Left Main Coronary Artery Diverticulum: Case Report and Review of the Literature

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Abstract

Coronary artery disease is a major cause of morbidity and mortality, and while most commonly is atherosclerotic, it can present with variable manifestations, both congenital and acquired. One such manifestation is coronary aneurysm, which is a localized dilatation of a coronary artery wall segment to greater than 1.5 times the adjacent normal segments. While a dilated outpouching of a coronary artery has been commonly classified as a coronary aneurysm, a non-dilated outpouching is rare and is referred to as a diverticulum, with only one previous case report in the literature. It is conceivable that other cases of coronary artery diverticulum may have been previously reported as an aneurysm, given the overlapping angiographic appearance. We present a case of a 72-year-old female patient with an incidental finding of left main coronary artery diverticulum on diagnostic coronary angiogram done for preoperative liver transplantation evaluation.

Keywords: Coronary artery diverticulum; Coronary artery anomalies; Coronary artery aneurysm

Introduction

Coronary artery disease is heterogeneous and while in most cases is acquired, can also be congenital. Congenital coronary artery anomalies are a diverse group of disorders which are rare and found in less than 1% of patients on angiography. Coronary artery diverticulum is rare, and was only recently reported in the literature. We present a case of a 72-year-old female patient who was found to have an asymptomatic left main coronary artery diverticulum on diagnostic coronary angiogram.

Case Report

A 72-year-old Caucasian female with past medical history of advanced liver cirrhosis, hypothyroidism, atrial fibrillation and mild systolic heart failure was being evaluated for liver transplantation. She had symptoms of shortness of breath on exertion and worsening lower extremity edema. She denied chest pain, palpitations, lightheadedness or syncope. Her electrocardiogram revealed atrial fibrillation with a rate of 58 beats per minute and left bundle branch block. An echocardiogram revealed a left ventricular ejection fraction of 45-50% with abnormal septal motion, severe tricuspid regurgitation and pulmonary artery systolic pressure of 54 mm Hg. She underwent elective left heart catheterization for evaluation of her symptoms prior to consideration for liver transplantation. Coronary angiography showed normal coronary arteries, with the exception of a non-aneurysmal outpouching arising from the left main coronary artery which seemed to fill completely with contrast, consistent with a coronary artery diverticulum.

Discussion

Coronary artery disease is a ubiquitous cause of major morbidity and mortality [1]. Commonly this is the result of atherosclerotic disease, which is well characterized with regards to its pathophysiology and treatment [2]. A wide variety of other less common acute and chronic pathologies can also affect the coronary arteries, which may be either acquired or congenital [3]. Such coronary pathologies often resemble equivalent aortic pathologies, yet on a smaller scale, and treatment can be comparable [4].

Obstructive endovascular lesions are most commonly caused by coronary atherosclerosis, which has long been recognized as a multifactorial disease leading to the deposition of lipids in the arterial wall [5], with well-described diagnosis, management [6] and prevention guidelines [7]. Less commonly, fibromuscular dysplasia (FMD) can affect the coronary arteries, predominantly in middle-aged women, but can also occur at any age and in either gender [8]. Coronary FMD has been reported to result in stenosis, aneurysms, dissection, or occlusion of the coronary arteries, and is often treated in a similar manner to coronary atherosclerosis [9].

Coronary vasospasm is a functional abnormality of mildly to moderately diseased segments of the coronary arteries [10].
The common pathophysiological phenomenon is vascular smooth muscle hyper-reactivity which causes the spasm [11], with a potential role for vascular inflammation [12]. Management, in general, has focused on removal of offending agents or triggers, in addition to the use of vasodilators including calcium channel blockers and nitrates [13]. Endovascular therapy is usually reserved for recurrent cases despite medical therapy [14] or persistent occlusion causing ongoing myocardial ischemia and infarction [15].

Coronary artery wall abnormalities which mimic aortic syndromes include coronary artery dissection, hematoma and ulcer. Coronary artery dissection is often spontaneous, with several reported predisposing and associated conditions [16]. Iatrogenic, especially catheter-related, coronary dissections have also been reported especially in the setting of coronary artery manipulations during angiography and intervention [17]. Treatment options for coronary dissection have included conservative management [18], endovascular therapy [19] and surgical intervention [20]. Similar to aortic hematomas, coronary artery hematomas have been reported with hemorrhage limited to the media-adventitia layers, resulting in hematoma formation without any identifiable intimal flap [21]. These can be spontaneous [22] or iatrogenic, following coronary stenting [23]. Although ulcerated atherosclerotic coronary lesions are not uncommon, a true coronary artery wall ulcer extending deeply into the intima is rare, with one case reported in the literature and treated conservatively [24]. Treatment of coronary hematomas and ulcers is individualized and is often conservative in the absence of symptoms, thrombosis or embolization.

Coronary artery fistulae have been described draining coronary artery flow into another vascular structure or cardiac cavity [25]. These are mostly congenital [26], with infrequent cases of iatrogenic coronary fistulae reported [27]. Various management options have been proposed based on size and physiology of the fistula, complications and symptoms [28]. Coronary ectasia is diffuse dilatation of the coronary arteries found in 1-6% of cardiac catheterization procedures, and is often associated with coexisting coronary artery disease [29]. The etiology of coronary ectasia remains unclear; however, histopathologic studies reveal extensive collagen and elastin degradation within the coronary arterial wall [30]. Although ectasia of the coronary arteries can be asymptomatic [31], it has been reported to cause myocardial infarction secondary to embolization [32], coronary artery vaso spasms [33, 34] and progression to coronary aneurysm [35]. Ectasia can also affect saphenous vein bypass grafts [36].

Coronary artery aneurysm is a localized dilatation of a
coronary artery wall segment to greater than 1.5 times the adjacent normal segments, with a reported incidence of up to 5% of coronary angiograms [37]. Although they are often interchangeably labeled as ectasia, coronary aneurysms tend to be more focal and are associated with several etiologies other than atherosclerosis, including inflammation, infection, trauma and congenital [38]. Inflammatory coronary artery aneurysms are a known complication of Kawazaki disease with well-established diagnostic and management guidelines [39]. Coronary aneurysms have also been reported to cause myocardial infarction [40]. Although they are most often isolated, they may be associated with fistulous connections [41]. Infectious coronary aneurysm has also been reported following coronary intervention [42]. Many factors influence management of coronary

| Pathology                     | Schematic | Description                                                                 |
|------------------------------|-----------|-----------------------------------------------------------------------------|
| 1   Coronary atherosclerosis | ![Coronary Atherosclerosis](image) | Lipid deposition causing plaque formation and stenosis. Well defined risk factors and treatment guidelines. Treatment is medical, endovascular or surgical intervention. |
| 2   Coronary fibromuscular dysplasia | ![Coronary Fibromuscular Dysplasia](image) | Segmental, non-inflammatory, non-atherosclerotic disease of the artery wall musculature, causing “beaded” appearance. Treatment is similar to atherosclerosis. |
| 3   Coronary spasm            | ![Coronary Spasm](image) | Endothelial hyper-reactivity causing abrupt interruption of flow and ischemia, rarely infarction. Treatment is vasodilators. Rarely requires endovascular intervention. |
| 4   Coronary dissection       | ![Coronary Dissection](image) | Spontaneous or iatrogenic intimal tear causing intramural hematoma that compresses the true lumen. Treatment is medical or endovascular intervention, rarely surgical. |
| 5   Coronary hematoma         | ![Coronary Hematoma](image) | Hemorrhage within the media-adventitia layers, resulting in hematoma formation without any identifiable intimal flap. Treatment is medical, occasionally interventional if obstructive. |
| 6   Coronary ulcer            | ![Coronary Ulcer](image) | Ulceration of an atheromatous plaque that extends deeply through the intima. Treatment of underlying atherosclerosis. Endovascular covered stent if at risk of rupture. |
| 7   Coronary fistula          | ![Coronary Fistula](image) | Vascular communication draining coronary artery flow into another vessel or cardiac cavity. Treatment is medical, if causes coronary “steal” and ischemia, endovascular coiling or surgery. |
| 8   Coronary ectasia          | ![Coronary Ectasia](image) | Diffuse dilatation of the coronary arteries, usually in atherosclerotic segments. Treatment of underlying atherosclerosis is medical, with endovascular intervention in stenotic segments. |
| 9   Coronary aneurysms        | ![Coronary Aneurysms](image) | Localized dilatation or aneurysmal outpouching of a coronary segment to > 1.5 × adjacent normal segments. Treatment is medical; interventional with covered stents or surgical if symptomatic. |
| 10  Coronary Diverticulum     | ![Coronary Diverticulum](image) | Localized non-aneurysmal outpouching of a coronary artery segment. Rare and can be treated similar to coronary aneurysms, medically or with covered stent, depending on presentation. |

Figure 3. Schematic presentation and brief description of coronary endoluminal lesions.
A coronary diverticulum refers to a non-aneurysmal (non-dilated) outpouching originating from the wall of a coronary artery. To our knowledge, there has been only one previous case report of coronary artery diverticulum in the literature. Shkolnik and Salka [44] reported a 33-year-old female with non ST elevation myocardial infarction who had a right coronary artery diverticulum containing thrombus which was successfully treated with thrombectomy and a covered stent. Our patient was found to have an incidental diverticulum of the left main coronary artery. She suffered advanced liver cirrhosis, which has been associated with various forms of acquired vascular malformations [45]; however, whether her coronary diverticulum is congenital or acquired cannot be discerned.

Conclusion

Coronary artery diverticulum is a very rare coronary abnormality. It can be asymptomatic and found incidentally on coronary angiography, as in our patient. On the other hand, it may lead to recurrent coronary thrombosis with acute coronary syndrome requiring endovascular intervention. Even though our patient is the second reported case with a coronary diverticulum in the literature, it is conceivable that other cases of coronary artery diverticulum may have been previously reported as an aneurysm or ectasia, given the overlapping angiographic appearance. A classification system of coronary endoluminal lesions with well-defined recognition criteria is called for, to avoid misdiagnosis and facilitate proper identification and implementation of necessary therapeutic modalities. Figure 3 provides schematic presentation of various coronary lesions with brief descriptions.

Disclosures

All authors declare no disclosures related to this manuscript.

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