CASE REPORT

Spontaneous Coronary Artery Dissection Accompanied with Antiphospholipid Syndrome and Leukemia

Xiaofan Peng, MM, Zhaowei Zhu, MD, Jianjun Tang, MD and Shenghua Zhou, MD

Summary
We report a rare spontaneous coronary artery dissection (SCAD) case accompanied by antiphospholipid syndrome (APS) and leukemia which was treated successfully with drug-eluted stents (DES) implantation. This young SCAD patient was initially diagnosed of acute myocardial infarction (AMI); however, except for 6 pack-years of smoking, there were no risk factors or family history of coronary artery disease. Subsequently, we screened other clinical status like autoimmune diseases and finally found APS. In general, APS was associated with thromboembolism events, not coronary artery dissection. Our case indicated that SCAD could be a rare manifestation of APS which should draw our attention. In addition, our bail-out therapy acquired the expected effect.

Key words: Percutaneous coronary intervention, Intravascular ultrasound, Antithrombins

SCAD is defined as spontaneous coronary artery layers separation producing true and false lumen without human intervention, triggered primarily by intimal-luminal interface disruption or vasa vasorum rupture. It remains an infrequent clinical entity, predominantly affecting young women. Interestingly, our SCAD case was a very young man with APS and a 19 year history of diagnosed leukemia. To our knowledge, this is the first time a SCAD case accompanied by APS and leukemia has been reported.

Case Report
A 29-year old man who had no risk factors for coronary artery disease, except for 6 pack-years of smoking and a past medical history of diagnosed leukemia 19 years ago, presented to our cardiology department with severe chest pain for 13 days. The severe chest pain occurred 13 days ago while he was at rest, radiating to the left upper limb with profuse sweating. The symptom did not relieve until he received morphine and nitrates in the local hospital. The troponin I test was positive and ECG showed ST segment elevation in the anterior and lateral leads (Figure 1), which led to a diagnosis of AMI. Accordingly, primary percutaneous coronary intervention was suggested, but it was refused by the patient. Standard drug therapy with low molecular heparin, double antiplatelet, statin, and other secondary prevention medicines was applied at the local hospital. Then, based on his wish, the patient was transferred 13 days later with a stable condition to our hospital (regional heart center). Cardiac catheterization was performed, which showed type-D dissection from ostium to the mid segment of the left anterior descending artery (LAD) with contrast retention and TIMI flow grade 2; the other coronary arteries were smooth without any stenosis. The cholesterol level was within normal range. The LDL-c level was 57.28 mg/dL. Echocardiogram showed severe hypokinesia of the anterior wall and the apical segment of left ventricle. The left ventricular ejection fraction value was 48%. Considering the low risk factors or family history of heart diseases, we screened further for other clinical status like autoimmune diseases. The ENA test was negative, while the anticardiolipin IgG and IgM were positive.

Two drug-eluted stents were implanted, covering segment from mid-LAD to the left main coronary artery via the application of intravascular ultrasound (IVUS). The IVUS explicitly revealed intimal flap true and false lumen before stenting which doubtlessly demonstrate a typical SCAD (Figure 2). IVUS showed also the shrinkage of false lumen after stenting (Figure 3). We administrated metoprolol, benazepril, ticagrelor, and aspirin during hospitalization. Moreover, the dosage of warfarin was to strengthen the antithrombotic effect. The patient was discharged, after 7 days of treatment, with no occurrence of chest pain or any other discomfort.

Discussion
SCAD remains an infrequent clinical entity with the incidence ranging from 0.1% to 1.1% among the coronary angiography group, but is not as rare as we believed pre-
Previously, along with the prevalence of catheterization and intracavitary imaging technology, SCAD is becoming widely recognized as an important cause of acute coronary syndrome (ACS), accounting for 1.7-4.0% among ACS and 0.5% among sudden cardiac death patients. The underlying predisposing factors of SCAD include fibromuscular dysplasia, pregnancy, connective tissue disorder, systemic inflammatory disease, hormonal therapy, and physical exertion. Our case was a young man associated with APS and leukemia which is beyond the stereotyped impression of SCAD for young women.

Does APS or leukemia cause SCAD? As we all know, the physiopathologic feature of APS is recurrent thrombosis in arteries or veins not with reference to SCAD; however, it does cause SCAD theoretically. Firstly, APS can cause endothelial dysfunction. The existing evidence has suggested a widespread endothelial dysfunction in APS patient, while coronary endothelial dysfunction must play a major role in SCAD pathogenesis along with other factors such as increased coronary shear stress and localized vasculitis. Secondly, APS is an important manifestation of various systemic inflammatory diseases, accelerating the process of atherosclerosis. Therefore, atheromatous plaque rupture can produce a tunnel between blood flow and coronary artery layers, which can be enlarged by increased shear stress and, consequently, form SCAD. Finally, APS may be a contributor to the rupture of the vasa vasorum, since the existing evidence has proved chronic inflammation will lead to the dysfunction of coronary microvascular circulation. It is reasonable for us to believe APS may cause vasa vasorum rupture which is a potential mechanism for the initiation of coronary arterial wall separation. With regards to leukemia, it was reported leukocytes can exert vascular ef-

---

**Figure 1.** ECG showed ST segment elevation in I, AVL and V2-V4, ST segment depression and T waves inversion in II, III, AVF.

**Figure 2.** CAG and IVUS showed a type-D dissection from ostium to the mid segment of LAD before stenting. Dotted arrow showed atherosclerotic plaque in the left main trunk. Solid arrow showed false lumen of SCAD.
ffects by adhesion, aggregation, platelet recruitment, and affecting blood viscosity. Moreover, leukocytes can release proinflammatory factors, vasculotoxic factors, oxygen species, protease, and other factors which exacerbate the adverse vascular effect. However, our patient’s leukemia was effectively treated 19 years ago and remained stable for 11 years; therefore, we think leukemia contributed less to the occurrence of SCAD.

For the treatment of SCAD, we implanted two drug-eluted stents in the coronary artery based on the intensive antithrombotic therapy. Although, conservative treatment was proved to be non-inferior to percutaneous coronary intervention (PCI) or coronary artery bypass grafting (CABG), this SCAD patient had ST segment elevation and a dissection very close to the left main coronary artery; therefore, we unhesitatingly administrated invasive therapy which should have been performed in the local hospital to restrict the myocardial necrosis area. It was suggested the patient receive aspirin, ticagrelor, and warfarin for 6 months, followed by aspirin and warfarin for another 6 months. No chest pain or other cardiac events were reported, 10 months after his discharge.

Conclusion

APS can lead to SCAD and the treatments for this clinical status should be based on the intensive antithrombotic therapy. Although most SCAD cases can be treated with conservative therapy well, invasive treatments including PCI or CABG will lead to a better result if the vital coronary segments or blood flow were affected.

Disclosures

Conflicts of interest: None.

References

1. Saw J, Mancini GB, Humphries KH. Contemporary review on spontaneous coronary artery dissection. J Am Coll Cardiol 2016; 68: 297-312.
2. Koga S, Ikeda S, Nakata T, Maemura K. Spontaneous spiral dissection of left internal thoracic artery graft. Int Heart J 2015; 56: 360-2.
3. Alfonso F, Bastante T, Garcia-Guimaraes M, et al. Spontaneous coronary artery dissection: new insights into diagnosis and treatment. Coron Artery Dis 2016; 27: 696-706.
4. Rashid HN, Wong DT, Wijesekera H, et al. Incidence and characterisation of spontaneous coronary artery dissection as a cause of acute coronary syndrome—A single-centre Australian experience. Int J Cardiol 2016; 202: 336-8.
5. Nishiguchi T, Tanaka A, Ozaki Y, et al. Prevalence of spontaneous coronary artery dissection in patients with acute coronary syndrome. Eur Heart J Acute Cardiovasc Care 2016; 5: 263-70.
6. Hill SF, Sheppard MN. Non-atherosclerotic coronary artery disease associated with sudden cardiac death. Heart 2010; 96: 1119-25.
7. Alexanderson E, Cruz P, Vargas A, et al. Endothelial dysfunction in patients with antiphospholipid syndrome assessed with positron emission tomography. J Nucl Cardiol 2007; 14: 566-72.
8. Vega-Ostertag ME, Pierangeli SS. Mechanisms of aPL-mediated thrombosis: effects of aPL on endothelium and platelets. Curr Rheumatol Rep 2007; 9: 190-7.
9. Rajendra N, Lim F, Shaukat N. Spontaneous coronary artery dissection presenting as an ischaemic stroke in a middle-aged man with anti-cardiolipin antibodies: a case report. J Med Case Rep 2010; 4: 94.
10. Higashi Y. Assessment of endothelial function. History, methodological aspects, and clinical perspectives. Int Heart J 2015; 56: 125-34.
11. Durante A, Bronzato S. The increased cardiovascular risk in patients affected by autoimmune diseases: review of the various manifestations. J Clin Med Res 2015; 7: 379-84.
12. Recio-Mayoral A, Mason JC, Kasiki JC, Rubens MB, Harari OA, Camici PG. Chronic inflammation and coronary microvascular dysfunction in patients without risk factors for coronary artery disease. Eur Heart J 2009; 30: 1837-43.
13. Harlan JM, Winn RK. Leukocyte-endothelial interactions: clinical trials of anti-adhesion therapy. Crit Care Med 2002; 30: S214-9.
14. Charo IF, Taubman MB. Chemokines in the pathogenesis of vascular disease. Circ Res 2004; 95: 858-66.
15. Thiagarajan RR, Winn RK, Harlan JM. The role of leukocyte and endothelial adhesion molecules in ischemia-reperfusion in-
16. Tweet MS, Eleid MF, Best PJ, et al. Spontaneous coronary artery dissection: revascularization versus conservative therapy. Circ Cardiovasc Interv 2014; 7: 777-86.