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Spontaneous coronary artery dissection secondary to intimal fibromuscular dysplasia (RCD code: I-1C.0)

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Abstract

Fibromuscular dysplasia (FMD) is a nonatherosclerotic, noninflammatory arterial disease of unknown etiology. Histologically, FMD is classified on the basis of arterial layer involved into intimal, medial and perimedial types. The medial form represents approximately 80% of the cases of FMD, perimedial and intimal forms are rare and account for 10% and 5% of the cases respectively. Clinically, FMD can manifest as arterial stenosis, occlusion, aneurysm or dissection. Coronary artery manifestations of FMD are rare but potentially fatal. Most cases present as a dissection of the involved coronary artery or its branch, which clinically leads to unstable angina, myocardial infarction, left ventricular dysfunction, or potentially sudden cardiac death. Various recent studies have established FMD as the leading risk factor for Spontaneous Coronary Artery Dissection (SCAD). We present autopsy findings of a 71 year old woman who suffered a spontaneous dissection of the posterior descending artery secondary to FMD-intimal type. The optimal diagnostic and treatment guidelines for SCAD secondary to FMD have not been established due to the lack of evidence. Further research into the pathogenesis, molecular and cellular biology, epidemiology and clinical management of FMD and its coronary artery manifestations is required. JRCD 2017; 3 (2): 56–58

Key words: rare cardiovascular disease, coronary artery disease, pathology, posterior descending artery, chest pain

Introduction

Fibromuscular dysplasia (FMD) is nonatherosclerotic, noninflammatory vascular disease that most commonly affects the renal and carotid arteries but can involve any arterial bed. The incidence of coronary artery involvement by FMD is not exactly known, but is considered to be very rare. Histologically, FMD is broadly classified on the basis of the arterial layer involved into intimal, medial and perimedial fibroplasia. Clinically, FMD can manifest as arterial stenosis, occlusion, aneurysm, or dissection [1–4]. A very strong association between FMD and Spontaneous Coronary Artery Dissection (SCAD) has been established in several studies [3,5]. This report describes autopsy findings of a patient who suffered a spontaneous dissection of the posterior descending artery secondary to intimal FMD.

Case presentation

A 71 year old woman with a past medical history of hypertension, hypothyroidism, depression, kidney donation for transplant, arthritis and obesity presented with chest pain. Her pain started when she was lifting groceries and was sharp, substernal, radiating to the back, with no relieving or exacerbating factors. Her electrocardiogram showed left axis deviation and poor R wave progression but no ST segment elevation. Her troponin I levels were 13.857 ng/ml on initial presentation and reached a peak of 47.573 ng/ml on the second post admission day. She was treated for Non ST Segment Elevation Myocardial Infarction (NSTEMI). Cardiac catheterization showed markedly tortuous coronary arteries but failed to show any occlusive atherosclerosis. Posterior descending artery was not examined during the angiography because no vascular disease was noted in the more proximal coronary arterial tree. She showed considerable improvement in her symptoms but suddenly died on the 5th post admission day due to ventricular free wall rupture leading to cardiac tamponade.

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Autopsy exam was requested and was restricted to lungs and heart only. Examination of the heart showed mild cardiomegaly. Coronary arteries were normally distributed, with the left-dominant pattern. Right coronary, left anterior descending and left circumflex coronary arteries were soft, patent and free of any calcification or atheromas. Left circumflex branch divided into two at the level of the posterior descending artery. Posterior descending coronary artery showed a dissection throughout its length causing complete obstruction of the true lumen (Figure 1). Tracing the dissected posterior descending artery led to a mottled yellow and red transmural area measuring 3 cm in width, representing an acute infarct in the posterior wall of the left ventricle. A perforated hemorrhagic track extended through the infarct to the epicardial surface causing cardiac tamponade.

Microscopic examination of the left anterior descending, right coronary, left circumflex and posterior descending arteries showed patent vessels without any prominent atherosclerosis, however there was intimal hyperplasia with irregularly arranged mesenchymal cells in a loose matrix of connective tissue and a fragmented internal elastic lamina (Figure 2). The findings were confirmed with an elastin stain. A dissection was seen in the posterior descending artery throughout its course (Figure 3). These histologic findings are consistent with intimal FMD leading to spontaneous dissection of the posterior descending artery.

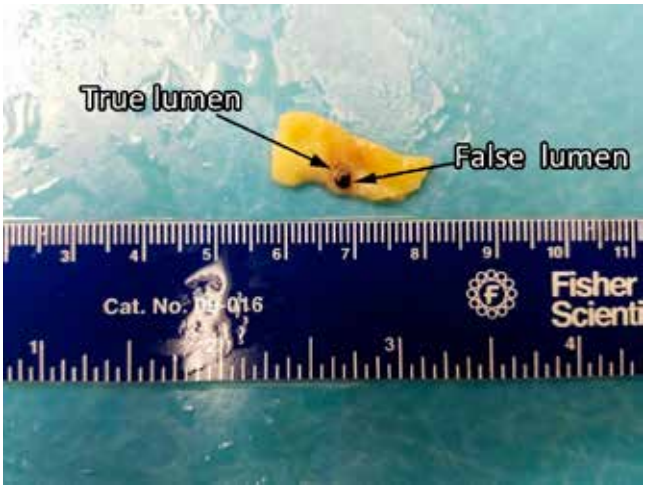

Figure 1. Cross section of the posterior descending artery showing a dissection – dissecting hematoma is occluding the true lumen

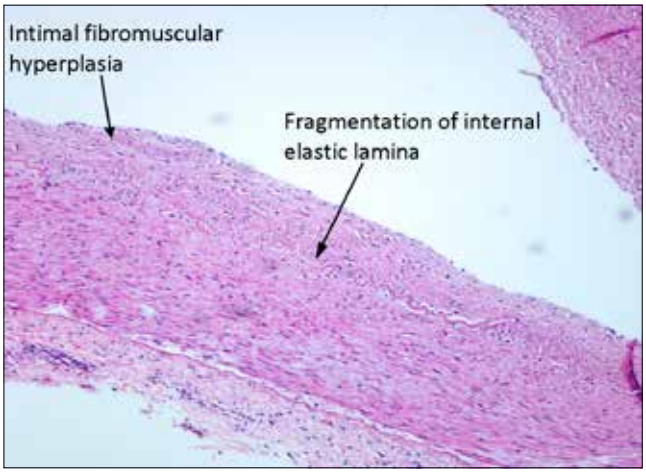

Figure 2. Hematoxylin and eosin stain on section of coronary artery wall showing fragmented internal elastic lamina and fibromuscular hyperplasia of the tunica intima

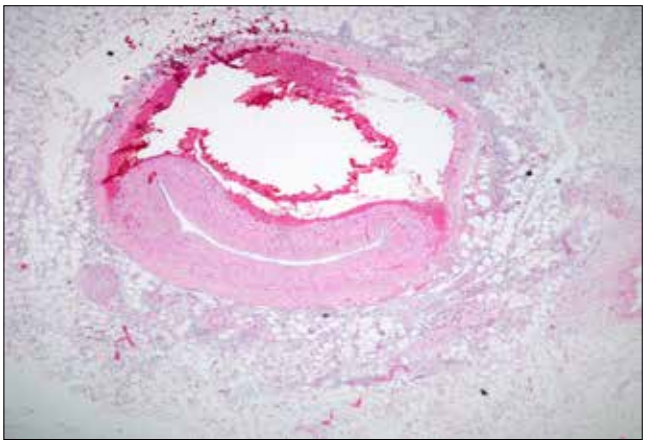

Figure 3. Hematoxylin and eosin stain on section of posterior descending artery showing a dissection

Discussion

FMD is a non-atherosclerotic, non-inflammatory vascular disease that has been reported in virtually every arterial bed, but most commonly affects the renal and extracranial carotid and vertebral arteries. Manifestation of FMD in coronary arteries is rare. The exact prevalence is unknown. In the U.S. Registry for FMD 91% of the patients with FMD are female and the mean age at diagnosis is 51.9 ±13.1 years [3].

Even though FMD was first described in 1938, the exact pathogenesis has not been elucidated to date. General consensus is, that it is a disease with a multifaceted etiology. Since majority of patients with FMD are women, exposure to endogenous or exogenous estrogens has been postulated, but the number of pregnancies and the frequency of oral contraceptive use does not differ between patients with FMD and matched controls. It has also been found that FMD is more prevalent in patients who smoke as compared to matched controls but mechanisms by which smoking contributes to FMD are not known [6]. The occurrence of renal FMD in identical twins suggests a possible genetic component [7]. Molecular genetic studies have also implicated autoimmunity and polymorphisms of the renin angiotensin system but none of these findings has ever been confirmed.

Histologically, FMD is broadly classified based on the arterial layer involvement, into intimal, medial and perimedial fibroplasia. In our case, the histologic sections of patient's coronary arteries showed intimal FMD which is the most rare form accounting for only 5% of all cases. It is characterized by irregularly arranged mesenchymal cells within a loose matrix of subendothelial connective tissue and a fragmented internal elastic lamina. Perimedial FMD affects approximately 10% of cases and involves excessive tissue

deposition at the junction of the media and adventitia [6]. These abnormal histologic changes cause structural and functional alterations in the vessel wall which can lead to stenosis, occlusion, aneurysm, or dissection.

Various recent studies have established FMD as the leading risk factor for SCAD, which is defined as an expanding hematoma leading to separation of layers of the coronary arterial wall. In a recent analysis of 738 patients enrolled in the U.S. Registry for FMD, 165 (22.4%) had a dissection of an artery other than the aorta. Of those 165 patients, carotid arteries were involved in 73%, vertebral arteries in 19%, renal arteries in 16%, and coronary arteries in 4.8% [8]. In a study of 50 patients at Vancouver General Hospital with non-atherosclerotic SCAD, 86% of all patients had imaging consistent with FMD of at least one non-coronary artery [9]. Coronary artery manifestations of FMD are rare but potentially fatal. Most cases present as a dissection of involved coronary artery or its branch, which clinically leads to unstable angina, myocardial infarction, left ventricular dysfunction, or potentially sudden cardiac death.

The optimal guidelines for management of a patient with FMD and its coronary artery manifestations have not been defined due to the lack of prospective trials and evidence based recommendations. The general consensus is to use a conservative approach in stable patient without active myocardial ischemia because most cases of SCAD heal spontaneously. Interventional approach in the form of intracoronary stent implantation and rarely coronary artery bypass grafting is reserved for unstable patients with active myocardial ischemia [10].

Conclusion

In conclusion, FMD with coronary artery involvement should be considered in all patients who present with acute coronary syndrome without significant coronary atherosclerotic disease. Even though various studies have established FMD as the leading risk factor for SCAD, exact mechanisms by which FMD develops and contributes to SCAD have not been defined. Further research is required to understand the epidemiology, etiology, diagnosis and treatment guidelines for FMD. Furthermore, investigation into the molecular and genetic etiology of FMD may open the doors to targeted therapies in the future.

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