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Clinical Case Report

Isolated fibromuscular dysplasia of the coronary ostium: a rare cause of sudden death. Case report and review of the literature

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ABSTRACT

We present a unique case of sudden death in a 21-year-old man with history of cocaine use and a solitary fibromuscular dysplastic lesion completely occluding the left coronary artery ostium. We document intimal proliferation of myofibroblasts at the opening of the left coronary ostium without other concomitant lesions. This report discusses the gross and histologic features of the lesion, explores in careful detail the possible etiologies, and gives a comprehensive literature review of isolated coronary ostial fibromuscular dysplasia presenting with sudden death.

1. Introduction

Fibromuscular dysplasia (FMD) is a nonatherosclerotic and noninflammatory arterial disease of unknown etiology, first described by Leadbetter and Burkland in 1938 [1]. It is characterized by a proliferation and disorganized arrangement of cellular and extracellular elements of an arterial wall, leading to distortion and narrowing of the vessel [2]. FMD typically affects young, white adults, between the third and fourth decades. The classification is based on the location of the lesion in the arterial wall. Pathologic classification originally described observations in patients with renal arterial disease [3]. This classification of renal FMD has not changed substantially since the original descriptions and is presently utilized in the classification of extrarenal lesions. Briefly, FMD is divided into three major types depending on the dominant arterial wall layer involved: (1) intimal fibroplasia (10%), (2) medial fibroplasia (80–90%) of which there are four subtypes (medial hyperplasia, medial fibroplasia with aneurysms, perimedial fibroplasia, and medial dissection), and (3) adventitial (periarterial) fibroplasia (<1%) [4,5].

Renal and carotid arteries are affected most often [4,6]. Coronary FMD is uncommon [7–41] and may present with sudden death [7–21], angina pectoris [42], or myocardial infarction [43–45]. Although newer classification systems based on angiographic appearance have been developed for diagnosis and treatment and are correlated with the histopathological designations [46], the original classification proposed by Harrison and McCormack [3] still remains the gold standard for coronary disease. Coronary FMD presenting as sudden death has been reported in three cardiac vascular anatomic locations: (1) major epicardial and subepicardial coronary arteries, (2) cardiac conduction system vascular supply and their branches, and (3) intramural "small vessel" arteries. Coronary FMD with involvement of the ostium is rare [13]. Two cases of isolated coronary ostial FMD have been documented [42,47]; neither presented with sudden death. We present a rare case of sudden death due to an isolated coronary fibromuscular dysplastic lesion occluding the left coronary ostium and review the literature to better characterize FMD and its relationship to sudden death.

2. Case report and scene investigation

A previously healthy 21-year-old white man was found unresponsive in the hallway of his apartment building by his brother. Emergency Medical Services responded and pronounced him dead. He was last seen alive in apparent good health approximately 4 hours prior by his girlfriend. It is unclear if the decedent was engaging in any exertional activity. There is anecdotal history of cocaine usage provided by his brother who said the decedent had been using cocaine regularly for the last 2 months. Investigation at the scene revealed no evidence of prescription medications, alcohol, drugs, or drug paraphernalia. The body was transferred to the local medical examiner office for postmortem examination due to suspicion of drug overdose.

3. Postmortem findings

The decedent was 1.82 m tall and weighed 83 kg. The heart weighed 410 g (reference range for a normal adult male heart weight: 0.45% of body weight in kilograms). For our case (83 kg), the normal weight would be 373 g (323–423 g) [48]. An irregular 0.5 cm×0.4 cm×0.3 cm,

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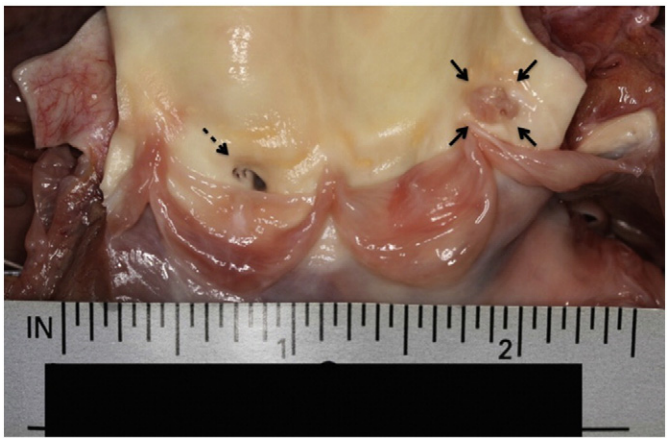

Fig. 1. Gross heart specimen demonstrating an irregular, tan-pink sessile polyp arising from the aortic wall with complete occlusion of the left coronary ostium (solid arrows). The polyp shows no connection with the aortic valve leaflets. The right coronary ostium is widely patent (dashed arrow).

soft, tan-pink, sessile polyp completely occluded the left coronary ostium. The ostium of the right coronary artery was widely patent (Fig. 1). No other lesions were present in the rest of the coronary arteries, the aorta, brachiocephalic, subclavian, common carotid, main pulmonary, or renal arteries. The heart, including cardiac valves and myocardium, was grossly and microscopically unremarkable. Comprehensive postmortem toxicology analysis was negative for ethanol, cocaine, opiates, and other substances of abuse.

3.1. Microscopic examination and ancillary tests

The left coronary ostium was occluded by a myofibroblastic cell proliferation in a background of a loose connective tissue matrix (Fig. 2A–C). The myofibroblasts were cytologically bland, spindle-shaped cells with no mitotic activity and abundant eosinophilic cytoplasm (Fig. 3A and B). No atheromas, inflammatory cells, nor acute or chronic ischemic changes were present within or surrounding the lesion. The media and adventitia were not involved. Elastic-Van Gieson (EVG) stain confirmed the intimal proliferation and revealed a fragmented and discontinuous internal elastic lamina (IEL) with focal reduplication (Figs. 4 and 5, A and B). The lesional cells showed immunoreactivity for smooth muscle actin and vimentin consistent with myofibroblastic origin. Masson's trichrome stain and immunoreactivity to collagen type IV highlighted lesional fibrocollagenous tissue (Fig. 6A–D). Immunostains for myogenin and CD 34 were negative, ruling out skeletal muscle and vascular origin, respectively. The microscopic findings of localized fibrointimal proliferation of the coronary ostium with associated disruption of the IEL were consistent with the diagnosis of isolated ostial coronary FMD, intimal subtype.

4. Discussion

Isolated or solitary coronary ostial stenosis has been defined as a localized narrowing of the coronary ostium, greater than 50%, with no evidence of obstructive distal vessel disease or any other coronary artery or aortic disease [47]. Common causes include atherosclerosis [47], aortic valve disease [49], cardiovascular syphilis [50], Takayasu's aortitis [51], and as a complication of heart surgery [52]. Isolated coronary ostial stenosis secondary to FMD is rare [42,47]. Yamanaka and Hobbs in 1993 [47] reported the Cleveland Clinic experience of 125,000 patients who underwent coronary arteriography between 1960 and 1988 and found 128 patients having 50–99% stenosis of one or both coronary ostia. FMD was found in only one patient who had surgical revascularization with a bypass graft and subsequently died in the immediate postoperative period. The patient was a 46-year-old female with 90% obstruction and perimedial type of FMD. The cause of death was "ventricular tachycardia" secondary to complete occlusion of the vein graft at the anastomotic site, most likely by thrombus. Another case of FMD involving the coronary ostium was described in a 28-year-old man who presented with angina on exertion that was relieved after coronary angioplasty [42].

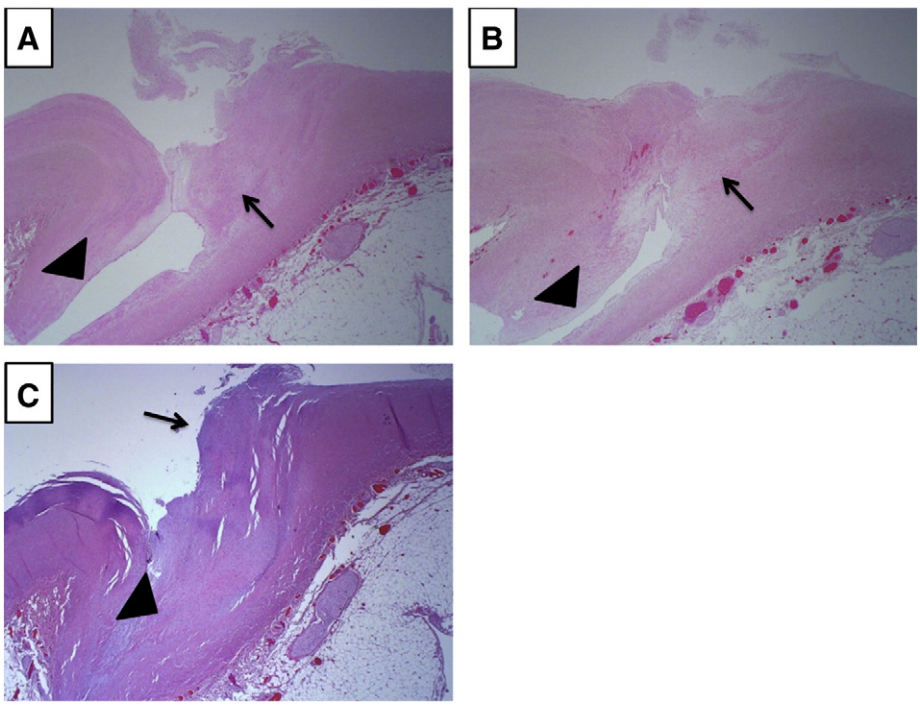

Fig. 2. A, B, and C: low-power view showing a complete occlusion of the left coronary artery ostium by the cellular proliferative lesion (arrow). The entrance of the left main coronary artery can be seen distal to the lesion (arrow head) (A and B). Low-power view of a deeper section demonstrating the polypoid nature of the lesion (arrow) with confinement to the ostium (arrow head) (C) (A, B, and C: hematoxylin and eosin, magnification ×4).