Case report
Asymptomatic right atrial leiomyosarcoma with tricuspid valve obstruction in a young female patient

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Received 7 May 1998; revised version received 9 September 1998; accepted 29 September 1998

Abstract

Primary cardiac leiomyosarcomas of the heart are rare tumors usually diagnosed post-mortem. Like other sarcomas located in the heart, it causes symptoms by obstruction or occlusion of cardiac cavities, local invasion, embolization or by systemic manifestations. We present an unusual case of a previously healthy young female patient who was accidentally diagnosed with a cardiac tumor of unknown origin during routine physical examination in May 1997. Until a few days prior to elective cardiac surgery for diagnostic purposes in June 1997, no clinical symptoms were present. To our surprise, a primary right atrial leiomyosarcoma was found which almost completely occluded the right atrium and destroyed the tricuspid valve. Despite the combination of surgical removal and adjuvant chemotherapy the patient died 3 weeks after the operation due to progressive tumor disease and development of congestive heart failure and lung embolism.

Keywords: Cardiac leiomyosarcoma; Heart tumor; Palliative resection; Adjuvant therapy

1. Introduction

Approximately 10% of primary cardiac tumors are malignant and most of them are sarcomas. They occur in either sex and may be found in patients of all ages [1,2].

Primary cardiac leiomyosarcomas usually originate within the major thoracic vessels [3].

We report an unusual case of a primary cardiac leiomyosarcoma evolving within the right atrium with subtotal occlusion of the atrial cavity and complete destruction of the tricuspid valve; there was a lack of clinical symptoms and early death due to massive tumor progression despite combined surgical and adjuvant chemotherapy.

2. Case report

A 21-year-old caucasian female was admitted to our hospital because of a rough diastolic murmur in the third left intercostal space for further medical examination. She did not complain of any clinical symptoms. Chest X-ray was normal, the electrocardiogram showed a regular sinus rhythm and a P-dextrocardiale.

Transthoracic and transesophageal echocardiography showed a tumorous mass of approximately 3 cm x 6 cm in size within the right atrium, invading the tricuspid valve and the interventricular septum.

Abdominal ultrasound demonstrated a thrombus of 1.3 cm x 1.6 cm x 4.1 cm extension in the inferior vena cava just below the diaphragm without extension to the right atrium.

Computed tomography of the chest and abdomen revealed a large hypodense tumorous mass of 3.5 cm x 6 cm within the right atrium, invading the right ventricle (Fig. 1). In addition, several suspicious foci in the right lung were identified. Due to the patient’s age and the suspected neoplasm’s aggressiveness, an elective thoracotomy for open chest cardiac biopsy and curative tumor resection after intraoperative rapid histological analysis was planned. During induction of anesthesia the patient became hemodynamically unstable and was put on cardio-pulmonary bypass.
A large atrial tumor including the completely tumorous obstructed tricuspid valve were partially removed in order to open the valve area and thus reduce an existing upper inflow obstruction. Only palliative resection could be performed. Postoperatively, the patient was hemodynamically stable with low dose catecholamines and a reduction of the central venous pressure from 30 mmHg to 14 mmHg.

Despite postoperative treatment with Adriamycin (120 mg/day) and Ifosfamid (2.5 mg/day) as adjuvant chemotherapy, the patient died from autopsically confirmed pulmonary thrombemboly and progressive tumor disease 3 weeks later.

3. Pathological findings

Histologically, the tumor cells had hyperchromatic, blunt-ended nuclei. Mitotic numbers ranged between two and four per ten high-power fields (HPF). Zonal necroses were present (Fig. 2).

Immunohistochemically, the majority of the tumor cells showed a positive reaction for smooth-muscle actin and a part of the cells also for desmin. Accordingly, the tumor was classified as leiomyosarcoma, grade 2.

At autopsy the main tumor mass originated in the right atrium (7 × 5 × 2 cm). Moreover, the leiomyosarcoma invaded the tricuspid valve, the atrial septum, the mitral valve and the base of pulmonary artery and aorta.

There was no evidence of peripheral abdominal or cerebral metastasis.

The cause of death was recurrent lung embolism, partly of neoplastic origin.

4. Discussion

Approximately 10% of primary cardiac tumors are malignant with angiosarcoma being the most common type [4]. Leiomyosarcoma of cardiac origin is uncommon and diagnosis has often been made post-mortem [4]. In the AFIP cohort 76% of these tumors were located in the left atrium, 16% in the right atrium, and 8% diffuse in the ventricles [4]. The mean age of presentation is within the fourth decade. Other large surgical series were even negative for this kind of tumor [5].

Primary leiomyosarcomas are highly aggressive and infiltrate adjacent tissues.

Although most patients die within 1 year of diagnosis, Pesotto et al. could recently document a 7-year survival after combined surgical and adjuvant radiochemotherapy [10]. The short survival time generally observed in these patients is due to the inability of complete tumor removal...
and surgical resection is often palliative in order to relieve symptoms caused by obstruction of cardiac cavities or major blood vessels. The clinical signs depend on the location of the tumor and its mass effect [2].

Echocardiography is a very useful non-invasive tool for imaging suspicious cardiac tumors and for further differentiation concerning the tumor’s dignity [7]. For the operative planning and surgical staging computed tomography or magnetic resonance imaging remain essential [6]. Indispensable for performing a differentiated therapy is the exact histological examination of tumor specimen gained by transvenous or open chest biopsy [1,10]. The therapy of choice is complete surgical resection in combination with adjuvant chemotherapy or radiotherapy [8,10].

As a last resort, heart transplantation is a possible therapy if the patient is in good condition and there are no signs of metastasis, unlike in our case [9].

Several unusual findings were observed in our case. First, the patient did not have any clinical symptoms at the time of diagnosis despite progressive tumor growth.

Second, diagnosis was made before clinical symptoms occurred but no sufficient therapy could be offered. This indicates, that considering the literature available, no adequate diagnostic tool exists in order to identify patients at risk in very early stages.

Third, the tumor was located primarily in the right atrium, leading to a destructive obstruction of the tricuspid valve. In addition, the tumor was invading the pulmonary vessels. A recurrent pulmonary thrombemboly was in part responsible for the patients poor prognosis. This case remarkably reflects the difficulty of early diagnosis in this kind of tumor disease and the necessity of immediate radical surgical therapy in combination with adjuvant radiochemotherapy.

References


Fig. 2. Tumor cells with blunt ended nuclei and the presence of a mitosis (H and E stain, magnification 400x).
