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Recurrent Left Atrial Botyroid Rhabdomyosarcoma

Günseli Abay, MD,[1] Cemal Kocaaslan, MD,[2] Rafet Gunay, MD,[2] Meryem Dogan, MD,[3] Mahmut Murat Demirtas, Assoc Prof[2]

Departments of [1]Anesthesiology and [2]Cardiovascular Surgery, Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Center, Istanbul; [3]Pathology Department, Haydarpasa Numune Hospital, Istanbul, Turkey

INTRODUCTION

Primary heart tumors are extremely rare and their frequency ranges from approximately 0.01-0.3% in autopsy series. Nearly one quarter of all primary cardiac tumors are malignant tumors such as sarcoma. Rhabdomyosarcoma is the second most common malignant primary tumor of the heart following angiosarcoma.

Primary cardiac tumors present with one or more of the symptoms of the classic triad: cardiac symptoms and signs resulting from intracardiac obstruction; signs of systemic embolization; and systemic or constitutional symptoms. The prognosis after surgery is usually excellent in case of benign tumors, but is unfortunately still limited in localized malignant diseases [Butany 2005].

In this case report we present a 45-year-old female patient operated three times in 9 years because of left atrial tumor.

CASE REPORT

The patient was operated by our cardiac surgery clinic because of intracardiac mass (at the left atrium) in 2004. The pathology was reported as primary cardiac rhabdomyosarcoma (botyroid embryonal type) so she was referred to the oncology clinic for further treatment. At the oncology clinic the patient received chemotherapy with a drug protocol of cyclophosphamide, doxorubicin, and vincristine with no adjuvant radiation.

In 2006 a malignant mesenchymal tumor was detected in the right lung. The tumor was resected by thoracic surgeons and the pathology was reported as rhabdomyosarcoma as a metastatic tumor from the heart.

The patient was operated again at 2010 because of a tumoral mass at the left atrium and the pathology was reported as rhabdomyosarcoma (botyroid embryonal type) again.

The final admission occurred in April 2013 with dyspnea and chest pain. A tumoral mass formation within the left atrium was detected with TEE and surgery was planned again. When the interatrial septum was exposed during cardiopulmonary bypass, several grayish-white tumoral masses of fibrous character were found, ranging from 5 x 4 to 0.8 x 0.5 in diameter (Figure 1). Immunohistochemical examination showed desmin positivity in addition to 90% necrotic areas within the tumor (Figure 2). A diagnosis of recurrent botyroid embryonal rhabdomyosarcoma was made by the pathology clinic. After the operation the patient was discharged and healing.

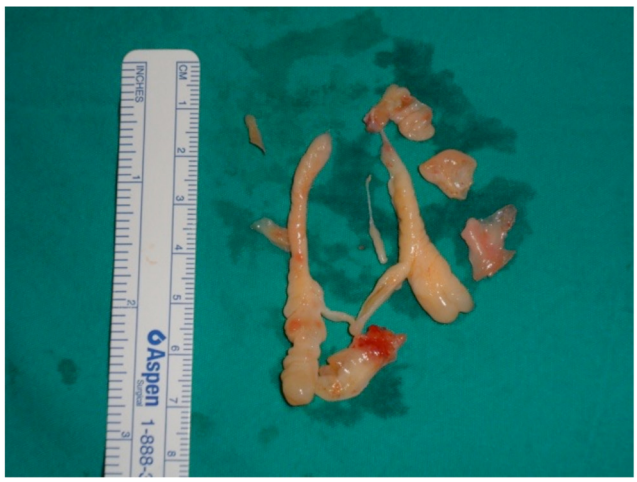

Figure 1. Rhabdomyosarcoma-recurrent (macroscopic appearance); tumoral mass pieces in dirty-white color with fibrous character.

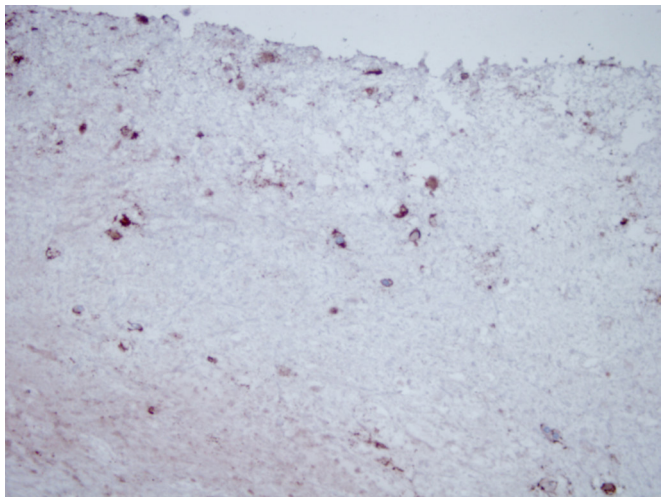

Figure 2. Rhabdomyosarcoma-recurrent (microscopic appearance); desmin (+) staining tumoral cross-section (desmin × 20).

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Correspondence: Dr. Siyami Ersek Hast. Tibbiye Cad, Üsküdar, Istanbul, Turkey; 00 90 505 391 98 12; fax: 00 90 216 544 44 44 (e-mail: cemalkocaaslan@yahoo.com).

DISCUSSION

Rhabdomyosarcomas are rare cardiac tumors comprising nearly 4% of all primary cardiac malignancies. A soft tissue tumor of childhood predominantly, rhabdomyosarcomas originate from embryonal mesenchyme and they are classified into further subtypes based on their histopathological cell type, such as embryonal, alveolar, and pleomorphic [Wexler 2002]. Botyroid rhabdomyosarcoma has a better prognosis than other subtypes and represents a subgroup of embryonal tumors. Embryonal rhabdomyosarcoma almost always occurs in children and young adults, with a rare occurrence in adults [Araoz 1999].

Although the lesion of our patient was located in the left atrium, which is the most common site for cardiac tumors, rhabdomyosarcomas generally do not exhibit specific site preference for an individual cardiac chamber and may develop in any of these, sometimes simultaneously [Karpuz 2007].

Despite advanced diagnostic techniques and therapeutic options, primary cardiac rhabdomyosarcomas in adults usually have a quickly progressive course, with a dismal prognosis usually resulting in death on average six months after the diagnosis [Steger 2012]. Metastases usually occur early and mostly involve the lung parenchyma, thoracic lymph nodes, and the mediastinum [Chlumsky 2001].

Although survival rates improve from 20-70% with surgical resection and subsequent chemotherapy, adequate surgical resection is usually not possible with a high risk of recurrence. Therefore, partial or complete surgical resection generally represents a palliative intervention that aims to improve the symptoms and the quality of life. In patients without metastases, orthotopic heart transplantation is another option.

Primary cardiac rhabdomyosarcoma is a very rare entity among adult patients and we believe that our patient represents an interesting case of primary cardiac rhabdomyosarcoma with three recurrences involving lung metastases over a 9-year period, and corresponds to one of the longest survival rates reported in the literature.

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