H82 A Case of Hamartoma of Mature Cardiac Myocytes (HMCM) Correctly Diagnosed at Autopsy

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Learning Overview: The goal of this presentation is to illustrate the clinicopathologic features of HMCM and its differentiation from other primary cardiac tumors.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by raising awareness of a rare primary cardiac tumor that may be detected or correctly diagnosed for the first time at autopsy.

Case: The decedent was a 35-year-old male with a history of cardiac fibroma clinically diagnosed in childhood and drug abuse who had an unwitnessed arrest at home with controlled dangerous substances found at the scene. He saw a cardiologist 16 months prior to death at which time he was asymptomatic; electrocardiogram showed normal sinus rhythm, left atrial enlargement, intraventricular conduction delay, and lateral T wave inversion; echocardiogram showed normal right ventricular size and function, normal left ventricular function, and a 7.1 x 5.5cm well-encapsulated echodensity on the left ventricular inferolateral-lateral wall. Autopsy and toxicology testing revealed the cause of death was fentanyl intoxication. Grossly, the decedent's heart had a 6 x 5 x 5cm, poorly-circumscribed, firm, white-gray mass involving the anterior, lateral, and posterior walls of the left ventricle. Microscopically, the mass consisted of mature hypertrophic cardiomyocytes with extensive fibromuscular disarray, areas of fibrosis and adipose tissue, and thick-walled arteries. Therefore, the mass that had been clinically diagnosed as a cardiac fibroma was actually a HMCM.

Discussion: HMCM is a rare benign primary cardiac tumor first described in 1988.[1] Between then and August 2019, there were 26 cases reported.[2-4] HMCM occurs more frequently in males (ratio 1.6:1) and affects all age groups (mean 32 years, range 6 months to 76 years).[2] HMCM most frequently occurs as a single mass in the left ventricle but may be multiple and involve the interventricular septum, right ventricle, and right atrium. Similar to other hamartomas, HMCM grows slowly and does not invade the surrounding tissue. Patients may be asymptomatic or have non-specific Electrocardiogram (ECG) findings, arrhythmia, palpitations, chest pain, dyspnea, syncope, or sudden death. In 46% of cases, the tumor was found incidentally at autopsy or during cardiac evaluation for other reasons.[2] HMCM is difficult to distinguish from other cardiac tumors such as fibroma and rhabdomyoma via imaging, and definitive diagnosis requires pathologic evaluation.[2] Grossly, HMCM is a discrete to ill-defined, white-tan, firm mass.[2,4] Microscopically, HMCM consists of disarrayed hypertrophic cardiomyocytes interposed with fibrous tissue, adipose tissue, and thick-walled arteries or dilated venules.[1-4] HMCM does not show significant inflammation, calcification, or vacuolization. In contrast, cardiac fibromas are well-circumscribed, white-tan, firm masses composed of bland fibroblasts and collagen, are often calcified, and are poorly vascularized.[5] Meanwhile, cardiac rhabdomyomas are well-circumscribed, white-gray, firm masses composed of immature cardiomyocytes with diffuse glycogen vacuolization.[6] In conclusion, HMCM is a rare benign primary cardiac tumor that may go undetected or be clinically misdiagnosed until autopsy, at which point routine histologic evaluation can provide a definitive diagnosis.

Reference(s):

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