Arteries within the Artery of Coronary Artery in an Adult Patient with Acute Coronary Syndrome

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Abstract

A 40-year-old man with acute coronary syndrome underwent coronary angiography, which showed a somewhat irregular contour with radiolucent lines in the left anterior descending artery. Intravascular ultrasound disclosed that the arterial lumen was separated by confining walls, yielding multiple inner lumens. Implantation of drug eluting stents resulted in slow coronary run-off, which was restored soon after intra-aortic balloon pumping support. The multiple inner lumens correspond to the histopathological finding of “arteries within the artery”. While “arteries within the artery” is seen exclusively in children with a history of Kawasaki disease, it is rare in adults with undiagnosed Kawasaki disease.

Key words: arteries within the artery, Kawasaki disease, coronary vasculitis, percutaneous coronary intervention, intravascular ultrasound

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Introduction

“Arteries within the artery” is a histopathological finding characterized by multiple inner lumens in a coronary artery. The lesion, which is explained by thrombotic occlusion and spontaneous reperfusion sequence of a coronary artery, has been found exclusively in Kawasaki disease (KD), (1, 2). KD is associated with an acute vasculitis that mainly affects small and medium-sized arteries such as coronary artery. Coronary artery aneurysms or ectasia develop in 15% to 25% of untreated children with the disease, and may lead to rupture or thrombotic occlusion in the acute phase of illness (3). These coronary artery aneurysms disappear angiographically within a few years after onset in up to 60% of the cases. However, long-term follow-up of the remaining lesions occasionally demonstrates further abnormalities such as stenotic or obstructive lesions, collateral vessel formation, and recanalized thrombosis (4, 5). “Arteries within the artery” lesion is relevant to recanalized thrombosis (2). We report an adult case of acute coronary syndrome associated with arteries within the artery lesion which was depicted by intravascular ultrasound (IVUS) (6). Percutaneous coronary intervention (PCI) was performed by implantation of drug-eluting stents.

Case Report

A 40-year-old man came to our hospital because of vague chest discomfort lasting for about 10 minutes. An electrocardiogram showed QS pattern in leads V1-3 and T-wave inversion in leads V5-6, and echocardiography demonstrated hypokinesis of the left ventricular apex. However, his cardiac enzymes were negative. He was prescribed low-dose aspirin and transdermal nitroglycerin, but two days later, he was transferred to the hospital because of anterior chest oppression refractory to sublingual nitroglycerin. He had experienced similar symptoms at the age of 30, when coronary angiography showed normal coronary arteries (Fig. 1) with a negative acetylcholine provocation test and left ventriculogram revealed no contraction abnormalities. He had been smoking 25-30 cigarettes per day, and he stopped smoking shortly thereafter. He had no other coronary risk factors such as hypertension, obesity, diabetes, or hyperlipidemia.
The patient did not have an apparent history of KD in childhood. On physical examination, his heart rate was 75 beats / min and blood pressure was 138/88 mmHg. Auscultation revealed no cardiac murmurs or rales. Cardiac enzymes were normal, and troponin T-test was negative. No additional ST-T changes were observed on the electrocardiogram. Coronary angiography performed the day after admission revealed a somewhat irregular contour with radiolucent lines in the proximal to mid left anterior descending artery (LAD) (Fig. 2, upper panels). Both the circumflex and right coronary arteries were normal. Percutaneous coronary intervention was subsequently performed. Because a guidewire could not cross the LAD lesion, a stiffer wire with a microcatheter support was used. IVUS disclosed that the arterial lumen of the middle segment of LAD was separated by confining walls, yielding multiple lumens (Fig. 2, lower panels). The inner lumens, varying in size, gathered together toward the ostial LAD. The left main trunk was spared, and no aneurysmal dilatation was observed in the affected segments. The entire length of the lesion was predilated with a 1.5×15 mm balloon and a 2.5×14 mm balloon with good primary result (Fig. 3A). Then, three drug eluting stents (3.0×33 mm, 3.5×18 mm, and 3.5×33 mm) were deployed to fully cover the entire lesion. However, coronary angiography revealed slow coronary run-off of contrast medium and an unexpected obstruction of a large diagonal branch accompanied by chest pain and ST-segment elevation in precordial leads. Intra-aortic balloon pumping (IABP) was initiated through the left femoral artery. Prompt relief of chest symptoms and disappearance of ST-segment changes were obtained soon after IABP support, which was removed the next day. The maximum CK-level was 415 IU/L. Coronary angiography was performed one week later, demonstrating normal run-off of the contrast medium and reappearance of the diagonal branch (Fig. 3B). The ostium of the diagonal branch was successfully dilated through the stent struts with a 3.0×14 mm balloon. The patient was discharged without further complications and remained free from chest symptoms.

The present case is unique in that the coronary artery lesion was comprised of multiple inner lumens rather than an organic stenosis. The IVUS finding corresponds to the pathological finding of “arteries within the artery” which has been seen almost exclusively in children with a history of KD (1, 2). The “arteries within the artery” lesion is thought to be caused by the coronary artery occlusion-spontaneous reperfusion sequences (7). As for KD, coronary arteritis in the acute phase of illness and subsequent consequences may result in coronary thrombosis (5). The coronary arteritis is characterized by swelling of the endothelial cells, inflammatory cell infiltrate in the subendothelial space, medial necrosis, and disruption of the elastic laminae. After months, as the inflammatory infiltrate diminishes, intimal thickening occurs due to proliferation of migrated smooth muscle cells through the damaged internal elastic lamina. The destruction of elastic laminae weakens arterial wall and leads to coronary aneurysm formation. Otherwise, proliferation of the intimal smooth muscle cells may produce severe coronary stenosis over a period of months to years. In either condition, the lesion is susceptible to coronary thrombosis (7-9). The coronary thrombosis may recanalize spontaneously and develop multiple intracoronary channels, which are surrounded circularly by proliferative smooth muscle cells (7). It is speculated that these recanalization processes eventually give rise to the “arteries within the artery” lesion (1, 2).

If the “arteries within the artery” lesion in the present case is to be attributed to KD, there are some important aspects that need to be discussed. Whereas a history of KD was uncertain in the present patient, KD itself was firstly described by Tomisaku Kawasaki in 1967, that is, only 40 years prior to the present date (3). It is conceivable that a patient over 40 years of age was undiagnosed as KD in their childhood. The other point is that the patient had a normal coronary angiogram 10 years earlier, without having abnormalities suggestive of KD. However, regressed large coronary aneurysms due to KD can be apparently normal angiographically (4, 10). Moreover, it is proposed that coronary arteritis in the acute phase of KD can lead to vascular dysfunction later in life (10). Tsuda et al reported that acute coronary syndrome developed in two young adults with regressed coronary aneurysms and normal coronary angiograms (11). If the coronary thrombosis-spontaneous reperfusion sequence is not necessarily specific to KD, it remains to be validated whether other non-atherosclerotic vasculopathies causing coronary thrombosis can lead to “arteries within the artery” lesion.

The “arteries within the artery” lesion has been evaluated by coronary angiography, IVUS, and coronary CT angiography. Suzuki et al angiographically demonstrated multiple tortuous vessels located in the lumen of a native coronary artery. These findings, which developed within 2 years after the onset of KD, were designated as a “braid-like” le-

Figure 1. Coronary angiogram 10 years prior to the presentation showing normal coronary artery.
Figure 2. Upper panels: Coronary angiograms at presentation demonstrating somewhat irregular contour in the proximal to mid left anterior descending artery (LAD). An enlarged view shows some radiolucent lines in the coronary lumen. Lower panels: Intravascular ultrasound (IVUS) images revealing multiple inner lumens within the LAD. The finding is histopathologically comparable to “arteries within the artery”. The arrows indicate the corresponding coronary segments of the IVUS images. A: Proximal segment of the LAD. B: Middle segment of the LAD. Note that the inner lumens are converging toward the ostial LAD.

sion (12). Terashima et al (6) performed IVUS examination of the braid-like lesion in a 26-year-old man with a history of suspected KD. The IVUS imaging revealed what they call a “lotus root” appearance, in which the segment was composed of multiple channels. Longitudinal reconstruction of the IVUS images demonstrated that each of the channels was connected with a specific branch, such as septal branch, diagonal branch, and distal LAD (6). Recently, coronary CT angiography has emerged as the most accurate noninvasive method for detecting coronary artery disease. In adolescent patients with KD, a good correlation has been reported between coronary CT angiography and coronary angiography for the detection of coronary artery aneurysms, stenoses, and occlusions. The braid-like lesion also has been detected by coronary CT angiography as a multi-layered structure in a coronary artery (13). Taken together, both the angiographic braid-like lesion and the ultrasonographic lotus root appearance coincide with the multiple inner lumens as were observed in the present case, and all of them are comparable to the histopathological finding of “arteries within the artery” lesion.

This is the first report that a “arteries within the artery” lesion was treated by PCI with stent implantation. Catheter intervention is occasionally performed for stenotic lesions in children with KD. Coronary artery lesions of KD are much stiffer due to dense fibrosis and calcification than those of adult atherosclerosis. Percutaneous transluminal coronary rotational ablation will be required for severely calcified lesions if balloon angioplasty has not been successful even with high-pressure balloons (14). However, we have experienced some difficulties other than described above in treating the “arteries within the artery” lesion. Contrary to angiographic findings which showed only subtle wall irregularity, guide wire passage from proximal to the periphery was not feasible, because the guide wire easily strayed off into one of the multiple lumens. A stiff guide wire with a microcatheter support was required to penetrate back into the true lumen of the distal artery. Whereas balloon predilatation of
the target lesion was performed without difficulties, stent implantation was followed by coronary slow flow phenomenon and unexpected obstruction of a large diagonal branch, necessitating IABP support. Stent dilation may have caused distal embolization of fragmented thrombus and compressed other recanalized vessels from which arterial branches arise. Although less invasive, we have found these to be potential drawbacks in performing PCI for the arteries within the artery lesion. Coronary artery bypass surgery may as well be considered as an alternative therapy (15, 16). One important prerequisite is that the “arteries within the artery” lesion must be diagnosed correctly especially in adult patients prior to PCI or surgery. Irrespective of a history of KD, the finding such as multi-layered structure in coronary CT angiography or multiple intraluminal vessels in coronary angiography should be interpreted as the braid-like lesion. Furthermore, IVUS imaging should be applied to identify the lotus root appearance which corresponds histopathologically to the “arteries within the artery” lesion.

References


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