Case 14518

Eurorad

Aortitis as a cause of acute coronary syndrome

Published on 09.03.2017

DOI: 10.1594/EURORAD/CASE.14518
ISSN: 1563-4086
Section: Cardiovascular
Area of Interest: Vascular
Procedure: Diagnostic procedure
Imaging Technique: CT-Angiography
Imaging Technique: Catheter arteriography
Special Focus: Inflammation Case Type: Clinical Cases
Authors: Emad Moussa FRCR/MD; Mohamed Andron MD, CCT (UK) FRCP; Ahmed Munir MD; Atheer Al-Ansari FRCP (UK)
Patient: 50 years, female

Clinical History:

53-year-old lady with 2 years history of fever, malaise, and chest pain. She was investigated in another institution, started to receive steroids for management but discontinued her treatment course. Recently the nature of her chest pain had changed to severe tightness with ischaemic changes noted at ECG.
Imaging Findings:

CT coronary angiography revealed diffuse mural thickening of the aortic root and ascending aorta extending to the proximal level of the aortic arch with subtle extension to the origin of the right brachiocephalic trunk, while the remainder of the aortic arch and the other arch branches are normal. This mural thickening severely involves the ostial levels of the left main coronary and right coronary artery with near 90% stenosis of both ostia. Post-contrast delayed CT scans revealed swelling of the aortic intima and enhancement of the media and adventitia layers of the involved segment.

Interventional coronary angiography confirmed the findings with successful stent placement at both coronary ostia.
Discussion:

Aortitis is the pathological term for inflammation of the aortic wall. The classification of aortitis broadly includes underlying rheumatologic and infectious diseases, along with isolated aortitis. The most common rheumatologic causes of aortitis are the large-vessel vasculitides GCA and Takayasu arteritis, aortitis also is associated with systemic lupus erythematosus, rheumatoid arthritis, the HLA-B27 associated spondyloarthropathies, anti-neutrophil cytoplasmic antibody–associated vasculitides, Behçet disease, Cogan syndrome and sarcoidosis. Infectious causes include tuberculosis, syphilis, salmonella and other bacteria. Inflammatory aortitis also may occur in isolation (i.e., no associated common rheumatologic or infectious disease is present) or accompany idiopathic retroperitoneal fibrosis or an inflamed abdominal aortic aneurysm. [1]

In general, the epidemiology of Aortitis is poorly understood, the two most common aetiologies of aortitis, namely GCA and Takayasu disease, are much more common in the female population. GCA mean age of diagnosing is 75 years while Takayasu disease is a disease of young females under 50 years of age. [1, 2, 3]

Clinical presentation of aortitis is quite variable and includes symptoms of systemic illness due to the inflammatory

process itself and symptoms related to the involved segment of the aorta and the nature of the aortic lesions due to the inflammation as aortic branches stenosis, aneurysms, dissections, thrombosis and systemic hypertension. Generally, GCA commonly results in aneurysmal lesions while Takayasu disease mostly results in aortic branches stenosis with much less common presentations by aneurysm formations. [1, 4, 5]

According to the American College of Rheumatology Classifications, the diagnose of GCA requires 3 or more criteria including age at disease onset 50 years or more, new headache, temporal artery course tenderness, elevated ESR > 50 and abnormal temporal artery biopsy. The diagnosis of Takayasu disease also requires three or more criteria including age at disease onset <40 y, claudication of extremities, decreased brachial artery pulse, blood pressure difference between upper limbs >10 mm Hg, bruit over subclavian arteries or aorta and arteriogram abnormality. [1, 6]

CT angiography and MRA have widely replaced conventional angiography for the diagnosis of aortitis, especially due to their ability to identify wall lesions, complications as well as periaortic changes. The addition of PET scanner to cross-sectional imaging also aids widely to determine areas of disease activity. [1, 7]

Our case didn't fulfil the criteria of GCA or Takayasu disease, other rheumatic disorders were excluded and the possibility of infectious aortitis was also excluded, subsequently this cases was considered a case of isolated idiopathic aortitis.
Differential Diagnosis List: Isolated idiopathic aortitis leading to acute coronary syndrome, Giant Cell Arteritis (GCA), Takayasu arteritis, Aortitis associated with other rheumatic disorders, Infectious aortitis

Final Diagnosis: Isolated idiopathic aortitis leading to acute coronary syndrome

References:

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Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, Edworthy SM, Fauci AS, Leavitt RY, Lie JT, Lightfoot RW Jr, Masi AT, McShane DJ, Mills JA, Wallace SL, Zvaifler NJ. (1990) The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 33:1122–1128. (PMID: 2202311)
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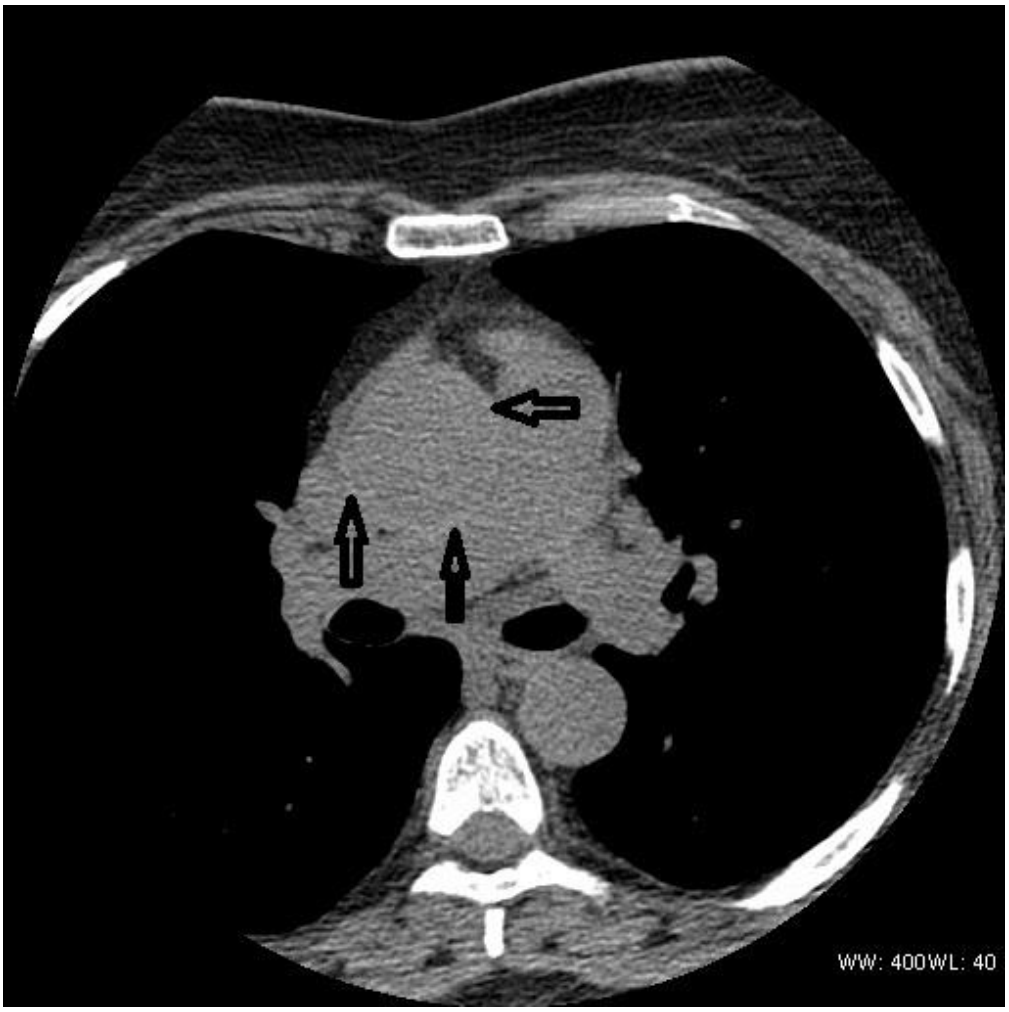
Figure 1

a



Description: CT pre contrast with diffuse mural thickening and increased density of ascending aorta walls (arrows). **Origin:** Radiology Department , Mediclinic Airport Road

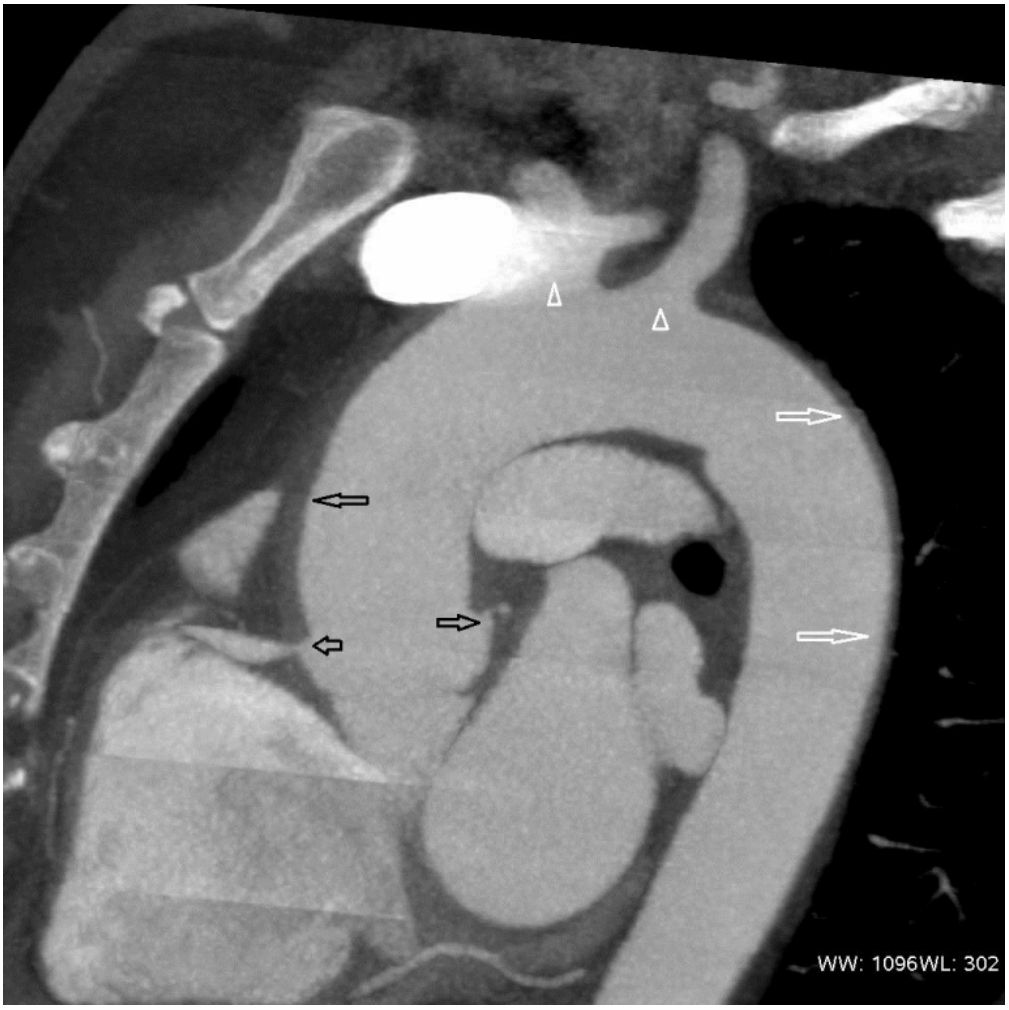
b

Description: Delayed enhancement CT with diffuse swollen intima and enhanced media and adventitia (arrows). **Origin:** Mediclinic Airport Hospital Abudhabi

c

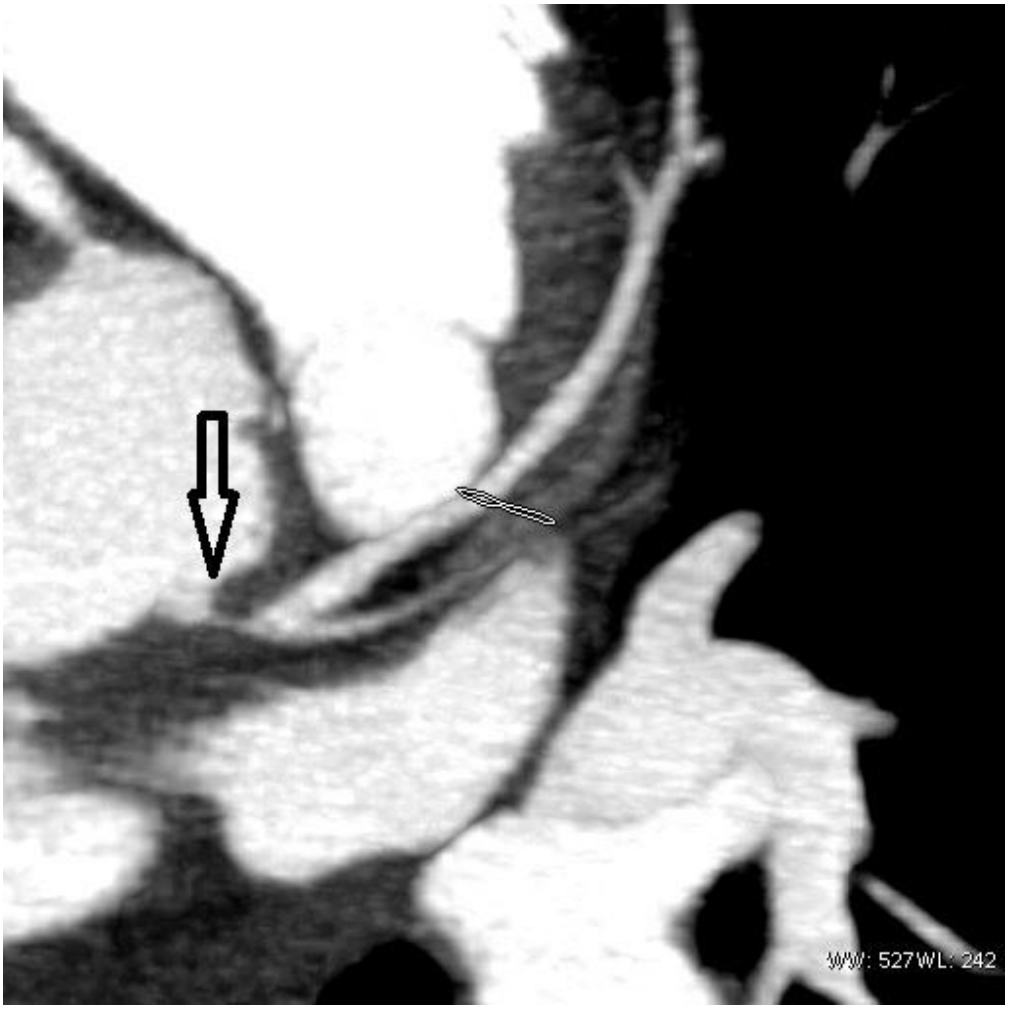
Description: Delayed enhancement CT coronal MPR of the ascending thoracic aorta with enhancement of the media and adventitia layers (arows). **Origin:** Mediclinic Airport hospital Abudhabi , UAE

d



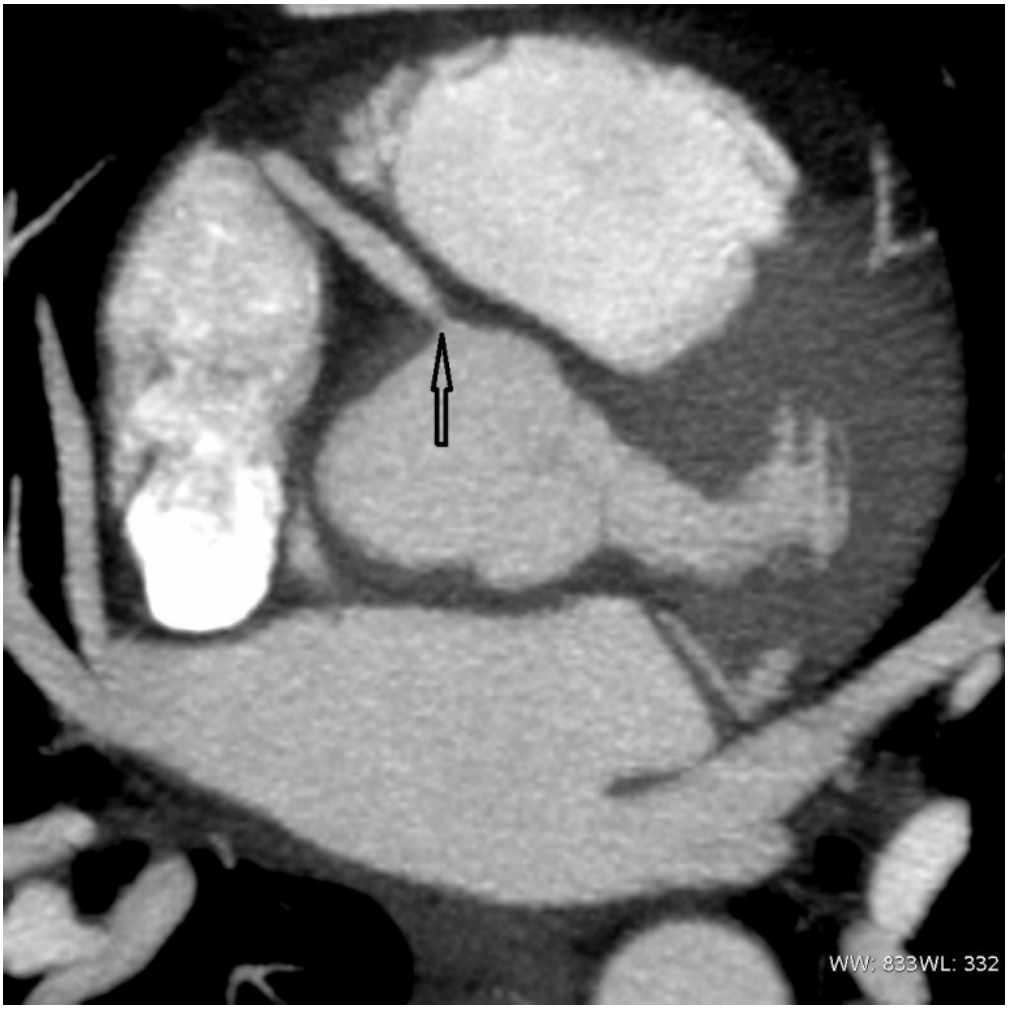
Description: MIP CT with mural thickening of the aortic root, narrowed coronary ostia (back arows), normal aortic arch main branches (white arrowheads) and normal remainder of the aorta (white arrows).
Origin: Mediclinic Airport Road Hospital , Abudhabi , UAE

e



Description: MPR of the left main coronary ostium with severe stenosis due to aortic wall thickening (arrow). **Origin:** Mediclinic Airport Hospital , Abudhabi , UAE

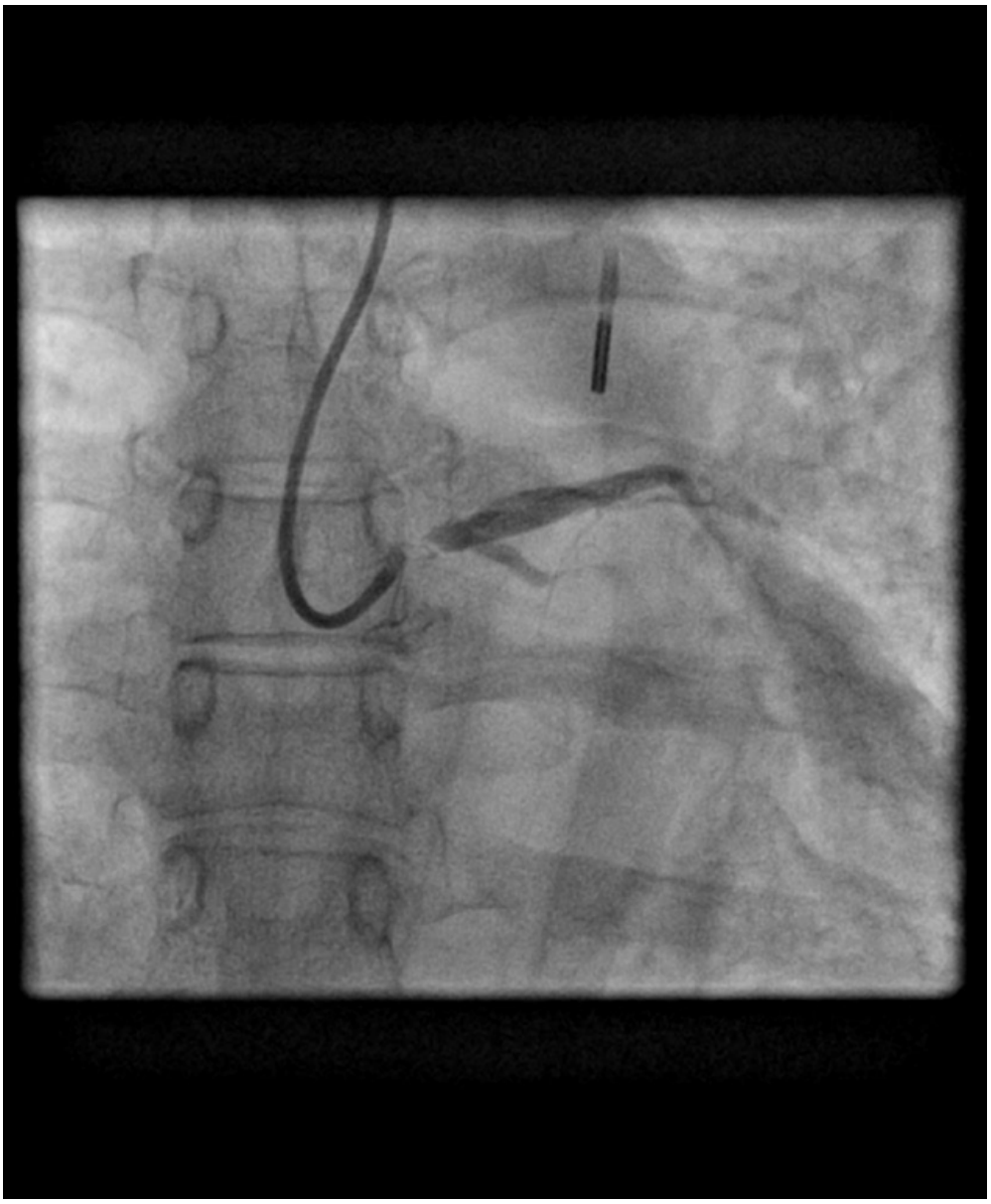
f



Description: CT MPR with RCA significant ostial stenosis due to aortic walls thickening (arrow). **Origin:** Mediclinic Airport Hospital , Abudhabi, UAE

Figure 2

a

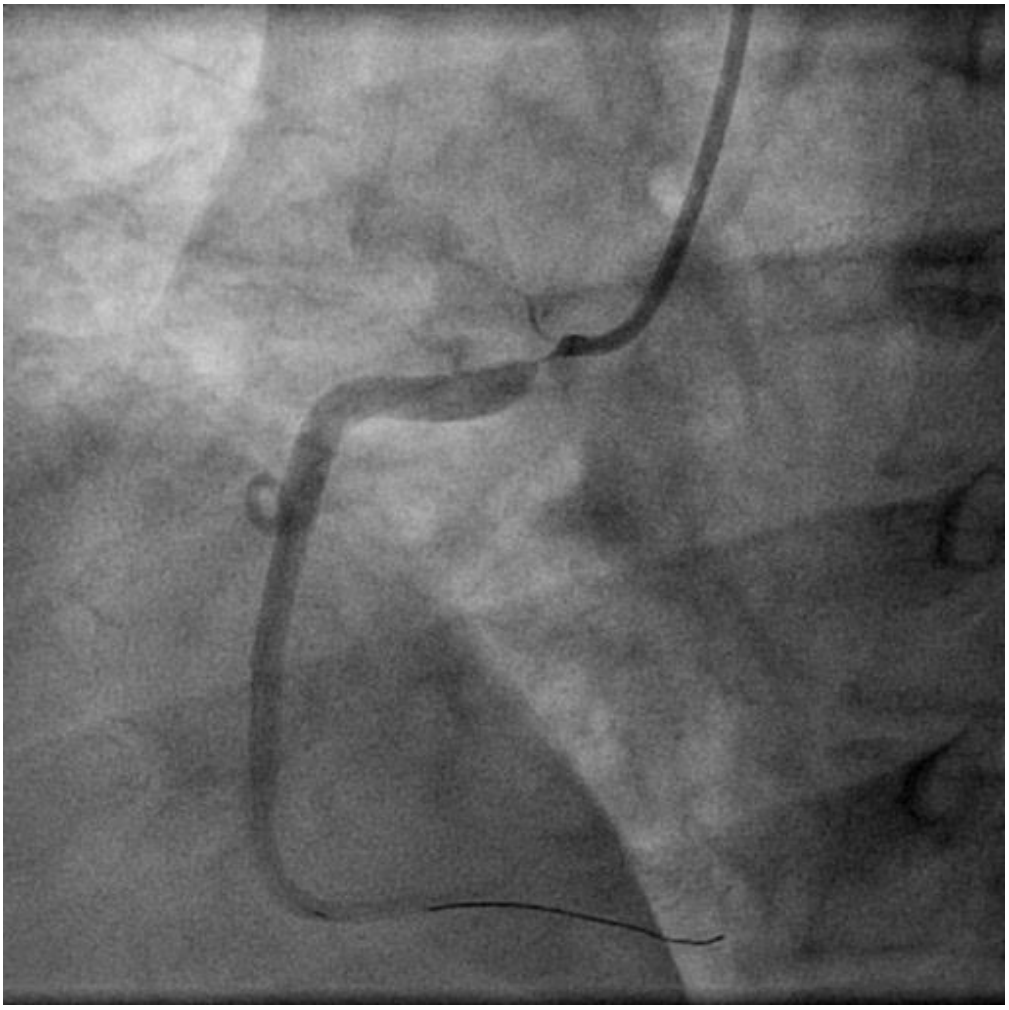


Description: Severe ostial stenosis of the LMA **Origin:** Mediclinic Airport Hospital , Abudhabi , UAE

b

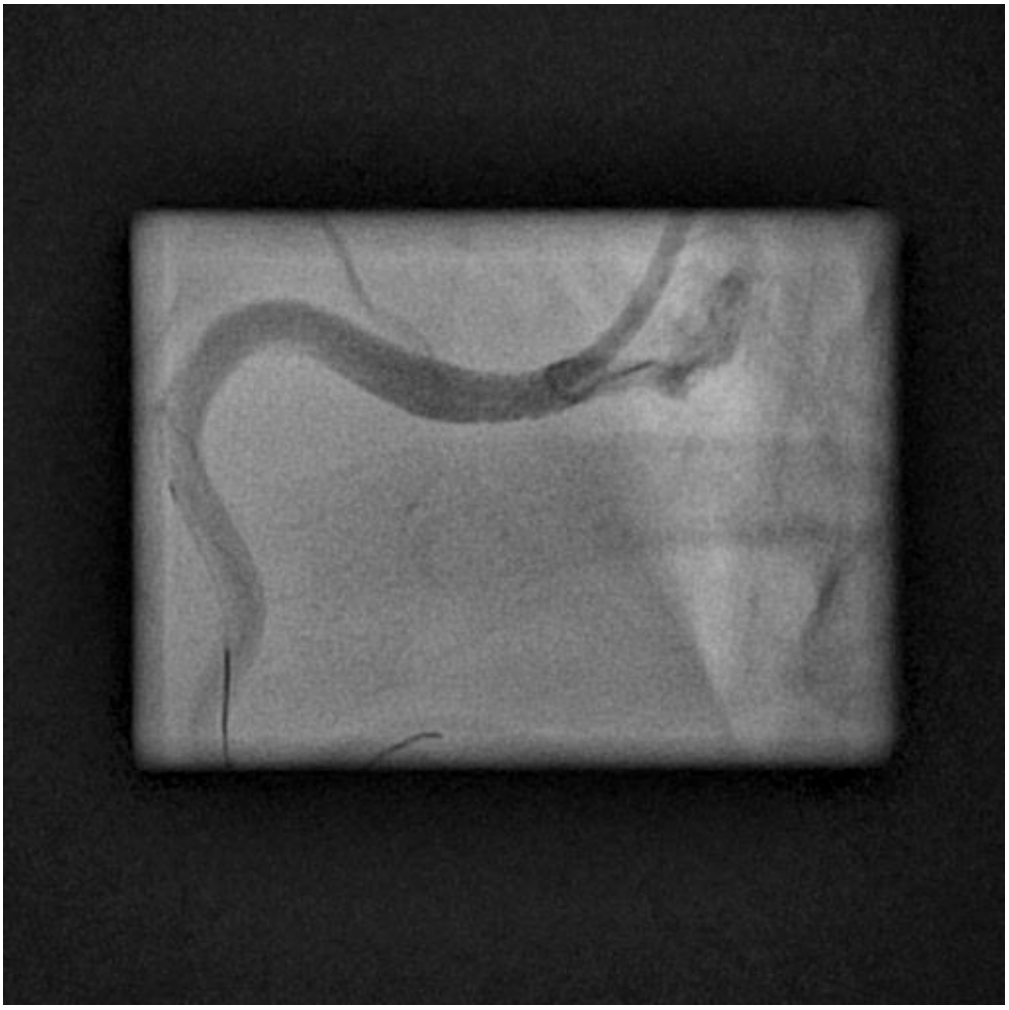
Description: Post LMA ostial stenting **Origin:** Mediclinic Airport Abudhabi , UAE

c



Description: Severe RCA ostial stenosis **Origin:** Mediclinic Airport Abudhabi , UAE

d



Description: RCA post ostial stenting **Origin:** Mediclinic Airport , Abudhabi , UAE