

Budd-Chiari syndrome and multiple coronary fistulae in a patient with Behçet's disease

Behçet hastasında Budd-Chiari sendromu ve çoklu koroner fistül

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A 32-year-old male patient with Behçet's Disease (BD) was consulted upon atypical chest pain. He had been diagnosed with BD 8 years ago (recurrent oral and genital ulcers, erythema nodosum, positive pathergy test, pulmonary embolism) and was treated with high-dose corticosteroids,

cyclophosphamide and colchicine. Due to abdominal swelling, abdominal sonography and computerized tomography that was performed showed findings compatible with Budd-Chiari syndrome (BCS) (Fig. A). Electrocardiography revealed sinus rhythm and no ischemic changes. Transthoracic echocardiography demonstrated normal left ventricular systolic function with normal segmentary wall motion and valvular functions. Multidetector computerized tomography coronary angiography revealed patent coronary arteries and multiple fistulae between the left coronary

system [both from left anterior descending (LAD) and circumflex (Cx) arteries] and the pulmonary artery (Figs. B-D). Also, conventional coronary angiography confirmed the multiple coronary to pulmonary artery fistulae (Figs. E-G, Video*). Because of the atypical symptoms and small fistulae diameter, myocardial perfusion scintigraphy was performed and showed no reversible perfusion defect. Eventually, the patient discharged uneventfully from the hospital with aspirin, warfarin, methylprednisolone, colchicine and trimetazidine treatment. During the 18 month follow-up visit, he was asymptomatic with a normal exercise test.



Figures– (A) Abdominal computerized tomography showing occluded inferior vena cava. Multidetector computerized coronary angiography revealed patent coronary arteries; **(B)** right coronary artery, **(C)** left anterior descending (LAD) artery, **(D)** circumflex (Cx) artery. **(E)** LAD to pulmonary artery fistula was demonstrated in MDCT coronary angiography. Conventional coronary angiography confirmed the diagnosis of LAD and Cx arteries to pulmonary artery fistulae **(F)** left oblique view, **(G)** postero-anterior caudal view (stars). *Supplementary video file associated with this case can be found in the online version.

