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Short Communication

Left Atrium Myxosarcoma: An Exceptional Cardiac Malignant Primary Tumor

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Summary

We report the case of a 32-year-old patient with a left atrium myxosarcoma, presenting with congestive heart failure. It is a rare cardiac malignant primary tumor that seems to derive from the same cellular line as myxomas, but the prognosis is very different. These tumors present local recidives and distance metastasis, so the mean survival is about 1 year, independent of any therapeutical option.

Key words: myxosarcoma, myxoma, sarcoma, heart

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Introduction

Myxosarcoma is a rare neoplasia very difficult to differentiate from myxoma. The ratio between the incidence of myxomas and myxosarcomas is approximately 0.8:100[1] and their prognosis are very different. Myxosarcoma most frequently originates in the left atrium (LA),[2–4] but it can also appear in other sites.[5–7] We report the case of a patient with a myxosarcoma. Although the patient had no metastasis, there was early local recurrence and a fatal end despite the association of chemotherapy to surgical resection.

Case Report

A 32-year-old man was referred to our hospital with congestive heart failure. Transthoracic echocardiograms (TTE) and transesophageal echocardiograms (TEE) showed a large mass (5 × 6 cm) occuping nearly all the LA. It was attached to the interatrial septum and protruded during diastole into the LV causing a severe flow obstruction between LA and LV (Fig. 1). The tumor was compact, with well-defined edges and with small echo-free spaces inside it. The location and its appearance made us think about malignancy. Pulmonary veins were not affected. There was no thoracic metastasis. The tumor was surgically excised, but a TTE performed 1 month later showed a 2.1 × 1.4 cm mass in the LA and the histopathologic study proved the diagnosis of myxosarcoma (Fig. 2). Chemotherapy was immediately initiated and the tumor size was reduced, but never disappeared completely. Our patient was asymptomatic during the next 9 months. He then suffered a left optic neuropathy attributed to tumoral infiltration. One year after surgical intervention we found a mass in the LA with the same size and characteristics of the first tumor. We proposed a new chemotherapy cycle, but the patient died suddenly that day and unfortunately we could not get the autopsy.

Discussion

Myxosarcoma is a rare form of primary cardiac malignant tumor with only a few cases reported. They have local recurrences, involving pulmonary artery, pericardium or pleura and distance metastasis with the

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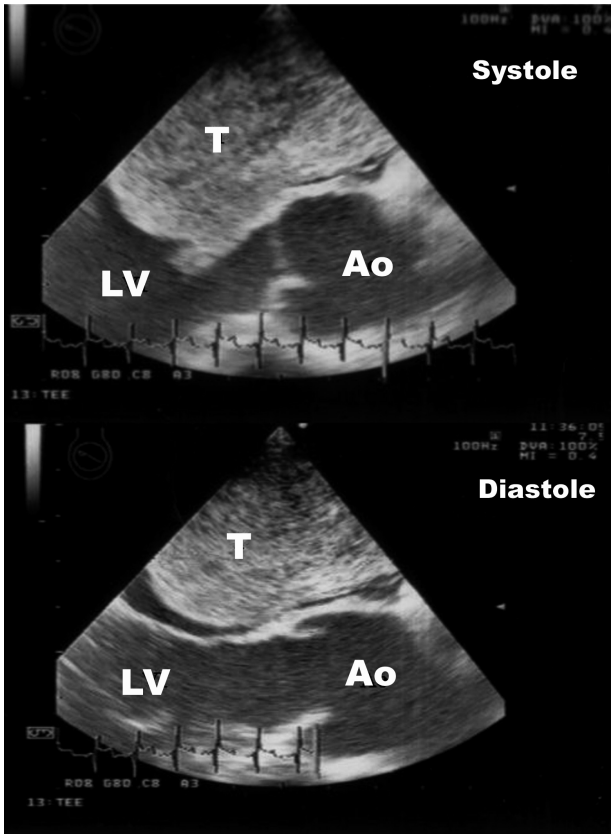

FIG. 1 Tranesophageal echocardiogram showing the mass, occuping all the LA, and protruding into mitral valve during diastole. Note the well-defined edges and echo-free zones. (T: tumor, LV: left ventricle, Ao: aorta).

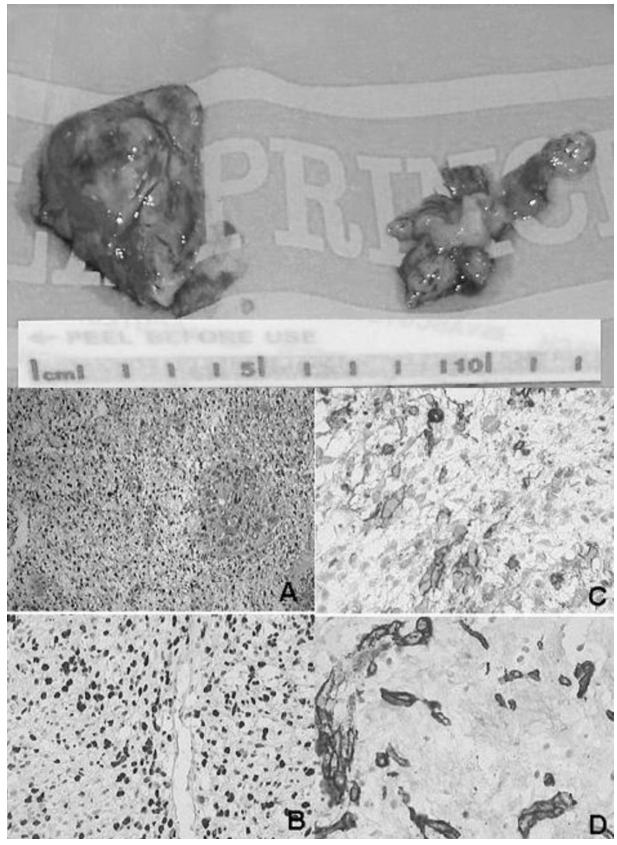

FIG. 2 **2a:** Macroscopic appearance **2b:** Histologic study: (A) Myxosarcoma with rounded cells areas into a mixoid stroma, distributed around vascular structures. (Masson trichromic 10x). (B) Growing fraction more than 50%. Immunohistochemical study with CD34 expression in a myxoma (D) and weaker, but significant in a myxosarcoma (C).

brain as the most affected organ.[8] Myxomas are usually smaller, multilobated and with noncompact appearance. Although Magnetic Resonance could give us more data suggesting malignancy,[9] our experience shows that image techniques are insufficient for a definitive diagnosis; the histologic study is mandatory.[8,10]

Resection is possible only in half of cases, and the most common cause of death is local recurrence (50%). Radiation and chemotherapy can be used with or without surgery but they do not prevent local recurrences and metastasis. Prognosis is poor with a mean survival time of about 1 year after diagnosis.[11–13] Cardiac transplantation could be an option but the reported cases have discouraging results.[14–18]

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