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Case Report / Olgu Sunumu

A cardiac myxoma detected during chemotherapy in an operated recurrent rectal cancer patient

Ameliyat edilmiş tekrar eden rektal kanser hastasında kemoterapi sırasında saptanan kardiyak miksoma

Ahmet Barış Durukan, Hasan Alper Gürbüz

Department of Cardiovascular Surgery, Memorial Ankara Hospital, Ankara, Turkey

ABSTRACT

Cardiac myxomas constitute over 50% of benign cardiac tumors and are the most common primary cardiac tumors. Concomitance with other primary tumors is rare. In this article, we report a 57-year-old male patient operated for recurrent rectal cancer who was diagnosed with concomitant cardiac myxoma and coronary artery disease during the third course of chemotherapy following colon surgery. Two-vessel bypass simultaneous with cardiac myxoma excision was performed with repair of atrial septum using Teflon patch. In patients with concomitant malignancy, timing of surgery for cardiac pathologies incurable by medical treatment or interventional measures is crucial, and there is still no consensus on this subject.

Keywords: Coronary artery disease; myxoma; rectal neoplasms.

ÖZ

Kardiyak miksomalar benign kardiyak tümörlerin %50'den fazlasını oluşturur ve en sık rastlanan primer kardiyak tümörlerdir. Diğer primer tümörlerle eşlik etme durumu nadirdir. Bu yazıda, tekrar eden rektal kanser nedeni ile ameliyat edilen ve kolon cerrahisi sonrası üçüncü kür kemoterapi sırasında eşlik eden kardiyak miksoma ve koroner arter hastalığı tanısı konulan 57 yaşında bir erkek hasta sunuldu. Atriyal septumun Teflon yama ile onarımı ile beraber kardiyak miksoma eksizyonu ile eş zamanlı iki damar baypas uygulandı. Eşlik eden malignitesi olan hastalarda, tıbbi tedavi veya girişimsel yöntemler ile tedavi edilemeyen kardiyak patolojiler için cerrahinin zamanlaması önemlidir ve bu konu üzerinde halen bir görüş birliği yoktur.

Anahtar sözcükler: Koroner arter hastalığı; miksoma; rektal neoplazmlar.

Primary cardiac tumors are rarely encountered in cardiac surgery practice with an autopsy incidence of 0.002-0.3% in population. Two-thirds of primary tumors are benign.[1] Of all heart tumors, cardiac myxomas (CMs) have an incidence of 83%. More accurately, the incidence of CMs is 0.5-1 case/1,000,000 individuals/year. Cardiac myxomas usually have female predominance, diagnosed at ages 40-60, and are 90% sporadic.[2] Patients may be asymptomatic or present with constitutional symptoms, obstructive cardiac symptoms representing as pulmonary edema/heart failure or coronary, cerebral or peripheral embolization.[2,3]

Cardiac tumors, particularly CMs are rarely concomitant with other primary malignant tumors and their diagnosis and treatment are challenging due to the present clinical condition of the patient affected by prior malignancy, surgery, and prior/ongoing chemotherapy.[4]

In this article, we report an operated recurrent rectal cancer patient diagnosed with CM and coronary

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Correspondence: Ahmet Barış Durukan, MD. Memorial Ankara Hastanesi Kalp ve Damar Cerrahisi Bölümü, 06520 Çankaya, Ankara, Turkey.
Tel: +90 312 - 253 66 66 e-mail: barisdurukan@yahoo.com

artery disease during third course of chemotherapy (CT). Two-vessel bypass simultaneous with CM excision was performed.

CASE REPORT

A 57-year-old male patient was consulted with the diagnosis of CM. He was operated on 14 months before for rectal cancer ($T_3N_0M_0$, grade 2 adenocarcinoma), (tumor excision + colostomy, soon closed). Six courses of CT and 28 days of radiotherapy were administered. Four months before, local recurrence with hepatic metastasis was found. Abdominopelvic-resection and hepatic metastasectomy were performed with open colostomy. Scheduled 12 courses of FOLFOX regime (Oxaliplatin-85 mg/m^2, one day; folinic acid-400 mg/m^2, 5-FU-2400 mg/m^2 46 hours infusion; every 15 days) were initiated. Transthoracic echocardiographic examination (TTE) did not reveal any cardiac mass three months before. During the third course of CT, routine TTE revealed a mass attached on the left atrium (LA) side of interatrial septum (26x28 mm). No valvular pathology was noted, but dyskinetic regions (ejection fraction: 40%).

Operation within five days was suggested by oncology. Coronary angiography was performed (30% stenosis in mid-left anterior descending artery, total occlusion of circumflex; increased thrombolysis in myocardial infarction two flow, 90% stenosis in right coronary artery).

Diagnosis of CM larger than 1 cm in diameter indicated early operation. A written informed consent was obtained from the patient. Chemotherapy was ongoing and was in the last 12 hours. The operation was scheduled to 24 hours following CT.

Median sternotomy and cardiopulmonary bypass (CPB) were performed (33 °C). After cardioplegic arrest, a horizontal left atriotomy, followed by vertical right atriotomy were conducted. A mass located on LA side of interatrial septum, solid in nature with compact consistency was observed (Figure 1a). The mass with interatrial septum including 2-3 mm tissue surrounding pedicle were removed (35x20x20 mm) (Figure 1b). A 25x25 mm defect formed on septum was repaired using Teflon patch. Biatrial incisions were closed primarily. Saphenous vein grafts were anastomosed to right posterior descending and circumflex posterolateral coronary arteries.

The patient was weaned from CPB with 5 mcg/kg/minute dopamine infusion. He was extubated after six hours, stayed in intensive care unit for 36 hours and discharged on fifth day. The whole postoperative course was uneventful. After 15 days, the fourth course of CT was administered. The patient had 12 courses totally, uneventfully. The patient is still alive after 21 months and on follow-up with no complaints.

DISCUSSION

Myxomas are usually located in the left atrium, in the region of fossa ovalis. They are derived from primitive endocardial/epithelial precursors.[3] More specifically, they develop from multipotent stem cells surrounding fossa ovalis and surrounding endothelium.[5] The St. John Sutton classification divides CMs as solid, which are smooth surfaced and have compact consistency (can be removed in one piece), and papillary, which are gelatinous (often piecemeal removal is necessary).[2] Our patient had a typical solid CM case that was removed as a

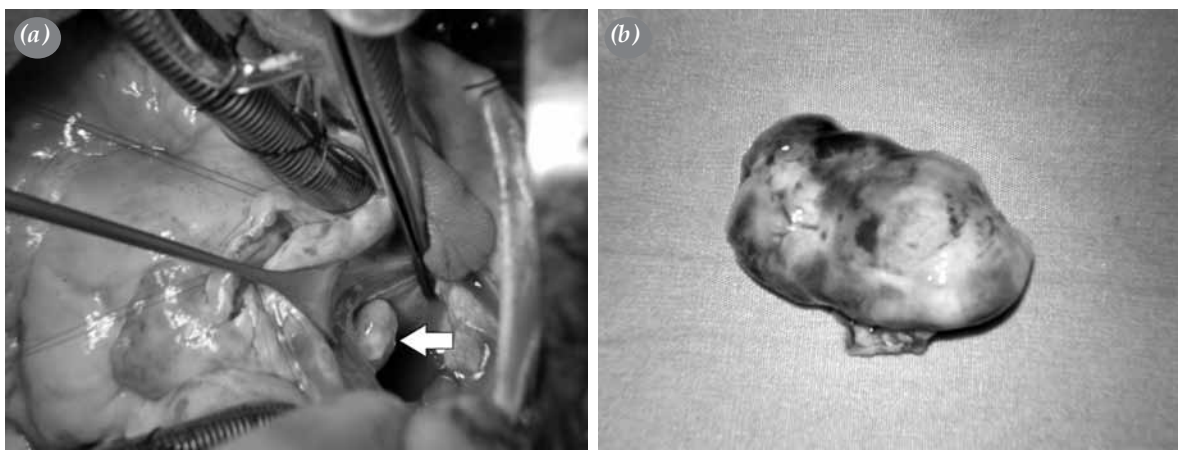

Figure 1. (a) Intraoperative image of mass seen from left atrial side. **(b)** Mass with dimensions of 28x26 mm after total removal.

solitary whole mass with ease without any particulate fragmentation. It was not possible to diagnose it as a solid CM prior to the operation, if so stenting the coronaries and postponing the operation for CM removal would be a good alternative option. However, there was no such modality to guarantee that no embolization would occur.

Up to 12% of primary cardiac tumors are asymptomatic and diagnosed incidentally. If clinically symptomatic, the classical triad is hemodynamic symptoms caused by intracardiac mobile mass mostly obstructive, embolism or constitutional symptoms.[5] The diagnosis is mainly by TTE, if the window is poor, transesophageal examination (TEE) is made. Either TTE or TEE have a detection rate over 95% and can also distinguish CM from LA thrombi.[2] The case we report here was incidentally diagnosed during routine TTE performed during the course of CT. The three months prior to TTE examination did not reveal any mass originating from the septum. To our knowledge, there is no time period documented for development of a CM. We had TTE three months before, so we believe that a 35x20x20 mm mass developed in three-month period. This cannot be a really definitive time for development of a CM, because the patient had recurrent rectal cancer, and some genetic and paraneoplastic mechanisms might have played role in its development.

The association of CMs with some other tumors has been defined as Carney complex which include mucosal/cutaneous myxomas, mucocutaneous hyperpigmentation, blue nevi, schwannomas, endocrine overactivity, and tumors.[3,5] It is X-linked autosomal dominant with complete penetrance. Mostly multicentric location and localization other than left atrial side of interatrial septum are seen. In sporadic cases, 60-80% are located in left atrial side of interatrial septum followed by 15-28% right atrial side, 8% right ventricle, and 3-4% left ventricle.[5] This complex does not include any gastrointestinal system tumor. Our case was CM located on LA side of interatrial septum concomitant with rectal cancer, and there was also no CM history in close relatives.

Metastatic cardiac tumors are 100 times more common than primary cardiac tumors, but are mostly documented on autopsy series.[5] Makhija et al.[4] published three cases of metastatic cancer to the heart mimicking thrombi or myxoma; particularly rectal, breast, and tongue carcinoma. Their report was the first case report to document invasion of endocardium from rectal primary. We have also hypothesized such a possibility prior to the operation, but the pathology report was certain on diagnosis of CM.

Fujisaki et al.[6] documented two cases with digestive cancer that required cardiac surgery during the course of CT after surgery for the primary malignancy. One was gastric cancer that developed CM and other was rectal cancer that developed endocarditis. In both cases, they have performed surgery during the course of disease and surgery was successful; however, both cases had recurrences which they thought were due to excessive surgical stress. Our patient had a similar course of disease, but time will show if recurrence will occur or not. However, there was no recurrence on first year.

The treatment of CMs is immediate excision if larger than 1 cm diameter due to the risk of embolism.[5] The most striking feature is not to handle the heart before aortic clamping. As soon as the patient is on CPB, the patient is cooled and aorta is immediately cross-clamped. Despite the ongoing chemotherapy, we scheduled early surgery due to the embolization risk. The chemotherapy was planned to last for 46 hours and there were 12 hours left for it to finish. Coronary angiography was performed since the patient was 57-year-old and had wall motion abnormalities on TTE. Totally after 36 hours, the tumor was removed. The decision for surgery despite chemotherapy was made based on expected survival of the patient and the potential risk of emboli.

Concomitant procedures during CM excision are not frequently encountered. Lin et al.[2] reviewed their 68 CM cases in a 16-year period. They have performed all under moderate hypothermia and with median sternotomy. They have performed coronary angiography if the patient was aged over 45 or had a history of chest pain. They have performed concomitant valvular procedures -either valvuloplasty or replacement in 23 cases- but coronary artery bypass grafting (CABG) only in one case. Pineda et al.[7] documented their experience on removal of benign cardiac masses; of 39 cases, they have preferred minimally invasive approach on 22. They have excluded 23 cases with concomitant cardiac procedures of which 15 was CABG. They have emphasized that the benefits of minimal invasive approach on other types of cardiac surgery (decreased blood loss, decreased hospital length of stay, decreased costs etc.) were also valid for benign cardiac masses. They have performed femoral cannulation and either right thoracotomy or transverse parasternal incision based on the location of the tumor. They have concluded that

minimal invasive approach was not inferior to median sternotomy approach. We have performed the surgery with median sternotomy (since we planned to perform bypass on circumflex and right coronary arteries) under mild hypothermia, we have not handled the heart until aortic cross-clamping.

In conclusion, we reported a cardiac myxoma case concomitant with rectal cancer managed successfully. The surgery and prognosis of cardiac myxomas are well documented and excellent. However, the management of cases with cardiac myxomas concomitant with other diseases, particularly malignancies are challenging. A decision for timing of surgery in these cases is quite difficult, and there is still no consensus on this subject.

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