H58 Primary Cardiac Tumor as a Cause of Sudden Death: A Report of a Rare Case of Atrial Lipoma

Stefano D'Errico, MD, PhD, Department of Legal Medicine, Lucca 55100, ITALY; Diana Bonuccelli, MD, Department of Legal Medicine, Lucca, ITALY; Alberto Mandoli, MD, Department of Legal Medicine, Lucca, ITALY; Massimo Martelloni, MD, Department of Legal Medicine, Lucca, ITALY; Francesca Maglietta, MD, Department of Forensic Pathology, Foggia 71122, ITALY*

Learning Overview: The goal of this presentation is to present the case of a sudden undiagnosed cardiac lipoma of the right atrium.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by discussing fatal cardiac rhythm disturbances caused by an atrial tumor infiltrating the free wall of the right atrium and the role of primary cardiac tumors as a cause of sudden death.

Primary tumors of the heart are rare, and the incidence varies between 0.0017 and 0.19% in unselected autopsy studies. Among these, more than 70% are benign, mainly myxomas. Other benign tumors are lipoma, papillary fibroelastoma, rhabdomyoma, and fibroma. It is extremely rare, with a reported incidence of approximately 10%–19% among primary tumors of the heart and pericardium. Histopathologically, cardiac lipoma can be classified into two types: lipomatous hypertrophy of the interatrial septum and true lipoma. Approximately 50% of cardiac lipomas arise subendocardially, 25% subepicardially, and 25% from the myocardium. The most common chambers involved are left ventricle and right atrium with a prevalence between 40–60 years of age without sex predominance and extremely variable in size. Patients with cardiac lipoma are usually asymptomatic and discovered incidentally; symptoms reported are fatigue, dyspnea, palpitation, syncope, and even chest pain, which are frequently a result of coronary artery or cardiac conductive system involvement. Tumors in the subepicardial region can create a mass effect on nearby structures. They can cause angina if they compress the coronary arteries, or they can reduce systolic function by compressing on the left ventricle. Tumors in the myocardium can infiltrate the electrical circuit and be a nidus for arrhythmogenesis. Depending on the chamber involved and the size of the mass, they can cause obstruction of flow and congestive heart failure.

Cases of sudden unexpected death attributed to myocardial tumors have been poorly described in forensic and clinical literature; in these cases, cardiac neoplasms cause atrioventricular or intraventricular conduction disorders, which are manifested by arrhythmias, interfering in the cardiac dynamic and leading to sudden death. It has been calculated that 0.0025% of all cardiovascular deaths may be sudden death caused by primary cardiac lesions and 0.01%–-0.005% of all sudden deaths could be due to primary cardiac tumors and 0.06% of cardiovascular deaths among the 0 to 34-year-old population may be the result of sudden death caused by a primary intracardiac tumor. These data indicate that primary cardiac lesions are uncommon, yet potentially lethal. Owing to the rarity of these lesions, it seems likely that many practicing autopsy pathologists will never encounter a primary cardiac lesion. It is also expected that several primary cardiac tumors causing sudden death will be missed each year because an autopsy is not performed.

Case Report: Presented here is the case of a 67-year-old man who was found unconscious at home and immediately taken to the emergency department of the local hospital. He had a history of hyperthyroidism and hyperlipidemia. One hour after recovery, clinical conditions improved, he was without pain and oriented (GCS 15); normal cardiac function was recorded at Electrocardiogram (ECG), and neurological examination was unremarkable. Two hours after recovery, he suddenly and unexpectedly died. A hospital autopsy was performed the day after death. External examination was unremarkable. Gross examination revealed an encapsulated tumor (40mm x 30mm x 30mm) in the external layer of the right atrium free wall. The section showed a yellowish and elastic solid mass. The heart was normal in size and shape and the coronary arteries were unremarkable. A heart examination was performed according to the inflow-outflow method; no further pathological findings were attributable to the heart except for the presence in the subendocardial wall of the right atrium of three yellowish nodular buttons 1cm in diameter. Mild pulmonary edema was also recorded, with white foam on the main bronchi. The histologic study was completed using formalin-fixed paraffin embedded tissue sectioned at 4mm and stained with hematoxylin–eosin. Microscopic findings were suggestive for a right atrial lipoma infiltrating through the atrium wall with mild intracardiac involvement. The diagnosis was confirmed by immunostaining that revealed immunopositivity for S100. Based on morphological and microscopic findings, a sudden arrhythmogenic cardiac death related to an atrial rhabdomyoma was diagnosed.

Sudden Death, Cardiac Lipoma, Fatal Arrhythmia