Surgical Treatment Of Pulmonary Artery Angiosarcoma Causing Severe Obstruction Of The Pulmonary Artery

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ABSTRACT

Pulmonary artery sarcomas are rarely seen and frequently misdiagnosed as pulmonary thromboembolic disease due to the presence of similar clinical manifestations and radiologic findings. As making the certain diagnosis is very important in order to determine the treatment strategy, one needs close follow-up to make a definite diagnosis. However, most of the cases are diagnosed during the surgery or autopsy. We report the surgical outcome of a patient with pulmonary artery angiosarcoma causing severe obstruction of the pulmonary valve and main pulmonary artery.

Key Words: Pulmonary artery sarcoma, cardiopulmonary bypass, pulmonary thromboembolism

ÖZET

Ciddi Pulmoner Arter Darlığına Neden Olan Pulmoner Anjiosarkomun Cerrahi Tedavisi

Pulmoner arter sarkomları nadir görülür ve benzer klinik tabloya sahip olmalarından dolayı sıklıkla pulmoner tromboembolik hastalık tanısı konulur. Tedavi stratejisini belirlemek açısından doğru tanının konması çok önemlidir. Bu nedenle bu hastalar doğru teşhiş konulması için yakın takip edilmelidir. Ancak olguların çoğuna cerrahi veya otopsi esnasında teşhis konulabilmektedir. Bu olgu sunumunda ana pulmoner arter ve pulmoner kapakta ciddi darlığa neden olan pumoner anjiosarkomlu birhastanın teşhis ve cerrahi tedavisini değerlendirdik.

Anahtar Kelimeler: Pulmoner arter sarkomu, kardiyopumoner baypas, pulmoner emboli

INTRODUCTION

Pulmonary artery angiosarcoma is a rarely seen and highly mortal malignant tumor. It can present with non-specific symptoms as dyspnea, chest pain, cough, or haemoptysis (1). Because of the similar clinical manifestations, it can be frequently misdiagnosed as pulmonary thromboembolism. When diagnosed median survival time only a few months without surgical resection. However, the surgical management improves clinical symptoms and presents the only chance for survival (2). As the pulmonary artery angiosarcomas imitate the pulmonary artery thrombus, the required prompt therapy may be delayed. We report a case of Pulmonary artery angiosarcoma, which was thought be chronic pulmonary thromboembolic disease preoperatively causing severe obstruction of the pulmonary valve and pulmonary artery.

CASE REPORT

A 37-year-old man who had progressive shortness of breath and fatigue for one year was referred to our hospital with the diagnosis of pulmonary artery embolism. He has no venous thromboembolism or chronic pulmonary disease history. Physical examination revealed normal pulmonary sounds on auscultation. On cardiac auscultation, grade 2/6

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mid-systolic murmur could be heard at the left second intercostals space. There were no significant abnormal findings concerning the blood count and chemistry. Chest radiograph showed a prominent main pulmonary artery. A baseline ECG revealed normal sinus rhythm and right ventricular strain pattern. The transthoracic echocardiogram showed a severe pulmonary valvular stenosis with an elevated right-ventricular-systolic-pressure to more than 70mmHg and enlargement of right-chambers. The ejection fraction was 0.70 and severe tricuspid insufficiency was detected. After the initial evaluation, the heparin therapy was initiated promptly. In order to define the precise character and expansion of the pulmonary thrombi Computed-tomography angiography (CTA) was done. The CTA showed a huge intravascular mass containing lipid density at the main pulmonary artery and proximal part of right pulmonary artery partially filling the lumen. Increased collateral circulation in the anterior mediastinum was also revealed. These findings were concluded in favor of a pulmonary artery tumor or pulmonary artery thrombus. Coronary artery angiography revealed normal coronary arteries. On cardiac catheterization pulmonary artery, right ventricular pressures and pulmonary valvular gradient were elevated. Because of high risk of embolization and suspicion of tumoral characterization of the intravascular mass on urgent operation was scheduled. The operation was initiated with median sternotomy and was followed by total cardiopulmonary bypass and cardioplegic arrest. Once the thickened and hard main pulmonary artery was incised, the tumor was seen to extend from right ventricular outflow tract (RVOT) to the distal part of the right pulmonary artery. Additionally, the pulmonary valvular structures were invaved by tumoral tissue. After the determination of the expansion, Intra-luminal tumoral mass was excised and pulmonary valve was resected because of the valvular infiltration (Figure-1,2). Pulmonary valve replacement with prosthetic valve was performed. The patholigical examination underlight microscope revealed malign tumoral infiltrations, which had necrotic areas, abundant vascular structures, and pleomorphic cells. Some of tumor cells had vesiculary nucleus others had hyperchromatic nucleus and clear or eosinophilic cytoplasm. The mitotic activity was high. Immunohistochemical studies revealed that CD31 and vimentin were positive while desmin was negative. Based on these findings a certain diagnosis of pulmonary artery angiosarcoma was made. After the consultation of oncology department, the patients' chemotherapy and radiotherapy was planned. On the first, second, sixth and twelfth month following operation computed tomography and echocardiograhy was repeated and no metastasis or recurrence of the malignancy was detected.

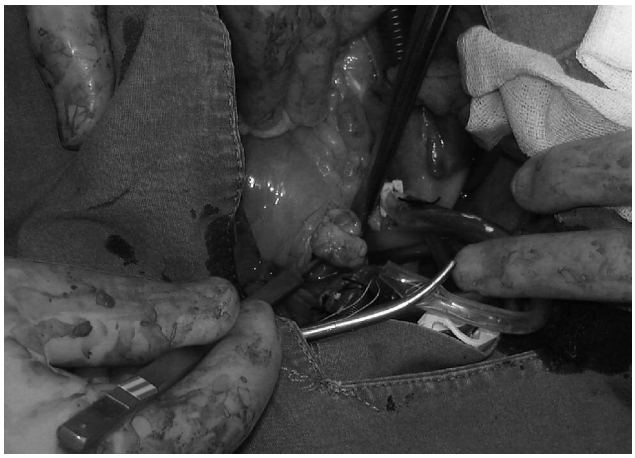

Figure 1: ???????? eksik tamamla

Figure 2: ???????? eksik tamamla

DISCUSSION

Pulmonary artery sarcomas are infrequent and usually carries a high mortality rate. Additionally, angiosarcoma is one of the rarest types of pulmonary artery sarcomas (2,3). There are few pulmonary artery anjiosarcoma cases reported in the literature. Generally, the diagnosis is difficult and delayed because symptoms are nonspecific and survival time is limited (2). Correct diagnosis was made during a postmortem examination in 59% and after surgical exploration in 31% of cases (4).

In the cases with pulmonary angiosarcoma the reported range of age is 38-69 years (3). Our case was relatively young in comparison with them. He was referred to our clinic with the diagnosis of pulmonary artery embolism with non-specific symp-

toms, which was also the case in reported cases in the literature (5,6).

Because pulmonary artery angiosarcomas are rarely seen, have insidious growth characteristics, and both clinical (dyspnea, chest pain, syncope, and palpitations) and radiological findings mimic pulmonary thromboembolic disease due to obstruction of the pulmonary artery, correct diagnosis tends to be mistaken or delayed (5,6). Therefore, inappropriate medical therapy such as prolonged anticoagulation or thrombolysis is usually applied (7). When an earlier and accurate diagnosis is made it gives the chance of an earlier interventions and improved patient's survival (8). Literature on this disease has reported a case who survived for 56 months after aggressive surgical resection in combination with the adjuvant chemoradiotherapy. However, without surgical resection the survival can be as short as 1.5 months.

Computed tomography angiography is useful for differentiation of pulmonary artery sarcoma from a pulmonary embolism. A filling defect sign seen on CTA occupying the entire luminal diameter of the pulmonary artery, expansion of the pulmonary artery branches leads to a suspicion of pulmonary artery angiosarcoma (1,9). Although the presence of these advantages of CTA in the differential diagnosis of pulmonary embolism, we could only made the certain diagnosis with histopathologic examination.

Pulmonary angiosarcomas generally arise from the main pulmonary artery. They can extend to the RVOT involving the pulmonary valve and more distal pulmonary artery branches (5,9). Similarly, in our case the tumor was seen to extend from right ventricular outflow tract to the distal part of the right pulmonary artery invasing the pulmonary valvular structures. Because this broad tumoral expansion including the majority of main and right pulmonary artery, and the pulmonary valve, the use of cardiopulmonary bypass was required in our case. All tumoral tissues were extracted from RVOT, main and right pulmonary artery. Additionally, pulmonary valve replacement with prosthetic valve was performed because of pulmonary valvular invasion by pulmonary angiosarcoma. After the specimens evaluation, pulmonary angiosarcoma certain diagnosis was confirmed by histopathologic and immunohistochemical examinations. At 15 months after surgery, no recurrence was observed in our patient, which supports our suggestion that aggressive surgical resection in combination with chemotherapy and radiotherapy can prolong survival.

In conclusion, the pulmonary artery angiosarcoma should always be included in the differential diagnosis of pulmonary embolism although it is seen rarely. Early and accurate diagnosis can be provided by meticulous physical examination and computed tomography scanning. It is possible to prolong patient's survival and improve quality of life by an aggressive surgical management of a malignant pulmonary artery tumor in combination with chemotherapy and radiotherapy. Therefore, pulmonary artery angiosarcoma may not carry a high mortality when aggresive surgical resection performed accepted as a fatal diagnosis.

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