A Rare Case of Triple Vessel Spontaneous Coronary Artery Dissection

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Introduction

- Spontaneous coronary artery dissection (SCAD) is a rare underdiagnosed phenomenon that presents as an acute, life threatening myocardial infarction, particularly in younger woman.
- Risk factors for SCAD include pregnancy, hormone therapy, stimulant drug use, connective tissue disorders and systemic inflammatory disorders.
- SCAD is defined as the separation of the coronary artery wall by hemorrhage with or without intimal tear. It causes acute coronary syndrome in 1.7% to 4% of cases.
- The actual prevalence of SCAD is difficult to quantify because, in some cases, it appears similar to atherosclerotic disease (Type 3 SCAD) by coronary angiography
- It is speculated that approximately 0.5% of myocardial infarctions are a result of SCAD. Some reports claim a prevalence of 25% in young females who experience myocardial infarction, particularly in pregnancy.
- The vast majority of reported cases are limited to a single vessel (84%) or two vessels (15%). We report a rare case of a young female patient with triple vessel SCAD presented after postpartum.

Case

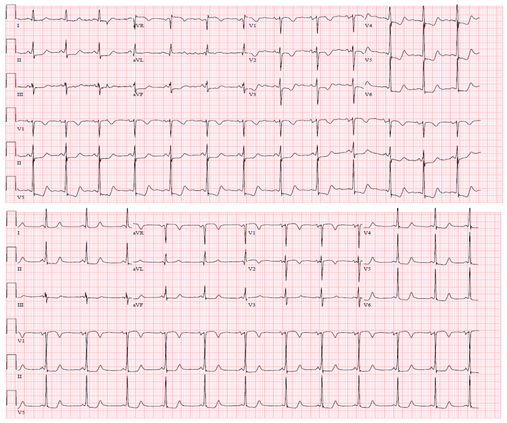
22 y/o woman, 2 weeks postpartum after her 4th pregnancy presented to emergency room with anginal chest pain. She did not have any known risk factors for atherosclerotic coronary artery disease.

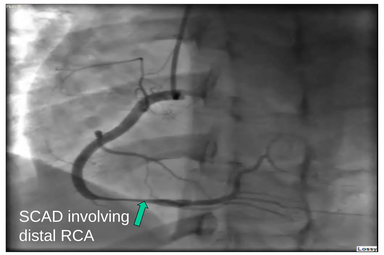
. EKG showed ST depressions in inferior & lateral leads and troponin was elevated to peak value of 4.932 (Normal Troponin I <0.040 ng/ml). Echocardiogram was consistent with mild decrease in LVEF (50%) without any focal wall motion abnormalities.

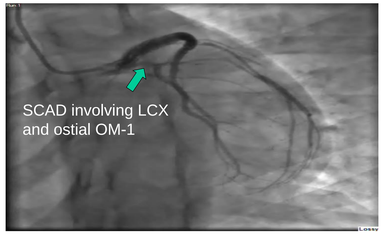
. Her coronary angiography revealed triple vessel SCAD involving distal LM extending to mid LAD, LCX and distal RCA. Further workup with CTA of the head/neck/abdomen revealed diffuse irregularity of vertebral arteries and intra cranial arteries, irregular caliber & focal wall thickening of common iliac arteries, and ectasia of the infrarenal abdominal aorta consistent with the diagnosis of Fibromuscular dysplasia (FMD).

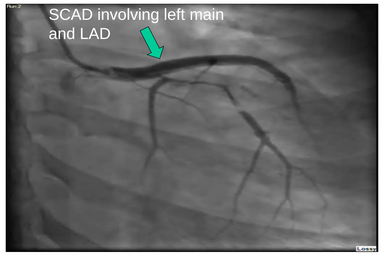
She was hemodynamically stable throughout, was managed conservatively with heparin during hospitalization, and was discharged home after 10 days on aspirin, plavix, metoprolol and captopril.

Images









Discussion

Three types of SCAD

- Type 1 is characterized by the appearance of a double lumen under contrast staining of the arterial wall.
- Type 2 appears as diffuse (typically 20 to 30 mm) and smooth narrowing that can vary in severity (due to intramural hematoma).
- Type 3 appears as focal or tubular stenosis that mimics atherosclerosis and can be confirmed only under intravascular visualization

Five angiographic features of SCAD

1. Absence of atheroma on other coronary arteries.
2. Radiolucent flap.
3. Contrast dye staining of the arterial wall;
4. Starting and/or ending of the angiographic ambiguity on a side branch; and
5. Long narrowing of the lumen caliber: smooth and linear, or stenosis of varying severity mimicking a “stick insect” or “radish” aspect.

Pregnancy and SCAD

- Peripartum-SCAD:
  - Highest incidence occurs during the postpartum period. Nearly all of the women diagnosed with P-SCAD presented within the 12 weeks following delivery, and the greater majority within one week postpartum.
- In United States, increased risk for P-SCAD was associated with
  - Pregnancy related risk factors: pre-eclampsia, prior treatment for infertility, multiparity.
  - Non-pregnancy related risk factors: Fibromuscular dysplasia, Antiphospholipid syndrome, Marfan’s syndrome, Connective tissue dysplasia, and Ehlers-Danlos syndrome.
- Mechanism
  - Unknown but possible that the physiological cardiac demands of pregnancy, hormonal changes in estrogen and progesterone, and the hemodynamic strains of labor and delivery contribute to the development of intimal wall tears and degeneration within coronary artery walls.
  - During pregnancy increased progesterone levels weaken the arterial wall by disrupting the normal corrugation of elastic fibers and degradation of medial wall collagen. In multiparity recurrent exposures to high levels of estrogen and progesterone can have additive effects to the degeneration of the arterial wall.

FMD and SCAD

- Fibromuscular dysplasia (FMD) is a nonatherosclerotic vascular disease that results in occlusion, aneurysm, or dissection of the artery. The most commonly affected vascular bed is the renal or carotid arteries . The exact frequency of FMD of coronary arteries is still unknown, as the diagnosis of coronary FMD is challenging.
- Genetic Link: Gene rs9349379-A allele of PHACTRI/EDN1 genetic locus (Chromosome 6q24) carries an increased risk of SCAD and FMD.

Discussion

Management of SCAD

- Most lesions heal spontaneously in 24-40 months per observational studies.
- Acute and sub-acute management of SCAD without ischemia is not well established.
- Therapy is aimed for support and symptom management. Includes heparin, beta blockers, calcium channel blockers, nitrates. Antiplatelet therapy is controversial.
- In patients where medical management fails or is not an option, PCI is an option for single vessel lesions. CABG is an option for Multivessel SCAD or complex lesions that fail PCI or are not amenable for PCI.

Conclusions

Multivessel SCAD is reported in a 22 y/o Female, 2 weeks postpartum, who presented with ACS.
Further workup revealed that patient meets criteria for FMD
Management of SCAD, particularly multivessel SCAD for postpartum patients, is not well established.

Clinical Relevance

- Multivessel SCAD should be suspected in young females with ACS, particularly in the postpartum period. Work up for FMD and genetic screening/counseling maybe warranted in these patients.

Disclosures

None

References

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