Sudden Nocturnal Death in Young Males from Ventricular Flutter

Masami Hayashi, M.D., Minoru Murata, M.D., Masahito Satoh, M.D., Yoshifusa Aizawa, M.D., Eiji Oda, M.D., Yuji Oda, M.D.,* Tohru Watanabe, M.D.,* and Akira Shibata, M.D.

SUMMARY

Two young males who had no organic heart disease died unexpectedly during the night. In one patient, the monitor ECG showed a sinoatrial block and the electrophysiologic study revealed a sinoatrial conduction time at the upper limit of normal and a prolonged PA interval. The surface ECG showed left axis deviation. Ventricular ectopic beats were confined to the night when he died from ventricular flutter. The ECG of the other patient was normal except for the change in QRS configuration found when the preceding RR interval was prolonged, suggesting a phase 4 block in intraventricular conduction. He was completely free from arrhythmia except for the ventricular ectopic beats which developed around 9:00 p.m. and the ventricular flutter following an R-on-T type ventricular ectopic beat which resulted in death during the night. The autopsy showed no organic heart disease. ST elevation suggestive of acute ischemia was not found and the QT intervals were normal in both cases. No electrolyte imbalances were found. These 2 cases can be diagnosed as Pokkuri disease which is well known in Japan. The victims are exclusively young males who have no apparent diseases to which death can be attributed.

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SUDDEN cardiac deaths have been reported during ambulatory electrocardiographic monitoring in Western countries and it was found that the majority were due to ventricular flutter and fibrillation.1)–7) The victims

From the First Department of Internal Medicine, Niigata University School of Medicine and *Minami Hospital, Internal Medicine, Niigata, Japan.
Address for reprint: Yoshifusa Aizawa, M.D., First Department of Internal Medicine, Niigata University School of Medicine, Asahimachi 1, Niigata 951, Japan.
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had coronary artery disease and were aged in the 5th decade or over except for one 21 year old patient with congestive cardiomyopathy. However, in Japan, young males have been known to collapse and die suddenly at night although they have lived a normal life prior to this. This is called Pokkuri disease as "Pokkuri" means a sudden and unexpected occurrence in colloquial Japanese. However, the details of such acute death are entirely unknown.

Recently, we happened to monitor 2 young males who died during the night. In both cases, the deaths were due to ventricular flutter following R-on-T ventricular ectopic beats.

**Case Report**

**Case 1**

The patient was a 25 year old male. He had been quite well until the night when his parents noted a noise in the bedroom and found that he was unconscious. Two min later, he regained consciousness but the same thing happened again 1 hour later. He was hospitalized on the next day. The physical examination disclosed no abnormality except for an irregular pulse. Chest X-ray showed no cardiomegaly or pulmonary congestion. The other noninvasive data, including two-dimensional echocardiography were non-contributory. The ECG showed atrial fibrillation (Fig. 1, left). Sinus rhythm returned in a few days (Fig. 1, right), but long sinus pauses followed by junctional escape beats were found frequently as shown in Fig. 2 (upper).

**ECG Case 1 T.Y.**

![ECG on admission in Case 1(left). Normal sinus rhythm with left axis deviation (right).]
He was paced for the sinus pauses for several days until normal sinus rhythm was observed regularly. The mean frontal axis was $-75^\circ$. No prolonged QT interval or abnormal Q wave was found. Blood pressure was 112/60 mmHg. Blood chemistries, including serum electrolytes (Na=140, K=3.8, Ca=5.4 mEq/L, Mg=2.4 mg/100 ml) were normal. No inflammatory sign was found.

The electrophysiological study was done in the post-absorptive state. The corrected sinus node recovery time was 540 msec and the sinoatrial conduction time (Strauss' method) was 150 msec. PA interval was prolonged to 70 msec. AH and HV intervals were both normal, 100 msec and 50 msec, respectively. The effective refractory periods of the atrium and AV node were 250 msec and 380 msec, respectively. He had been under

![Monitor ECG](image)

Fig. 2. The monitored ECG. A long sinus pause was followed by a junctional escape rhythm in Case 1 (upper). ECG from Case 2. A notch is apparent as indicated by an arrow (lower).

![Monitor ECG](image)

Fig. 3. The terminal ECG of Case 1. Although sporadic ventricular ectopic beats first appeared at 1:47 a.m., no warning arrhythmia was found even 10 min before death (upper). Following polymorphous ventricular tachycardia, unimorphous ventricular tachycardia developed which soon degenerated to fibrillation (lower).
continuous monitoring after admission. On the 10th day, transient polymorphous ventricular tachycardia was found at 3:00 a.m. He lost consciousness for a brief time. No warning premature beat was found before the episode. He was then completely free from arrhythmia until the 16th night when ventricular ectopic beats appeared spadically during the night (beginning at 1:47 a.m.) followed by polymorphous ventricular tachycardia at 2:12 a.m. (Fig. 3). The latter resulted in death. Cardiopulmonary resuscitation was tried but failed.

Case 2

The case was a 32 year old male whose past history was noncontributory. He noted an attack of dizziness during treatment with antibiotics for a sore throat which had developed 2 weeks before admission. On the day of admission, he complained of dizziness, cold sweats and palpitations. The physical examination disclosed an irregular pulse but otherwise was noncontributory. Chest X-ray was normal. Blood count showed mild leukocytosis (WBC=10,500 cmmm) and mild liver dysfunction was found. Serum electrolytes were normal (Na=142, K=4.0, Ca=5.0 mEq/L). The leukocytosis and the abnormal liver function normalized a few days after admission.

ECG on admission showed atrial fibrillation (Fig. 4, left). QRS complex was normal and no abnormal Q wave or ST-T change was found. QT interval was normal. When the preceding RR interval was prolonged, a notch was found on the QRS complex as shown in Fig. 2 (lower). The longer the preceding RR interval, the clearer the notch became. The atrial fibrillation was treated with quinidine sulfate given orally and normal sinus rhythm returned (Fig. 4, right). The drug was then changed to verapamil.
Case 2 S.T. Holter ECG
0:58 a.m.

1:07 a.m.

1:10 a.m.

Fig. 5. The terminal ECG in Case 2. Following an R-on-T ectopic beat, polymorphous ventricular tachycardia or flutter appeared.

(120 mg/day) to prevent possible APCs and the patient was discharged on the 12th day.

In the outpatient clinic, he was noted to have a Wenckebach type A-V block and verapamil was discontinued at that time. Otherwise, the ECG was the same as previously. He went home with a Holter monitoring recorder. On the next morning, he was found out to be dead in the bedroom. The Holter ECG showed the terminal event. One ventricular ectopic beat developed at 9:47 p.m., 6 at 11:10 p.m. and 17 around 0:42 a.m. Following an R-on-T type ventricular ectopic beat at 1:07 a.m., ventricular fibrillation developed as shown in Fig. 5. No ST-T change was apparent. This ventricular arrhythmia was thought to be the cause of his death. An autopsy was done but no organic heart disease was found. The histological study revealed a normal conducting system. The coronary arteries were normal.

DISCUSSION

In Japan, young males without apparent disease are known to die suddenly and unexpectedly during the night.8)–10) The cause of death is controversial but is generally believed to be cardiac in origin. A histopathologic study showed fibrosis and a significant reduction of the conducting fibers in and around the sinus node in 6 of 7 patients.8) Abnormal course and branching of the node artery was seen in 6 of 7 patients. Four cases had fibrosis or lipomatous interruption of the His bundle or bundle branch.9) Similar
findings of fibrosis in the sinus node region with minor anomalies of the sinus node artery have been seen by other workers. However, how these minor abnormalities result in the death of these young men is entirely unknown.

The present cases seem to be examples of " Pokkuri disease " in which apparently healthy young Japanese males suffer sudden unexpected nocturnal death. Electrocardiographic monitoring showed that both cases experienced ventricular flutter and fibrillation following R-on-T ectopic ventricular beats immediately prior to death. It is to be stressed that the warning arrhythmia was confined to within 1 or 2 hours before death. The deaths were therefore completely unexpected. Both patients showed atrial fibrillation when they were admitted following attacks of dizziness or unconsciousness at home, possibly from cardiac arrest. Case 1 had slightly abnormal sinus nodal function but normal A-V nodal function. His PA interval was prolonged and his atrium might have been electrophysiologically abnormal, but there was no direct relation between these abnormalities and the death. In Case 2, phase 4 block below the His bundle was suggested because of the change in QRS configuration which became clearer when the preceding RR interval was more prolonged. The autopsy was, however, non-contributory. QT intervals were normal in both cases. Acute ischemia as reported by Savage et al was unlikely because there were no ST-T changes during ECG monitoring.

This is the first report where sudden nocturnal unexpected death in young males was actually demonstrated to have been precipitated by ventricular flutter and fibrillation. We conclude that death in so-called " Pokkuri disease " in Japan is most probably cardiac in nature. More aggressive electrophysiologic studies as well as screening studies need to be established to deal more effectively with this disease.

References

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