuscitation was performed successfully. ECG following this attack revealed complete A-V block with frequent bigeminy from 2 foci in the ventricles (Fig. 2). Procaine amide was given twice and the bigeminy ceased. Next day the patient's dyspnea increased so severely that the pleural fluid was drawn by 1,400 ml. It was not bloody. The activity of LDH returned to normal level (465 units). Though all of the pleural fluid could not be drawn, the fluid began to decrease spontaneously, and only small amount of the fluid was seen several days later (Fig. 3).

Fig. 2. Repetitive electrocardiograms. The first row, sinus arrhythmia and the first degree of A-V block (PQ, 0.22 sec) with normal ventricular conduction. Second row, complete A-V block with junctional escape rhythm. Third row, bigeminy from 2 foci in the ventricle with complete A-V block. From the fourth to eighth row, atrial fibrillation, flutter, and flutter-fibrillation were shown in the supraventricular conduction, and right bundle branch block with transient normal conduction were observed in the intraventricular conduction.

Fig. 3. Chest roentgenogram after the administration of mitomycin C. Note the marked decrease of pleural effusion.
The patient remained relatively free of symptoms for 26 days. But the pattern of ECG changed markedly. In auricular mechanism, abnormal conductions were shown, such as atrial fibrillation or flutter-fibrillation. QRS complex changed from the normal conduction pattern to complete right bundle branch block, and the normal conduction was seen transiently (Fig. 2). On May 25, epigastralgia and slight melena appeared, and then he died.

*The autopsy findings: (TP: 27246)*

In the thorax the pericardium exhibited diffuse fibrino-fibrous adhesion with no effusion. There was no remarkable change in the position, size and shape of the heart except the rough surfaced epicardium. The focal slight nodular protrusion with some increase in consistency was seen at the left atrial wall. After dissection of the heart out of the pericardial cavity, a 10.5×8.0×4.5 cm sized tumor was found at the left atrial wall of the non-septal part. The cut surface of the tumor exhibited milky-white color with a map-like necrosis and was rather well circumscribed between the endocardium and the epicardium except the focal invasive appearance, adjacent to the left ventricular myocardium (Fig. 4).

Also the left circumflex coronary artery and coronary sinus were invaded by the tumor, which involved the wall of the latter with a 2.2×1.8×1.5 cm sized polypoid tumor thrombus formation in the right atrium. The invasion of the atrial septum was also noticed after serial cuts of the atrial septum, where continuous invasion occupied the head of the A-V node. The head of the A-V node was completely replaced by the tumor tissue. But the more peripheral conduction system

![Fig. 4](image-url)
Fig. 5. Involvement of the atrial septum and pars membranacea. The ventricular septum was free from the tumor, but relatively recent myocardial infarction scar was observed.

was not invaded by the tumor. The bundle of His remained intact (Fig. 5). All of the valves showed no remarkable change and the narrowing of the cardiac chambers was not considered to be present. No metastasis was found. Poorly aerated lungs (left, 650 Gm; right, 485 Gm) with focal fibrino-fibrous pleural adhesions at the left segment-3, -10, and the right segment-2 were noticed and slightly

Fig. 6. The tumor consisted of various type of pleomorphic cells. Hematoxylin-eosin stain, $\times75$. 
bloody pleural fluid (left, 480 ml; right, 50 ml) was present without any evidence of malignancy.

Microscopic findings:
The tumor consisted of diffuse proliferation of pleomorphic cells, of which cytoplasm were round or polyhedral and deeply eosinophilic. The nuclei had dense chromatin and 1 or 2 nucleoli. Numerous mitoses and bizarre multinucleated giant cells were scattered in many parts. Each of tumor cells proliferated in dense arrangement or in alveolar pattern with close connection to fine network of argentaffin fibers (Figs. 6–8). Cross striation was not shown by phosphotungstic acid
hematoxylin stain and electron microscopy. Parietal pleura adjacent to the epicardium showed non-specific inflammation without evidence of tumor infiltration. The final pathological diagnosis was rhabdomyosarcoma of pleomorphic type.

Discussion

Whorton collected 9 cases of cardiac rhabdomyosarcoma, which had been reported between 1909 and 1947. Twenty-one cases were added by Porter et al. In 1967 Dell'Acqua added 11 more cases. Saint-Florent et al described 5 more cases, chiefly from French report in 1969. The present case revealed the replacement of the A-V node by the rhabdomyosarcoma proved on autopsy; the similar case had only once been reported by Porter et al.

The ECG changes in cases of rhabdomyosarcoma, were described in 21 cases. They are summarized in Table I. All kinds of arrhythmias were shown. No specific changes of ECG were found in rhabdomyosarcoma, but cardiac tumor may be suspected when the various changes of ECG are found without the history of rheumatic fever or coronary sclerosis.

As to histological findings, cross striation had been considered to be essential in the diagnosis of the rhabdomyosarcoma. Phosphotungstic acid stain has been recommended to show the cross striation, but the presence of the cross striation is now considered not to be essential. According to Horn's classification, cross striations were not shown in 7 out of 39 cases in generalized rhabdomyosarcoma. Of the 26 reports of the rhabdomyosarcoma of the heart, cross striation was clearly seen in 16 cases, almost invisible in 6 cases and invisible in 4 cases, to which our case belonged. Some of the round cell or spindle cell sarcoma cases in the old literatures might be included in the category of the rhabdomyosarcoma. Stains using an immunological method would be more beneficial.

Various histological findings were reported in the cases of the tumors which invaded atrioventricular node. These tumors included angioma, lipoma,
mesothelioma, and rhabdomyosarcoma.\textsuperscript{5,15-23} In Porter's case the thoracotomy was performed 3 times to implant the electrode of pacemaker, and the biopsy of the heart muscle was performed. The diagnosis during life of their patient was myocarditis. Systemic lymphadenopathy was present at the late stage, and multiple metastases to the various organs were revealed in that case. In both their case and present one, hyperemia and edema of the lungs, multiple adhesions of the pleura, and bilateral hydrothorax were encountered on autopsy. The ECG of both cases demonstrated the same pattern of complete A-V block with idioventricular rhythm, but the course of ECG was far more complex in the present case than their case. Abnormal auricular conduction was the prominent finding of ECG of our case. To authors' knowledge, this is the report of the eldest case of rhabdomyosarcoma.

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