

A Catastrophic Picture; Recurrent and Multiple Embolisms of Left Atrial Myxoma: Case Report

Katastrofik Bir Tablo: Sol Atriyal Miksoma Kökenli Multipl ve Rekürren Emboliler

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ABSTRACT This report describes an unusual case of extensive multiple peripheral embolisms caused by a left atrial myxoma involving cerebral arteries, abdominal aorta and its branches in a 46-year-old patient. Embolism of left atrial myxoma is an uncommon but a well-known cause of peripheral and cerebral ischemia. Distal embolization is often the first presentation in most cases, with the central nervous system being the most common site. Distal embolization generally results from tumor fragmentation, or, less often, from complete tumor detachment, causing syncope, dyspnea, neurologic symptoms, or ischemic limb pain. Myxomas might give rise to embolism large enough to cause vascular occlusion, as in this case. This is the first case experienced initial peripheral embolism followed simultaneously by subsequent cerebral and abdominal/renal arterial embolisms originating from left atrial myxoma. Diagnosis was previously based on the transthoracic echocardiography and confirmed with histopathological evaluation of the resected material. We present a very interesting case of a myxoma leading to multiple embolisms concurrently with a catastrophic consequence despite aggressive attempts.

Key Words: Heart atria; myxoma; embolism

ÖZET Bu yazıda, 46 yaşındaki bir hastada sol atriyumdaki bir miksoma kaynaklı serebral arterler, abdominal aorta ve dallarının da dahil olduğu geniş bir alanda multipl periferik embolilerin gözleendiği, nadir görülen bir tabloyu sunuyoruz. Sol atriyal miksoma sık görülmemekle birlikte serebral ve periferik iskeminin iyi bilinen nedenlerinden birisidir. Çoğu olguda periferik embolizasyon ilk bulgu, santral sinir sistemi tutulumu ise en siktir. Distal embolizasyon genelde tümör fragmentasyonu ile, daha seyrek olarak da tümör dokusunun bütün olarak duvardan kopması sonucu ortaya çıkarak senkop, dispne, nörolojik semptomlar, ekstremitelerde iskemik ağrıya neden olabilir. Miksomalar, bu olguda olduğu gibi, vasküler oklüzyona neden olabilecek kadar büyük embolilere yol açabilirler. Bu sol atriyal miksoma olgusu, başlangıçtaki periferik emboliden hemen sonra serebral ve abdominal/renal arteriyel embolilerin eş zamanlı gözleendiği ilk örnektir. Tanı, transtorasik ekokardiyografi ile konarak rezeksiyon materyelinin histopatolojik incelemesi sonucu doğrulanmıştır. Biz burada birden çok emboliye yol açarak agresif girişimlere rağmen katastrofik bir gidiş izleyen oldukça ilginç bir sol atriyal miksoma olgusu sunuyoruz.

Anahtar Kelimeler: Kalp atria; miksoma; embolizm

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Cardiac myxomas are an uncommon but well-described cause of peripheral embolization and stroke in younger individuals. Cardiac myxomas are the most common benign tumor of the heart in adults and represent approximately 83% of primary cardiac tumors.¹ Cardiac myxomas arise from the endocardium and can occur in any of the cardiac cham-

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bers, but up to 75% to 80% of them occur in the left atrium, near the fossa ovalis. The mean age of presentation has been reported between 30 and 70 years.² There is a female-to-male predominance reported between 3:2 and 2:1. The classic triad of presenting symptoms includes obstructive cardiac signs,¹ embolic signs,² and constitutional or systemic manifestations.³ Clinically apparent cerebral emboli have been reported in 25% of the cases.³

CASE REPORT

A 46-year-old male patient was referred to our Emergency Department suffering from sudden onset of bilateral leg pain, muscle weakness, and foot cyanosis. He had a previous stroke history of four years ago. On physical examination, the patient was conscious, blood pressure was 150/90 mmHg, and the pulse was regular with rate of 80 per minute. Systolic or diastolic murmur was not heard at the apex of the heart. Peripheral pulse examination revealed normal carotid and upper extremity arterial pulsations while the left and the right femoral arterial pulses were absent. Both lower extremities were cold and cyanotic. The patient was diagnosed with acute arterial ischemia and emergent embolectomy was conducted. To avoid further delay of treatment, preprocedural imaging was not performed.

A gelatinous-like material was removed from both femoral and iliac arteries with Fogarty balloon catheters. Macroscopic examination of the embolectomy material showed pinkish-red colored, shapeless gelatinous clot-like tissue fragments. Following bilateral embolectomies, all peripheral pulses of lower extremities were restored and the symptoms disappeared.

After the embolectomy, a computed tomography (CT) angiogram of both lower legs was taken. The angiogram revealed a good runoff in the right femoral and iliac arteries. However, the proximal left superficial femoral artery was totally occluded, most likely previously by the chronic embolic disease. In addition to this, extensive collateral circulation originated from the developed left deep femoral artery towards the left popliteal artery, with a good distal runoff.

A prompt echocardiogram taken after the surgery for determining the origin of embolism showed incidentally an echodense spherical, mobile homogenous mass (4.5 x 4.0 cm) originating from the interatrial septum in the left atrium (Figure 1A). The tumor had irregular surface and had a peduncle that allowed it swing during the cardiac cycle. Echocardiographic evaluation led us to consider the diagnosis of left atrial myxoma. Overall, left ventricular functions and valvular functions were preserved. Left atrial mass causing emboly forced us immediately to resect it under cardiopulmonary bypass. While preparing the patient to operation, he suddenly lost his consciousness and had a weak withdrawal on the right side. Cerebral embolism was considered and medical therapy was initiated. The initial cerebral CT imaging was negative for positive findings. There was no evidence of early radiological signs of ischemia such as dense middle cerebral artery (MCA) sign or loss of gray-white differentiation. An abdominal and pelvic CT angiogram obtained before the surgery demonstrated a saddle embolus totally occluding abdominal aorta with infrarenal involvement distally to the bilateral iliac arteries (Figure 1B). Additionally, a large infarcted area in the lower pole of the left kidney and small infarcted areas in the right kidney were also observed (Figure 2). Final evaluation of the patient revealed simultaneous formation of cerebral and abdominal/renal arterial embolisms stemming from left atrial myxoma.

After improvement of the patient's clinical condition, he was operated under cardiopulmonary bypass. The mass in the left atrium was totally ex-



FIGURE 1: (A) Echocardiographic examination shows an intracardiac mass originating from the interatrial septum (white arrow). (B) Sagittal reconstruction of CT angiogram of the abdomen demonstrates complete occlusion of the infrarenal aorta (white arrow).



FIGURE 2: Coronal reconstruction of CT shows a large infarct in the left kidney (white arrow) and multiple small infarcted areas in the right kidney.

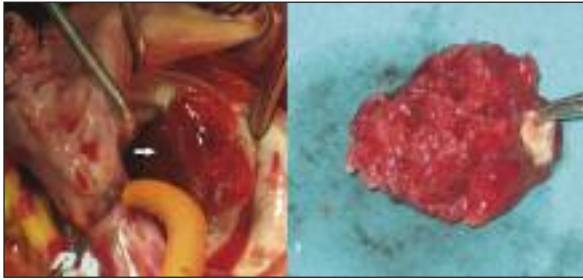


FIGURE 3: (A) Intraoperative appearance of the mass (white arrow). (B) Gross appearance of the mass excised with the interatrial septal tissue involved.

cised (Figure 3A). The resected mass had a fragile smooth surface and pinkish-red color (Figure 3B). Pathologic examination of the mass was compatible with cardiac myxoma. After median abdominal incision, synchronous direct embolectomy with an arteriotomy for bilateral renal arteries and infrarenal aortic embolectomy via femoral arteries were performed in the surgery. A great amount of embolus was removed from the left renal artery and aorta. After the operation, the patient had palpable peripheral pulses in his both legs.

After the operation the patient ameliorated initially, however despite close follow-up he progressively deteriorated. A cerebral CT scan repeated 16 days afterwards the surgery demonstrated a big left hemispheric subacute infarct (Figure 4). The patient's neurologic status did not improve, and he passed away 21 days after the surgery.

DISCUSSION

We have presented here an unique case of the simultaneous formation of cerebral embolism along with abdominal aortic/bilateral renal arterial embolisms originating from a cardiac myxoma.

Cardiac myxomas most commonly arise in the left atrium (80%), with 15% to 20% occurring in the right atrium, and 4% in the ventricles; approximately 5% of patients have multiple myxomas.⁴ Embolic phenomenon in cardiac myxoma is common, with the incidence ranging from 30% to 40%.⁵ The emboli generally display a predilection for the central nervous system,⁶ but can also affect other organs such as the liver, spleen, kidney, retina, coronary vessels, and distal arterial tree.⁵ In a retrospective review of 112 patients by Pinede et al.,⁷ it was found that the most common initial presenting symptoms of atrial myxoma were related to mitral valve obstruction; however, 33 patients presented with signs of embolism, including 24 to the central nervous system.² The other studies suggest that atrial myxoma occurs in 0.5% of acute stroke patients, women in the fifth decade being at greatest risk.⁸

The central nervous system is one of the most susceptible areas of embolization, resulting in multiple ischemic strokes, and at times, independent metastatic growth occurs in the brain.³ Several other reports describe symptoms of cerebellar infarction, multiple cerebral infarcts, and even fulminant brain necrosis after multiple myxoma embolism. In addition, myxoma emboli may manifest as systemic or cerebral metastases even years after removal of the cardiac tumor and pulmonary hypertension when the tumor originates from the right atrium.²

Diagnosis is usually made with echocardiography. The sensitivity of transthoracic echocardiography is 95%, compared with 100% for transesophageal echocardiography. Magnetic reso-



FIGURE 4: CT with a massive large hypodense area indicating subacute infarct in the left cerebral hemisphere (White arrow).

nance imaging may be useful to help determine tumor characteristics and composition.¹ When found, left atrial myxomas should be surgically removed in a timely fashion to prevent emboli or other complications. Surgery carries a relatively low morbidity of less than 5%, and is usually curative.²

CONCLUSION

This case presented a cardiac myxoma partially detached from the left atrial septum and caused aortic saddle embolization, renal infarction and cerebral infarction. As far as is known, this appears rarely.

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