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Left circumflex coronary artery to hepatic vein fistula: a case report and brief review of coronary vasculogenesis

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Abstract Coronary artery fistulas are rare coronary artery anomalies. Their clinical significance varies from a long asymptomatic course to overt heart failure and death. They are often detected incidentally with diagnostic coronary angiograms. Cardiologists increasingly encounter coronary artery fistulas secondary to recent improvements in cardiovascular imaging modalities. Management is still controversial, especially in asymptomatic cases with less significant shunts. Here, we describe a 62-year-old woman with a left circumflex artery to hepatic vein fistula found on coronary angiography. The patient is being managed conservatively using nuclear imaging studies and echocardiographic evaluation.

Keywords: Vasculogenesis; Fistulae; Coronary angiography

1. Introduction

Coronary artery fistulas (CAF) are uncommon coronary artery malformations. They can be detected in various age groups with different presentations. Conventional coronary angiography is the diagnostic procedure of choice which allows management with coil embolization if needed.

2. Case report

The patient is a 62-year-old white female who was referred to cardiology outpatient clinic for substernal chest pain. This patient started having chest pains 3 years prior to presentation; however, she did not seek any medical attention until recently when the frequency and intensity of her pain progressively increased. She described the chest pain as substernal, nonradiating, "sharp," and not associated with exertion. Patient denied having dyspnea, orthopnea, paroxysmal nocturnal dyspnea, or lower extremity swelling. She reported palpitations occasionally at rest and under exertion. Her past medical history was nonsignificant. She was not using any medicine. She denied cigarette, alcohol, or illicit drug use. On physical examination, the blood pressure was 140/80 mmHg, with a heart rate of 80/min. The patient had a normal S1 and S2, and a 2/6 midsystolic murmur best heard at right sternal border. No rubs or gallops were appreciated. Point of maximal intensity was not displaced. Lungs were clear to auscultation with no crackles or wheezing. Abdomen was nontender, nondistended with no hepatosplenomegaly. She did not have peripheral edema.

Complete blood count and comprehensive metabolic panel (blood urea nitrogen, creatinine, serum electrolytes, and liver function tests) were normal. Thyroid function tests

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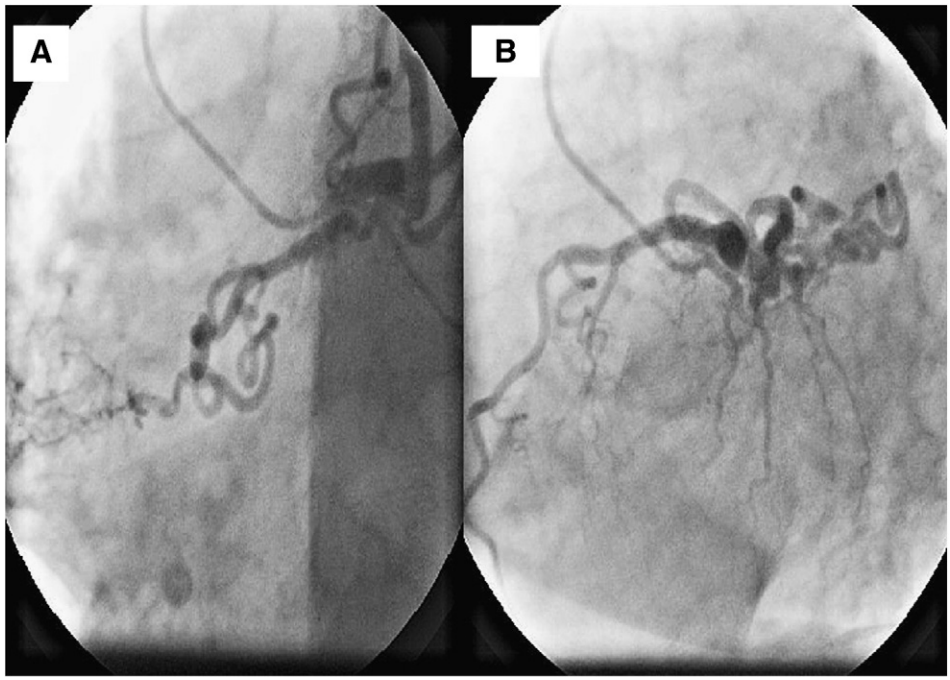

Fig. 1. (A and B) Coronary angiogram of the patient in left anterior caudal (30°) (A) and lateral (B) view demonstrating the left circumflex artery hepatic vein fistula.

including thyroid-stimulating hormone, free T3, and free T4 were within normal range. Her low-density lipoprotein level was 110 mg/dl.

Resting electrocardiogram revealed normal sinus rhythm with no ST-T changes. Initial cardiac workup included a transthoracic echocardiogram, which showed concentric left ventricular hypertrophy, sclerosis of the aortic valve with mild aortic insufficiency, and a normal ejection fraction of 60%. Transmitral flow pattern revealed diastolic dysfunction (E/A ratio of 0.7). All segments contracted normally without any evidence of hypo- or akinesis. Treadmill exercise test was nondiagnostic. Diagnostic coronary angiogram revealed circumflex artery to hepatic vein fistula (Fig. 1A and B) The coronary arteries were otherwise normal. We decided to proceed noninvasive ischemia evaluation and shunt detection. Patient underwent pharmacological single photon emission computed tomography using thallium 201, which did not show any ischemia. Transthoracic echocardiogram revealed small left to right shunt (Qp/Qs:1.2) with normal right heart dimensions. After discussion of her condition with the patient, we decided to manage her conservatively with regular follow-ups.

3. Discussion

CAF are communications between a coronary artery and the systemic or pulmonary circulation or a cardiac chamber. Many CAF result from congenital anomalies during coronary development, which begins in the third week in the embryo [1] (Fig. 2). The cells that generate coronary arteries originate from an epithelium associated with septum transversum. This outgrowth from septum transversum, called the proepicardial organ (PEO), is composed of epithelial cells and is located near the sinoatrial pole of the heart. This is the critical initial stage in the formation of coronary vasculature and is independent from myocardial and endocardial development. The PEO contacts with the surface of the heart followed by epithelial cell migration spreading over the heart (Fig. 2). Thus, coronary vessels originate from subepicardial coalescence of the mesenchymal cells originating in the atrioventricular sulcus in the embryo. At this point, there is massive number of cells since every myocyte is in contact with cells of coronary vasculature. Subsequently, these vasculogenic epithelial cells link to form networks called “plexi,” which are the precursors of coronary arteries. Coronary arteries were originally thought to be outgrowths of aortic root. However, this belief was discarded after demonstration of arterial precursors’ self organization and vascular plexus formation in the subepicardial space. The drive for coronary arterial patterning is not clearly understood, but several mechanisms, such as growth factors and hypoxia play a key role. Transforming growth factor, vascular endothelial growth factor A, and basic fibroblast growth factor stimulate vasculogenesis [1]. Hypoxia, by itself, promotes growth factor expression, which, in turn, stimulates vasculogenesis. However, apoptosis and pressure changes also contribute to this process. A unique feature of coronary vessel development is the final connection of the coronary arteries to the