Woven Coronary Artery Anomaly: A Review of Current Evidence

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Abstract

Woven coronary artery (WCA), also known as woven coronary artery anomaly or malformation, is a vascular malformation with an unclear pathogenesis and unknown incidence. On coronary angiography, WCA manifests as intertwined branching conduits with or without distal reanastomosis that most commonly involves the right coronary artery and predominates in males between the fifth and sixth decade. WCA typically follows an insidious clinical course and is associated with a normal coronary flow (i.e. TIMI grade 3 flow). Slit-like radiolucency between entangled channels may be mistaken as the filling defects observed in spontaneous coronary artery dissection, recanalization of an organized thrombus, or chronic occlusion with bridging collaterals. Recognition of this benign entity is crucial for interventional cardiologists in order to avoid the "oculo-radiolucent reflex", the temptation to perform unwarranted angioplasty on intraluminal radiolucency. As turbulent flow at the entrance of intertwined channels may predispose to atherosclerotic plaque development, prophylactic antiplatelet therapy is usually advisable for patients diagnosed with such malformation. More data from large observational studies and angiographic registries will be required to inform clinicians regarding the optimal management for WCA.

Keywords: Woven Coronary Artery; Woven Coronary Malformation; Coronary Vessel Anomaly; Coronary Angiography; Atherosclerosis; Coronary Artery Dissection

Abbreviations

OCT: Optical Coherence Tomography; SCAD: Spontaneous Coronary Artery Dissection; TIMI: Thrombolysis in Myocardial Infarction; WCA: Woven Coronary Artery

Background

Woven coronary artery (WCA), manifesting as intertwined branching conduits with distal reanastomosis, is a relatively rare malformation with an unknown incidence. In 1988, Sane., et al first reported this condition as a “figure-of-eight” pattern without associated atherosclerosis on coronary angiography [1]. Over the next three decades, subsequent WCA publications consist mostly of discrete case reports or case series. With the advance of coronary imaging modality and the emergence of research on coronary artery diseases, the diagnosis of WCA has gained prominence in the medical literature. As such, among all the reported cases to date, more than one-half were published in the past 10 years. WCA is usually detected incidentally on coronary angiography while assessing patients with ischemic equivalents [2,3]. In certain instances, these anomalies represent epiphenomena and do not impede coronary flow or jeopardize myocardial perfusion. Per contra, the twisted conduits may accelerate the development of atherosclerosis and heighten the risk for thrombus formation, which eventually leads to acute coronary syndrome [2,4]. Constant vigilance coupled with advanced intracoronary imaging is essential for interventional cardiologists to establish a definitive diagnosis and avoid undue angioplasty. The article summarizes the contemporary body of knowledge on woven coronary artery in hopes of raising the awareness of this under-recognized disease.
Pathophysiology

The exact pathophysiology of WCA remains largely unknown. WCA has been suggested as a congenital malformation without a clear genetic association [5]. Coronary artery dissection, either spontaneous or related to Kawasaki disease, has also been implicated in the pathogenesis [3,6]. Furthermore, the split conduits observed in the WCA were postulated to occur as a consequence of coronary thrombus recanalization [3].

Although WCA generally follows a benign clinical trajectory, ischemia referable to this anomaly has been observed in several reported cases. It was postulated that at the branching point proximal to the divided conduits, fluid mechanics play a central role in the development of a proatherogenic endothelial phenotype. Specifically, non-laminar flow patterns with a disrupted shear stress may serve as a signal that influences atherogenesis, thereby resulting in vascular wall dysfunction and adaptive arterial remodeling as the disease progresses [7,8].

Histopathology

On histopathology examination, WCA is featured by: (1) preserved integrity of endothelial lining with inward remodeling; and (2) exuberant intimal thickening mostly composed of irregular fibrosis [9]. Current knowledge concerning the natural history and pathophysiology suffers from the scarcity of cases and delayed or missed diagnosis due to physician unawareness. Further studies are warranted to elucidate the etiopathogenesis of woven coronary artery.

Epidemiology and Demography

The true incidence and prevalence of WCA remain unclear. Among the reported cases in the English literature, WCA was found to predominate in males between the fifth and sixth decades [1-6,9-20]. Risk factors for coronary artery disease were commonly observed in WCA patients, including dyslipidemia, hypertension, and tobacco use. Association with Kawasaki disease was reported in one pediatric patient [3]. Little to none is known about the racial predilection for WCA.

Clinical Presentation

The majority of patients with WCA present for evaluation of symptoms or findings suggestive of ongoing myocardial ischemia, with chest pain as the most common manifestation. Patients may also be free of anginal symptoms or present with atypical chest discomfort and palpitations. Conversely, woven coronary artery malformation could be detected incidentally on coronary angiography when evaluating other cardiovascular disorders such as perioperative assessment of previous myocardial infarction and heart failure.

Coronary Angiography

Coronary arteriography remains the gold standard for diagnosing woven coronary artery anomaly. However, universal diagnostic criteria for WCA have not been established. Woven coronary artery is usually defined as a coronary artery malformation characterized by intertwined branching channels with distal reanastomosis. Among the reported cases, the right coronary artery is the most commonly affected epicardial vessel. The malformation can involve one or more epicardial arteries or their branches, where they divide into a variable number of thin channels traversing distally with a twisted or parallel course. These channels eventually rejoin the main lumen or, less commonly, subdivide into smaller conduits. The site of reanastomosis may be absent or difficult to localize despite the use of multiple orthogonal projections and digital zooming on coronary angiograms. The affected segment is usually limited to several centimeters in length.

In the absence of concomitant stenosis or other pathology, the flow in the coronary artery afflicted by woven anomaly is typically normal (i.e. Thrombolysis in Myocardial Infarction [TIMI] grade 3 flow). There may be associated luminal stenosis, atherosclerosis, or thrombus formation. Of note, in five of the reported cases, the diagnosis of WCA was not considered initially and was mistaken for recanalization of organized thrombus or coronary artery dissection with or without plaque encroachment [4,6,12,15,16].

Optical Coherence Tomography

Intracoronary imaging, particularly with optical coherence tomography (OCT), may be useful in establishing a definitive diagnosis of woven coronary artery [16]. Enhanced spatial resolution of OCT allows cross-sectional delineation of the vessel microstructure and concomitant lesions. WCA typically manifests as multiple lumens with preserved integrity of the intima and media without coexisting pathology such as plaque or thrombus [18]. In contrast, recanalized thrombus is identified as a lotus root- or honeycomb-like structure with communicating cavities separated by signal-rich, high backscattered tissue [21,22]. However, OCT may not always be readily available, and advancing the imaging probe across tortuous, thin channels could pose a risk for procedural complications [5].

Differential Diagnosis

The differential diagnosis of woven coronary artery includes spontaneous coronary artery dissection (SCAD), recanalization of organized thrombus, and chronic occlusion with the development of bridging collaterals, all of which may be mistaken as filling defects on coronary angiography [10]. WCA is commonly considered as a non–infarct-related malformation that does not impede coronary flow or jeopardize myocardial perfusion [12,15]. Angiographically, SCAD can be classified into type 1 (evident arterial wall stain with multiple radiolucent lumens), type 2 (diffuse smooth stenosis of varying severity), type 3 lesions (focal or tubular stenosis mimicking atherosclerosis), type 4 (dissection leading to abrupt total occlusion, usually of a distal coronary segment) and intermediate type 1/type 2 lesions [23,24]. Notably, type 1 SCAD appears as a filling defect associated with the intimal tear. Propagation of the dissection may compress the true lumen and compromise the blood flow. Anterograde coronary flow in dissected arteries may be minimally disturbed if the false lumen reunites with the true lumen. Natural history provides an important clue for differentiating WCA from SCAD lesions. Spontaneous healing has been observed in the vast majority of SCAD lesions, whereas the angiographic appearance of WCA remains unchanged when followed by repeated angiography. Recanalized thrombus manifests as a linear, intraluminal radiolucency associated with variable resistance to flow [25,26]. Bridging collaterals represent tortuous, intracoronary conduits of microvessels bypassing the site of chronic occlusion. In this context, it is prudent to acquire previous medical and radiographic records for comparison. Another distinctive feature of woven coronary artery is the absence of classic angina symptoms in a patient with a normal stress test assessed by treadmill, echocardiography, or myocardial scintigraphy [6]. Multiple orthogonal views with digital zooming on coronary angiography should be obtained to discriminate a woven coronary artery from other entities. Intravascular ultrasound, optical coherence tomography, or computed tomography coronary angiography may be considered when diagnostic uncertainty exists.

Therapeutic Considerations

Thus far there has been no consensus or practice guideline on the optimal management strategy for WCA. Among the reported cases with more than one year duration of follow-up, none had adverse cardiovascular events associated with woven coronary arteries or angiographic progressions in the malformation. Therefore, woven coronary artery malformation has been regarded as a benign entity associated with normal coronary flow and myocardial perfusion. Certain considerations should be undertaken while determining treatment strategies. First, disrupted hemodynamics at the entrance of intertwined branching channels may be conducive to plaque development, making prophylactic antiplatelet therapy advisable for persons with the WCA malformation. Second, although this anomaly could serve as the basis for atherosclerosis and subsequent thrombus formation [2,15], revascularization should be reserved for selected cases with ischemia attributable to luminal occlusion within the woven coronary artery. Lastly, the structural configuration carries interventional implications. Percutaneous coronary intervention on the culprit lesion at or distal to the woven segment may lead to periprocedural complications, as advancing the guidewire or dilating device across the divided channels could be challenging in cases with a severely intertwined morphology [18]. Thus far, there is one report of successful recanalization of occluded woven segments [4]. Mostly, patients with WCA have a generally favorable prognosis and may be managed medically with antiplatelet agents.
Conclusions

Woven coronary artery is an uncommon anomaly with an uncertain pathogenesis. This benign malformation appears as a non–infarct-related filling defect on coronary angiography. Orthogonal, close-up views should be obtained to differentiate the anomaly from spontaneous coronary artery dissection, recanalization of organized thrombus, or chronic occlusion with bridging collaterals. Recognition of this anomaly is crucial for interventional cardiologists to avoid the “oculo-radiolucent reflex,” which denotes the temptation to perform unwarranted angioplasty on intraluminal radioluency. Given the benign nature of WCA anomaly, most patients do not require revascularization and may be medically managed with antiplatelet therapy.

Bibliography


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