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Total Occlusion of the Infrarenal Aorta by Cardiac Myxoma: Emergent Surgical Management

İnfrarenal Aortun Kardiak Miksoma Nedeniyle Total Oklüzyonu: Acil Cerrahi Tedavi

ABSTRACT

Cardiac myxomas are the most common primary tumors of the heart and can cause systemic embolization, particularly when located in the left atrium. We present a rare case of a left atrial myxoma resulting in total occlusion of the infrarenal abdominal aorta. A 60-year-old man was admitted to the hospital with back pain, paresthesia in the lower extremities, and subsequent development of paraplegia. Computed tomography revealed total occlusion of the infrarenal aorta. Emergent surgery was performed, and the intraoperative specimen removed from the aorta was histologically identified as myxomatous material. Transthoracic echocardiography revealed a mass located in the left atrium. Despite a second surgery for the removal of the cardiac mass, embolic episodes caused by the myxoma could not be prevented. Prompt diagnosis and timely treatment are essential to improve clinical outcomes by preventing embolization-related complications.

Keywords: Abdominal aorta occlusion, atrial myxoma, cardiac mass, thromboembolism

ÖZET

Kardiyak miksomalar en sık görülen primer kalp tümörleridir ve sol atrial yerleşimli olduklarında sıklıkla sistemik emboliye neden olurlar. İnfrarenal abdominal aortun total tıkanıklığına neden olan nadir bir sol atriyal miksoma olgusunu sunuyoruz. 60 yaşında erkek hasta sırt ağrısı, alt ekstremitede parestezi ve ardından gelişen parapleji nedeniyle hastaneye başvurdu. Bilgisayarlı tomografide infrarenal aortun total oklüzyonu görüldü. Hasta acil cerrahiye alındı. Aorttan intraoperatif alınan örnegin histolojik olarak miksomatöz benzeri bir materyal olduğu tespit edildi. Hastaya yapılan transtorasik ekokardiyografide sol atriyum yerleşimli kitle saptandı. Kalp içi kitlenin çıkarıldığı ikinci cerrahiye ragmen, miksoma kaynaklı emboli atakları önlenemedi. Hızlı tanı ve dolayısıyla hızlı tedavi, embolizasyon kaynaklı komplikasyonları önleyerek klinik sonuçları iyileştirebilir.

Anahtar Kelimeler: Abdominal aort oklüzyonu, atriyal miksoma, kardiyak kitle, tromboembolizasyon

Myxomas are the most common primary tumors of the heart. Although their location can vary, they are most frequently found in the left atrium, typically attached to the atrial septum, with an incidence of 70–90%. It has also been reported in the literature that approximately 20% of cases can occur in the right atrium and about 5% in the left and right ventricles.¹ Although these tumors are histologically benign, they can lead to complications with a poor prognosis for the patient.²

Cardiac myxomas may present with obstructive, embolic, or constitutional symptoms. In addition, less common presentations include fever of unknown origin, endocarditislike findings in the presence of infection, acute myocardial infarction, and pulmonary embolism. In some cases, the first symptom of a myxoma may be an embolic event, and the diagnosis of cardiac myxoma can be made through histopathological examination of embolic material removed from a peripheral artery.^{1,3} When embolization occurs, the cerebral arteries are typically the most affected, followed by the renal artery, iliac artery, and femoropopliteal arteries. In rare cases, if the embolic myxoma material is particularly large, it can obstruct the infrarenal aortic bifurcation.⁴ In this case report, we present a patient with cardiac myxoma who died due to a cerebral embolic event following surgery for infrarenal abdominal aortic embolism.



CASE REPORT

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Figure 1. Computed tomographic angiography image showing total occlusion of the abdominal aorta, starting from the infrarenal level and extending to the bilateral femoral arteries (white arrow in A and B).

Case Report

A 60-year-old male patient with no history of chronic disease presented to the emergency department with complaints of back pain, paresthesia in the lower extremities, and subsequent development of paraplegia. The absence of distal pulses prompted a consultation with the cardiovascular surgery team. Initial physical examination in the emergency department revealed a Glasgow Coma Scale (GCS) score of 15, clear consciousness, blood pressure of 170/90 mmHg in the upper arm, heart rate of 90 beats per minute, and a normal sinus rhythm on electrocardiography (ECG). Both lower extremities were cold, pale, and cyanotic. Computed tomographic angiography (CTA), performed prior to the consultation, showed occlusion of the aorta at the infrarenal level (Figure 1). The patient underwent emergent surgery, and thromboembolectomy was performed. During the procedure, the retrieved material was found to be gelatinous and soft in consistency (Figure 2). Postoperative physical examination revealed palpable femoral, popliteal, and distal pulses, with warming and improved coloration of the bilateral lower extremities.

The patient underwent cardiac computed tomography (CT), transthoracic echocardiography (TTE), and transesophageal echocardiography (TEE) to investigate the source of emboli and identify any additional embolic events. Cardiac CT revealed a thrombus-like mass approximately 4 cm in diameter located in the left atrium (Figure 3A), as well as renal infarcts in the upper and lower poles of the right kidney, and splenic infarcts in the lower pole of the spleen. Brain CT revealed a hypodense nodular lesion measuring 10 mm in the left parietal lobe and 7 mm in the right parietal lobe, both located at the level of the centrum semiovale. TTE showed a mobile mass in the left atrium with irregular borders, measuring 4 × 2 cm, along

ABBREVIATIONS

- ASD Atrial septal defect
- CT Computed tomography
- CTA Computed tomographic angiography
- ECG Electrocardiography
- GCS Glasgow Coma Scale
- TEE Transesophageal echocardiography
- TTE Transthoracic echocardiography



Figure 2. Embolectomy material with an irregularly shaped, solid, dirty beige myxoid appearance, measuring $2.7 \times 2.5 \times 1.5$ cm and $1.8 \times 1.6 \times 1$ cm, with some areas containing maroon-colored hemorrhage (marked with a blue arrow). A maroon-colored thrombus-like tissue fragment measuring $2.5 \times 1.5 \times 1.3$ cm (marked with a red arrow).

with second-degree mitral regurgitation. TEE demonstrated a mobile mass with irregular borders, measuring 4.8 × 1.8 cm, originating from the septal region of the left atrium (Figure 3B). The patient was urgently taken to surgery for removal of the intracardiac mass.

Surgical intervention was performed via median sternotomy under cardiopulmonary bypass with standard aortic and bicaval cannulation. A soft gelatinous mass measuring approximately 4.5×3 cm, occupying the left atrium and attached to the atrial septum, was accessed through left and right atriotomy incisions. A right atriotomy with a transseptal approach was performed to provide better access to the mass within the septal wall. An atrial septal defect (ASD) was created, the mass was completely excised, and the ASD was subsequently closed. The excised mass was sent for pathological examination (Figures 4, 5).

On the first postoperative day, the patient was awake, and the GCS score was 11. He responded to verbal stimuli but presented with left-sided hemiplegia. In addition to the preoperative cerebral infarct areas, control brain CT and magnetic resonance imaging showed (Figure 6) extensive acute diffusion restriction in the right hemisphere. The neurology team initiated treatment with enoxaparin sodium.

Despite medical management, brain death occurred during follow-up, and the patient was declared deceased on the fourth postoperative day.



Figure 3. (A) Thoracic computed tomography image showing a thrombus-like mass approximately 4 cm in diameter in the left atrium (shown with white arrows in the axial section). (B) Transesophageal echocardiography image showing a mobile, irregularly bordered mass measuring 4.8 × 1.8 cm in the left atrium (circled in yellow).



Figure 4. The mass removed from the left atrium measured $3 \times 2.5 \times 1.8$ cm, and a smaller mass measured $1.1 \times 0.8 \times 0.2$ cm. The majority of the tissues appeared maroon and hemorrhagic, with some areas showing beige-colored, soft-consistency myxoid features.



Figure 5. (A and B) Pathological specimen image showing stellate-shaped stromal cells in a myxoid, vascular-rich stroma (x100, hematoxylin-eosin staining). (C) Pathological specimen image showing CD34 positivity in stromal cells (x40, immunoperoxidase staining).

Discussion

Myxomas may present clinically with nonspecific symptoms such as weakness, fever, weight loss, and myalgia, in addition

to symptoms related to obstruction within the heart or cerebral and peripheral embolization.⁵ In more than 50% of cases, myxomas are diagnosed only after systemic embolization or



Figure 6. (A and B) Magnetic resonance images taken on postoperative day 1 (blue and red arrows): Diffuse diffusion restrictions in the region corresponding to the right middle cerebral artery (MCA) territory; additional diffusion limitations in the bilateral lateral ventricles, left frontobasal region, left occipital lobe, and bilateral cerebellar hemispheres. (C) Computed tomography image (yellow arrows): Diffuse hypodense areas in the right cerebral hemisphere consistent with the MCA territory, along with localized hypodense areas in the left cerebellar hemisphere.

when the tumor reaches a size that obstructs the heart valves and causes hemodynamic disturbances. Systemic embolism is the most significant complication of myxomas located in the left atrium.^{1,3}

The typical location for myxomas is the fossa ovalis in the interatrial septum. The cardiac chamber in which the mass is located and the surface to which it is attached provide important clues for differentiating myxomas from thrombi, vegetations, and other tumors. In this distinction, the patient's clinical presentation, along with TTE and TEE findings, play a critical role.^{1,3} In our case, the myxoma was localized in the left atrium and had caused embolic events in multiple organs. Once the diagnosis of cardiac myxoma is established, surgical excision should be performed as soon as possible, given the ongoing risk of embolic complications. Careful handling of cardiac structures and the tumor during removal reduces the risk of intraoperative embolic events as well as the likelihood of tumor fragmentation.²

Macroscopically, if the tumor is soft, villous, and has papillary projections, it is more prone to embolization. In contrast, tumors with a smooth and more fibrotic surface are less likely to cause embolization. Myxomas localized in the right atrium more frequently exhibit calcification and, as a result, are reported to cause fewer embolic events. Embolism occurring in the absence of cardiac arrhythmia is considered characteristic of myxomas.⁶ In our case, the thrombectomy material was macroscopically soft, easily fragmentable, and had an irregular surface. The patient did not have any cardiac arrhythmia.

Left atrial myxomas are a rare but recognized cause of stroke and acute limb ischemia.⁶

Tumor cells seeded within the embolus may proliferate and form metastatic lesions.⁷ Emboli most commonly affect the brain but may also involve other organs, such as the liver, spleen,

kidneys, retina, coronary arteries, and peripheral arteries.⁸ In rare cases, a left atrial myxoma may completely detach and obstruct the aortic bifurcation. Emboli of this size at the aortic bifurcation have been reported in the literature to mimic the clinical presentation of aortic dissection and may require immediate surgical intervention.^{8,9} It is generally recommended to remove the primary cardiac lesion before addressing brain metastases, unless there are urgent surgical indications, such as hemorrhage with significant mass effect or progressive neurological deficits.⁷ There have been reports of cases where myxoma emboli manifest as systemic or cerebral metastases, even years after removal of the cardiac tumor, as well as cases of pulmonary hypertension when the tumor originates from the right atrium.^{7,10} Due to the rarity of metastatic myxomatous lesions, data on their treatment are limited, and there are no established standards of care.⁷ Early diagnosis is crucial, as it significantly reduces complications related to arterial occlusion and increases survival rates.

In our case, peripheral embolizations occurred in the splenic artery, renal artery, cerebral arteries, and abdominal aorta. Despite prompt intervention with embolectomy and atrial myxoma removal, the development of extensive peripheral organ emboli in the preoperative period ultimately led to mortality in this case.

Conclusion

In patients presenting with sudden arterial embolism and no prior medical history, cardiac myxoma should be suspected, particularly in the absence of cardiac arrhythmia, atherosclerosis, or previous thromboembolic events. In cases of embolization affecting multiple regions, intracardiac masses, especially myxomas, should be considered in the differential diagnosis. Prompt diagnosis and timely treatment are essential to improve clinical outcomes and prevent embolization-related complications. Çelebi et al. Silent Killer within the Heart: Cardiac Myxomas

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