ACUTE CARDIAC DEATH OF UNKNOWN ETIOLOGY IN JAPAN

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Since the medical examiner's system was established in Tokyo in 1948, almost a hundred thousand cases were examined and about fourty thousand cases were autopsied during the past 21 years up to 1970. Annual number was shown in slide.

Around 60% out of the autopsied cases were determined as sudden and unexpected natural deaths.

Therefore our examiner office have experienced a large number of cases, more than 1000 of sudden death every year.

The cardiac diseases were most frequent. The percentage was 50 to 60% from the examination of sudden and unexpected death. This is nearly identical with reports made by other investigators both in Japan and other countries.

As to incidences of the types of heart diseases in acute cardiac death, coronary sclerosis was most frequent and nearly 50%, as similar to that in other countries, while the disease of myocardium 15 to 20%, the disease of heart valve and endocardium 15% and disease of aorta or pulmonary artery 7% respectively.

OUTLINE OF ACUTE CARDIAC DEATH OF UNKNOWN ETIOLOGY

It has been a great concern to us that there are curious death, in which we could not find any evidence of remarkable change in all organs to cause acute death. This is found in almost 100 cases every year, which is about 15% of the total number of acute cardiac death.

Now, we would like to focus our remarks upon the death of this type, acute cardiac death of unknown etiology in more detail. It is called "Pokkuri disease" in Japanese language.

The majority of these deaths occurred almost unexclusively in young male individuals who seemed apparently healthy and lived their lives as usual until the day before death. In general they died suddenly during sleep with a groan as if having a dreadful dream and, agonal deep respiration with stretching of limbs were often noticed.

(a) EPIDEMIOLOGICAL ANALYSIS

A large scale of investigations has been made upon 525 cases in 2 separate periods, one from 1959 to 1961 and another from 1969 to 1970. Now we are going to mention several items upon the clinical and pathophysiological features of this death in more detail from these investigations.

1. Hereditary disposition was not so dominant. However, in more than 6 family trees, 2 cases of this death were observed in the same family. One example is presented in slide as a case report.
2. As to the annual incidence, almost 100 cases, 15 to 20% out of total acute cardiac death were seen annually in the Tokyo area. This percentage was similar to that in Osaka area, by Matsukura's report.
3. In general, the physical condition of these cases was apparently healthy and they worked as usual until the day of death, except about 1/3 cases had complained of easily becoming fatigued before death.
4. The age distribution is shown in the next slide. It ranged from 13 to 48 years old and almost 60% of the cases were around 20 years old. It contrasts to that of sudden death due to coronary heart disease, the age group were mainly over 40 years old.
5. As to the time of death, it was interesting that most cases died in the night time, especially between 12 p.m. to 6 a.m. as compared to that the sudden death from any other organic heart diseases occurred mostly in the day time.
6. As to monthly incidence, the occurrence of these deaths was most frequent in the summer time between May and September; while the organic heart disease in the winter season.
7. The sex incidence dominated mostly in male and the ratio of male to female about 15:1. Furthermore, it was interesting that the state of death was much different between the male and the female. For example, the majority of male died at night during sleep (84%), while this was less than 10% in the female. Other example was that most of the males had no complaint but females had some complaints or illness, such as pregnancy, common cold etc. before death.
8. The objective symptoms observed by the people presented at the moment (spot) are shown in slide. According to frequency, groan 67%, cramp 23%, abnormal respiration 18% and so on.
9. On the other hand, a few cases complained or screamed their subjective symptoms just before death. The most frequent symptoms were chest pain like anginal attack.

(b) LIFE & PHYSICAL SITUATION JUST BEFORE DEATH:

10. The trigger of cardiac attack and the immediate pre-death situation such as physical condition and life situation were investigated as much in detail as possible during the subject's final few minutes, final 24 hours and final few days. These results were compared between cardiac deaths of unknown etiology and those due to coronary disease. In the group of unknown etiology, any special events considered to be a trigger was not recognized because of the subject being asleep in the majority. It was interesting that in most of the coronary group, the attack leading to death was provocated by some moments such as muscle work, exercise, taking a bath, walking or taking a meal.

Most of the subjects without organic disease did not complain of any physical trouble before death, except easily becoming fatigued. To know the reason for this fatigue, the life situation was examined in some cases of unknown etiology. The frequency of life stress were as follows:

1. The change of their life pattern such as moving.
2. Insufficient sleep for some reason.
3. Trouble at home.
3. Emotional tension due to their business.
5. Physical trouble like pregnancy, common cold, loss of appetite, neurosis.

It was suspected that many of the subjects of unknown etiology were exhausted from several reasons before the death.

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SOME ASPECTS OF PATHOLOGICAL FINDINGS

1. Coronary artery: Any abnormality to cause the death in the coronary artery was not recognized. Both the main arteries and small branches were examined completely in some cases. Especially the small branches were followed grossly as far as possible.

One example of coronary artery of 28 years male, examined by Sugai, one member of our office is presented.

In the next place the distribution type of the coronary artery was classified by means of dissection method and plastic casting method. The type, in which the right coronary artery predominating the left were observed in a high percentage, as compared with the control group.

2. Heart and aorta

Slight under-developed heart and aorta were recognized in some cases.

In the microscopic studies of the myocardium, the general findings observed were slight pathological changes as a consequence of oxygen deficiency of the cardiac muscle. However. these minor changes did not seem to cause the death.

3. The interesting pathological changes of several organs other than heart were as follows.

In $\frac{1}{4}$ of the subjects, the hypertrophy of thymus was seen.

The microscopic studies were done on the automatic ganglion and adrenal gland in some of the subjects at random. A considerable degree of atrophy and degeneration of ganglion were seen and the thinning, the scantiness of lipid in adrenal cortex and the decrease of weight of adrenal gland were seen in the majority.

SOME CONSIDERATIONS ON THE MECHANISM OF THIS DEATH

In spite of many efforts made by several investigators both morphologically and biochemically, the mechanism is still obscure at this moment.

We have also done several biochemical analyses previously such as the estimation of some enzymes, minerals, Vitamins and so on. But no information were obtained.

In order to clarify the real mechanism of this death, from our experiences, we believe that the most important thing would be understanding the mechanism of cardiac arrest in any kind of sudden death. This might be the final answer for the mechanism of sudden death without organic disease mentioned in this session, because even in sudden death due to coronary disease, nobody knows the detail mechanism of the stop of the heart beat at the moment of their death.

From all information analysed of all types of sudden death, on their state of death, pre-death situation and pathophysiological features, the total sudden death might be divided into two groups: namely death during sleep and during awake. These observation might be points to be investigated in the future. The next step to be studied in general is the final analysis upon (1) cardiac conduction system and (2) neurohumoral system in relation to sudden death.