Editorial Comment

Tumors of the cardiac conduction system: are they an explanation for otherwise unexplained sudden cardiac death?

Brian Olshansky

University of Iowa Hospitals, 200 Hawkins Drive, 4426a JCP, Iowa City, IA 52242, USA

Cardiac tumors are well described in the literature. The first reports of cardiac tumors date back hundreds of years. The prevalence of primary cardiac tumors at autopsy ranges from 0.001% to 0.3% with secondary tumors more common than in primary tumors. 1

Recently, Yu et al. from the People’s Republic of China, reported a series of 234 patients diagnosed with primary cardiac tumors from 33,108 who underwent cardiac operations between 1996 and 2005. 2 The most common benign tumor was a cardiac myxoma. Mesenchymomas and angiosarcomas were among the most common primary malignant cardiac tumors. Rhabdomyomas and fibromas were more common in children.

Tumors of the conduction system are known to be small and can increase the risk of sudden death even if the tumor is benign. 3,4 Such tumors have been well described many years ago. 5 The issue remains: how often do tumors of the cardiac conduction system occur and what is their risk? These questions remain unknown.

Song et al. in this issue, provide new provocative information regarding tumors of the cardiac conduction system based on pathological post-mortem evaluation. In their report, two groups of patients were evaluated. One group included 198 cardiac related deaths, and the second group included 838 non-cardiovascular related deaths. With careful sectioning of the heart, 12 cases (1.2%) were found to have tumors of which ten were primary and two were metastatic tumors. This is a relatively high number compared to other reports in the literature. Tumors were located in the AV node or His bundle in eight of the 10 primary tumors. The cardiac conduction system tumors all arose from tissue in the ventricular septum or from residual embryonic cells. Of the 12 cases, two individuals died suddenly presumably related to the tumor. The other patients died from other causes.

What is not completely clear is whether these findings were incidental findings or the cause for death. From a clinical standpoint, the key issue is how to apply these findings and to assess what they mean for a patient. The investigators complete a pathological morphological examination of the tumors of the conduction system but it is not clear if any patients that have these tumors the conduction system could potentially survive with transient heart block or other problems that could be corrected by a pacemaker or even a defibrillator. It is not clear the clinical impact of these tumors in those patients who died from other causes. It is not clear what would need to be done if they were or could have been identified before death (but it seems unlikely that this could occur). The clinical impact of these tumors remains unknown. Even in the two cases of sudden cardiac death, it is not clear that they are directly linked to the tumor of the conduction system.

The investigators contend that tumors of the conduction system may serve as a basis for electrical instability of the heart. The investigators have not yet proven the point that these tumors were the cause of death. Even if they caused complete heart block or bradycardia, that does not necessarily indicate that sudden death is inevitable. The patient could present with complete heart block and symptoms such that a pacemaker would reduce the risk of sudden death. On the other hand it is possible that a tumor of the conduction system may cause ventricular fibrillation that would not be prevented by a pacemaker. It is possible that conduction system tumors may have long-term arrhythmogenic affects that may not be ameliorated by a pacemaker alone. The authors mentioned “cardioneuropathy” a term coined by James et al. 6 The investigators have not shown that cardioneuropathy existed in or pertained to any of the patients in their series.

The jury is out with regard to the clinical importance of these microscopic tumors found in the cardiac conduction system. The information reported is interesting as it points out that tumors could explain otherwise idiopathic heart block or even the possibility of sudden cardiac arrest, but the investigators have not proven a relationship between the two. Others have shown this to be the case yet these findings may simply be incidental. On the other hand, these findings may be a ready explanation for individuals with otherwise apparently normal hearts.

More work needs to be undertaken to determine the mechanism and cause for sudden cardiac death in those who have no explainable cause. Most likely, some genetic abnormality, such as, long QT interval syndrome, Brugada
syndrome, catecholaminergic polymorphic ventricular tachycardia or another, as yet, poorly defined channelopathy may be the most common explanation of otherwise idiopathic sudden cardiac arrest.

Genetic screening may help identify the cause for patients who have sudden cardiac arrest and have no other explainable problems. Nevertheless, this manuscript opens up the possibility that small, otherwise unidentifiable tumors of the cardiac conduction system may be another mechanism responsible for sudden cardiac arrest in individuals who otherwise have no obvious explanation for their problem.

I congratulate the authors for addressing this issue in such a careful fashion and encourage them to focus on clinical correlation between their pathological findings and a potential mechanism by which death may occur in these individuals.

References
1. Roberts WC. Primary and secondary neoplasms of the heart. Am J Cardiol 1997;80:671-82.