

## ANJİYOGRAFİDE ABARTILI NEOVASKÜLARİTE İLE KENDİNİ GÖSTEREN VE SOL ATRİYUMA DRENAJ YERİ ATİPİK OLAN BİR KORONER-KAMERAL FİSTÜL OLGUSU

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### ÖZET

**GİRİŞ:** Bir koroner arterle bir kalp boşluğu arasında miyokardiyal kapiller yatağı köprüleyerek oluşmuş belirgin bağlantı koroner-kameral fistül olarak tanımlanır. Mitral darlık hastalarında, sirkumfleks koroner arterden köken alan fistül ve koroner revaskülarizasyon varlığı, sol atriyal trombüsün bir belirtisidir.

**OLGU:** 51 yaşında eski bir maden işçisi olan erkek hasta hastanemize nefes darlığı ve atipik göğüs ağrısı yakınmaları ile başvurdu. Transtorasik ekokardiyografide mitral darlık ve sol atriyumda trombüs tespit edildi. Transözefajeal ekokardiyografide trombüsün sol atriyum apendiksinden köken alarak interatriyal septum ve atriyum tabanına doğru uzanım gösterdiği gözlemlendi. Koroner anjiyografide sirkumfleks koroner arterden çıkan ve sol atriyum içindeki kitle lezyonuna drene olan yaygın damar ağı görüldü. Hastaya mitral kapak ve koroner-kameral fistülün kapatılmasına yönelik cerrahi planlandı. Operasyonda, atriyum tabanından başlayarak sağ pulmoner ven ağız seviyesinden interatriyal septum ve mitral kapak anulüsü seviyesine kadar olan mesafeyi kaplayan dev trombüs saptandı. Trombüs dikkatli bir şekilde atriyum duvarından diseke edilerek ayrıldı. Trombüsün altında posterior mitral anulüsten yaklaşık 2-2,5 cm mesafede –süperior pulmoner venler ve atriyal apendiks arasındaki hattın orta noktasında– yerleşimli olan koroner-kameral fistül ostiumu bulundu. Ostium primer olarak sütüre edildi ve mitral kapak replase edildi.

**SONUÇ:** Mitral darlık olgularında sol atriyal trombüse eşlik eden koroner-kameral fistüller değişik şekillerde ortaya çıkabilmektedir. Bu tip koroner-kameral fistüller en sık atriyal apendikse drene olmakla birlikte, fistülün açıldığı yerin kesin olarak saptanabilmesi için sol atriyumun diğer bölgeleri de dikkatli bir şekilde incelenmelidir.

**Anahtar sözcükler:** Koroner-kameral fistül, sol atriyal thrombus, mitral stenoz

### A Coronary Cameral Fistula Displaying Exaggerated Neovascularity on Angiogram with Atypical Drainage Site in Left Atrium

#### SUMMARY

**OBJECTIVE:** The term coronary cameral fistula describes a sizeable communication between a coronary artery and a chamber of the heart that bypasses the myocardial capillary bed. In patients with mitral stenosis, coronary revascularization and fistula originating from the circumflex coronary branch is an indicator of left atrial thrombus.

**CASE:** A 51-year-old male who was a former coal miner admitted to our hospital with complaints of shortness of breath and atypical chest pain. Transthoracic echocardiography revealed mitral stenosis and left atrial thrombus. Transesophageal echocardiography showed that the thrombus was spreading out from the left atrial appendix towards the interatrial septum and the base of atrium. Coronary angiogram revealed extensive network of vessels arising from the circumflex coronary artery draining into a solid lesion originating from left atrium. He was scheduled to undergo mitral valve surgery and surgical closure of the coronary cameral fistula. In the operation, giant thrombus extending from the base of atrium to the origin of the right pulmonary vein and down to the interatrial septum and mitral valve annulus was detected. Thrombus was carefully dissected and peeled off from the left atrial wall. Underneath the thrombus –situated at the center of the line between the superior pulmonary veins and atrial appendage– approximately 2-2,5 cm. distant from the posterior mitral annulus, ostium of the coronary cameral fistula was found. The ostium was closed by simple sutures and the mitral valve was replaced.

**CONCLUSION:** Coronary cameral fistulas accompanying left atrial thrombus in cases of mitral stenosis may emerge in variable forms. Although the most common drainage site for these fistulas is atrial appendage, the other regions of left atrium should be carefully inspected for exactly localizing the ostium of the fistula.

**Key words:** Coronary cameral fistula; Left atrial thrombus; Mitral stenosis

The term coronary cameral fistula describes a sizeable communication between a coronary artery and a chamber of the heart that bypasses the myocardial capillary bed<sup>1</sup>. Although these fistulas are most likely congenital in origin and usually remain asymptomatic<sup>2,3</sup>, acquired fistula secondary to trauma or invasive cardiac procedures have also been reported<sup>4</sup>. Coronary cameral fistula accounts for 0,08-0,4% of congenital cardiac anomalies and are observed in 0,1% of patients undergoing coronary

angiogram<sup>5,6</sup>.

Coronary revascularization and fistula originating from the circumflex coronary branch is an indicator of left atrial thrombus in mitral stenosis<sup>7,8</sup>. Typical angiographic finding for such a thrombus is the collection of tiny vessels coursing through left atrial appendage which terminate in a fashion of network and form a “blush” of contrast media. This arteriographic finding is reported to be 72% sensitive in detection of left atrial thrombus and is particularly

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useful in patients with previous embolic events or failure of echocardiography in diagnosing thrombus<sup>9</sup>. In this paper, we present a case of coronary cameral fistula with atypical drainage site in left atrium and co-existing giant left atrial thrombus presenting with extensive neovascularisation on coronary angiogram.

## CASE REPORT

A 51-year-old male who was a former coal miner applied to a chest physician with complaints of shortness of breath and atypical chest pain of one month duration. Physical examination revealed 2/6<sup>o</sup> diastolic murmur near the apical border and chest radiography demonstrated cardiomegaly. Atrial fibrillation was observed on electrocardiogram. Transthoracic echocardiography revealed mitral stenosis with a valvular area of 1cm<sup>2</sup>, mild mitral insufficiency, pulmonary hypertension and a hyper-echogenic mass in the left atrial cavity. Thereafter, transesophageal echocardiography was performed and spontaneous contrast in the left atrium which is an indicator of thrombus was detected. The thrombus was observed to occupy the left atrial cavity from the border of left atrial appendix to interatrial septum and the base of atrium (Fig.1). The patient underwent coronary angiography and catheterization as to clarify the coronary anatomy. Coronary angiogram revealed extensive network of vessels arising from the circumflex coronary artery which drain into the left atrial thrombus. The angiographic appearance was similar with that of a tumor with neovascularisation (Fig.2).

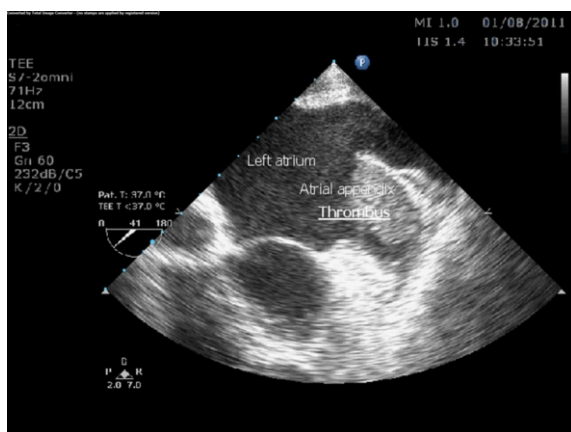


Figure 1. TEE image of the thrombus in the left atrium.

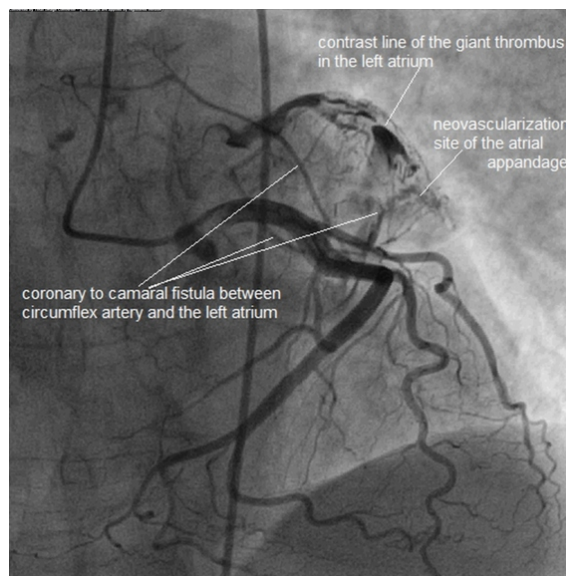


Figure 2. Angiographic appearance of left atrial thrombus and coronary cameral fistula. Giant thrombus surrounded by numerous vascular structures mimics a tumoral lesion.

He was scheduled to undergo mitral valve surgery and surgical closure of the coronary cameral fistula. Median sternotomy was performed. Standard aortic and bicaval venous cannulation was used to institute cardiopulmonary bypass. The aorta was cross clamped and heart was arrested by means of antegrade cardioplegia. Then, left atrium was opened and the giant thrombus extending from the base of atrium to the orifice of the right pulmonary vein and towards the interatrial septum and mitral valve annulus was exposed. Thrombus was observed to be well organized and strictly adherent to the left atrial wall. It was carefully dissected and peeled off from the left atrial wall. The ostium of the fistula was located underneath the thrombus –situated at the center of the line between the superior pulmonary veins and atrial appendage– and approximately 2-2,5 cm. distant from the posterior mitral annulus. It was 0,4-0,5 cm. in size and closed by simple sutures. Mitral valve was replaced with 31mm. St. Jude mechanical prosthesis. The postoperative course was uneventful and the patient was discharged on the seventh postoperative day.

## DISCUSSION

A direct communication between a coronary artery and a cardiac chamber is described as a coronary cameral fistula. The feeding artery of the fistula may drain from a main coronary artery or one of its branches and is usually a dilated and tortuous artery terminating in one of the cardiac chambers<sup>2</sup>. These fistulas commonly originate from the right coronary artery (55%) and in 35% of cases from the left coronary artery<sup>4</sup>. The right side of the heart is the major drainage site and right ventricle is the most

frequent receiving chamber (45%) followed by right atrium (25%). Rarely, the drainage site may be coronary sinus or pulmonary vasculature<sup>10,11</sup>. Most of the coronary artery fistulas are small, asymptomatic and are usually undiagnosed until a coronary arteriography is performed. However the larger ones may lead to myocardial steal phenomenon and ischemia of the myocardial segment which is perfused by the involved coronary artery<sup>12</sup>. In this case, aim of the treatment is the occlusion of the fistula, whilst preserving normal coronary blood flow. Occlusion of the fistula can be performed via catheter techniques or surgery. Surgery involves internal closure of the fistula within the receiving chamber whenever feasible<sup>2</sup>.

Left atrial thrombus is frequently encountered in mitral stenosis and may be along with a coronary cameral fistula. The fistula arising from the circumflex branch to left atrial appendix in association with neovascularisation can be detected in coronary angiography. This neovascularisation is typically observed as a collection of small vessels which branch to form a network of vascular channels and finally terminate in a lacunar pattern. Such a pooling of vessels yields the image of a blush of contrast medium and this is considered as a positive predictor of thrombus. However, localization of the exact drainage site of fistula in the left atrium by coronary angiography is difficult.

The angiographic appearance of neovascularisation and position of the fistula drainage site in the left atrium is quite atypical in this case. The giant left atrial thrombus surrounded by numerous exaggerated vessels and an accompanying coronary cameral fistula mimics a tumoral lesion with feeding vessels from the adjacent tissues. This angiographic appearance is not similar with tiny vascularity accompanying a left atrial thrombus. The fistula arising from the circumflex branch is usually drained to appendage in majority of the cases with left atrial thrombus, however in this case the drainage site is the base of left atrium.

In conclusion, this case is an example of extensive and exaggerated neovascularisation associated with a coronary cameral fistula and left atrial thrombus. This type of vascularity established on coronary angiogram may recall a tumor; however it can be a marker of thrombus. Although the most common drainage site for coronary cameral fistula in patients with mitral stenosis and left atrium thrombus is atrial appendix, the other regions of left atrium should be carefully inspected for exactly localizing the ostium of the fistula.

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