CLINICOPATHOLOGIC CORRELATIONS

De Subitaneis Mortibus

I. Fibroma Compressing His Bundle

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SUMMARY

Necropsy studies of sudden death should always include examination of the conduction system of the heart but seldom do. This case is an example of sudden unexpected death in which there was a fibroma compressing the His bundle. Almost by definition it will be necessary to reconstruct probable clinical events retrospectively in such cases and several such considerations are made here. By studying individual examples and appropriate groups of cases in this way, it should be possible to reduce the number of “unexplained” cases, to formulate reasonable hypotheses for prospective examination, and ultimately to devise means of preventing some of these deaths.

Additional Indexing Words:  
Sudden death  Electrical stability of the heart

SUDDEN DEATH is a very old subject.¹ Being struck dead or found dead unexpectedly is a recurring theme in epics, operas and legends. In many respects the associated mystery is the most compelling feature of the subject. Those sudden deaths which were expected or which are readily explained seldom become epic themes. But whatever charm this subject may hold for poets and playwrights, it is a vexing burden for physicians.

From careful necropsies and the increasingly sophisticated analyses now available to forensic pathologists, the number of previously mysterious deaths is gradually diminishing. However, despite the fact that many mysterious deaths are logically attributable to lethal electrical instability of the heart, the cardiac conduction system is too seldom included in forensic pathologic studies. This conspicuous omission recently led Schwartz and Walsh² to recommend that examination of the conduction system and its blood supply become routine in every necropsy study of sudden death, and that those studies without such an examination must be considered as significantly incomplete.

For nearly ten years we have conducted a continuing series of studies of the electrically specialized tissues of the heart from examples of sudden death in which neither the clinical history nor a careful routine necropsy provided a suitable explanation, and in which the known circumstances just before death suggested that the terminal event was lethal electrical instability of the heart. Although we are keenly aware of some limitations inherent in such studies, such as the absence of any specific knowledge concerning cardiac rhythm or conduction, there is no presently suitable means to circumvent these limitations when the subject is a study of sudden unexpected death. One may also invite certain restrictive definitions, such as what is meant by “sudden.” We take the colloquial definition, meaning an unexpected or unusual death which was sudden in general terms, and which may or may not have been witnessed, but which poses a mystery for explanation even after careful routine necropsy. Specific events concerned with a particular case can regularly be provided in detail as known.

Having the available facts concerning such deaths, and necropsy material to examine, we
focused our attention on the sinus node, atrioventricular (A-V) node, His bundle and its proximal branches in every case. The studies routinely included careful assessment of the blood supply to these structures. From this accumulated and continuing experience, certain examples lend themselves most suitably to individual analysis, while others are best considered in small or larger groups of cases. This initial example is best considered individually.

Available Clinical Information

A forty-year-old man was considered in excellent health except for having developed a cold one day prior to death. He was in good spirits at the time of his evening meal and went to bed normally a short time thereafter. His wife was out of the room briefly after he had retired, but he appeared to be normally asleep when she returned. At about 4 a.m. she was awakened by a gurgling noise and found her husband lying on his back in bed beside her. She shook his hand and asked him if he was all right but he did not answer. She then got out of bed and turned on the light and heard the gurgling noise again, now noticing that his mouth was purple and she thought that his tongue was swollen. She then lifted his eyelids and realized that he was dead.

He had been in hospital three years previously with stomach ulcers but had had no recent medical attention.

 Necropsy Findings

There were no marks of violence. Toxicological examination of the blood, urine, gastric contents, liver and kidney was negative. Concerning the history of a cold, there was no evidence of any complicating infection which could have caused death. Gall stones were present and a chronic duodenal ulcer as well but neither of these conditions could have played any part in the death. The lungs were congested and slightly edematous with some recent bleeding into the alveoli; this appearance was one of terminal congestion. There was no pneumonia or other disease.

The heart appeared grossly normal and many routine sections of myocardium were unremarkable. The coronary arteries, including those supplying the conduction system, were normal as were all four cardiac valves and the septa of the heart. The sinus node was normal. From the A-V node anteriorly the tissue was serially sectioned at 8 μ intervals because a small tumor was visible.

An encapsulated fibroma lay adjacent to the anterior half of the His bundle, compressing it from the left side (figs. 1 to 3). The central fibrous body and the membranous interventricular septum were thicker than normal but the fibrous tumor was discretely circumscribed, measuring about 3 by 4 mm in an elliptical shape similar to a bean, and being slightly over one millimeter in maximal thickness. There was no His bundle anterior to the tumor, the left bundle branches having completed their departure into the left septal endocardium and the rather large right bundle branch continuing on to penetrate the interventricular septum on its right side. The posterior two millimeters of the His bundle which included virtually all of the undivided portion was entirely normal, as was the A-V node. In the region where the tumor compressed the His bundle, the tumor lay directly beneath the left ventricular endocardium just below the aortic valve and displaced the His bundle to the right. Except for a thin margin of smooth muscle at its surface adjacent to the His bundle, the tumor was composed of mature collagen and elastin. It was relatively sharply demarcated from the adjacent collagen of the central fibrous body and membranous septum, although it was a component of these structures. No other similar tumors were found elsewhere in the heart.

Discussion

There are two facts in this case: the patient died suddenly and unexpectedly, and a fibroma was found compressing the His bundle. It would of course be helpful to know with certainty if any form of rhythm or conduction disturbance did precede his death, but the circumstances strongly suggest that the two facts are causally related rather than coincidental. This interpretation is supported by the numerous reports of sudden death having occurred in subjects later found to have a fibroma of the heart, particularly when the interventricular septum was involved, although we are unaware of any previous study in which the conduction system was described. There are three recent good reviews of the subject of fibroma of the heart, including lists of reported cases and the postulated causes of death.3-5

It may be useful to consider briefly some possible mechanisms of lethal electrical instability of the heart in this patient. Although there was no history
Figure 1

These two photomicrographs compare the His bundle described in the present report with a normal one from an adult of similar age (both sections prepared with Goldner trichrome stain). The left ventricular cavity (LV) is on opposite sides of the two sections due to the way they were mounted. Cavities of the right atrium (RA) and right ventricle (RV) are indicated and the tricuspid valve is seen between them. In each section the His bundle is indicated by three open arrows. The upper part of the small fibroma is indicated by three black arrows. Note that the left ventricular endocardium in this region of a normal heart is comparatively thin. See figures 2 and 3 for more details.

of symptoms suggesting previous failure of A-V conduction, the information concerning this possibility is meager and there were no prior electrocardiograms. If terminal electrical instability was the cause of death, one may ask why it occurred at the time it did. Three factors which may be contributory are the recent onset of a cold (possibly some fever or slight toxic reaction), the fact that he was asleep (when oxygen content normally diminishes in the blood, and carbon dioxide normally increases, and when frightening dreams may occur), and the likelihood that at some time in the life history of the compression of His bundle by the tumor some clinical point would eventually be reached when the His bundle malfunctioned. The tumor here was adjacent to the His bundle at an area where the bundle is normally encased in collagen which limits its mobility, and the tumor was in a location subject to the highest pressure generated by the contracting heart.
These two sections are about 2.5 mm apart and come from near the anterior and posterior margins of the tumor. Adjacent His bundle is indicated with a large open arrow. The section with Goldner tri-chrome stain (above) is anterior to the one with Verhoeff-Van Gieson elastic stain (below). The upper section is a higher magnification of that in figure 1. The small open arrows indicate the left bundle branch passing between the fibroma and the crest of the interventricular septum, and the small black arrows indicate a rim of smooth muscle at the margin of the tumor adjacent to the His bundle. Magnification in both photomicrographs is the same.

If His bundle malfunction was represented by longitudinal dissociation of conduction and the development of a re-entrant arrhythmia, this may have been at a rate so rapid or slow or sufficiently irregular that cardiac action no longer sustained life. On the other hand, total failure of A-V conduction may have been followed by an irregular escape rhythm, or ventricular fibrillation, or cardiac standstill, any of which if sustained could have been the terminal event. The presence of pulmonary congestion and the dying gurgle which was heard suggest a brief period of pulmonary edema, but any of the several postulated electrical events may have caused this. While there is no way in such cases to know what the electrophysiological disturbance truly was, it is nevertheless instructive to consider the plausible possibilities.

Finally we may consider the etiology of this isolated small but critically placed fibroma. Most cases of fibroma of the heart have been found in children, and only very few in adults as old as the present case. While it has been suggested that the predominance in early ages is due in part to the frequency with which fibroma of the heart "causes" sudden death, it also seems likely that this abnormal behavior of cardiac connective tissue is an
early developmental abnormality. In the present case the location of the fibroma and the unusual thickness of most of the membranous septum and central fibrous body suggest that this tumor occurred at the junction of forward-moving dorsal endocardial cushion with the anlage of the roots of the great vessels. In the human fetus and newborn the central fibrous body is normally of a much greater relative thickness than it is in the adult, and the postnatal condensation of this loose connective tissue into a thinner and more compact structure is associated with a conspicuous process of molding and shaping of the His bundle in the newborn period.6,7 This being precisely the location of the present fibroma, it seems likely that both the tumor and the thick adjacent collagen represented some failure of progression of a normal process, although the presentation of clinical consequences so late in life (comparatively) is unexplained.

The important lesson from this case is that a distinct abnormality was found in the cardiac conduction system of a man dying suddenly and unexpectedly, and in whom a careful routine necropsy had provided no explanation of the death. Whatever the exact electrophysiological malfunction may have been, the evidence seems best interpreted as death being due to a terminal electrical instability of the heart. How often
significant abnormalities of the cardiac conduction system are associated with such deaths, and whether such abnormalities are similar or widely different in nature can only be known by making histopathological examination of this crucially important part of the heart an essential part of such necropsies.

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