CASE REPORT

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Right atrium myxoid chondrosarcoma

Condrossarcoma mixóide de átrio direito

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Abstract

We are reporting a case of a 46-year-old woman, Caucasian, with hypertension and a primary diagnosis of infectious endocarditis. A transthoracic echocardiogram was performed, suggesting right atrium myxoma. The patient was submitted to surgery, which found a tumor mass with a jelly-like exterior. The mass was sent for anatomopathological analysis, which diagnosed a myxoid chondrosarcoma tumor. After the surgical resection, the patient achieved complete recovery with no signs of recidivation after 14 months.

Descriptors: **Heart neoplasms. Heart atria/surgery. Chondrosarcoma.**

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Resumo

Relatamos o caso de uma paciente de 46 anos, cor branca, hipertensa há 20 anos, com suspeita de endocardite infecciosa. Foi realizado ecocardiograma transtorácico, o qual levou à hipótese de mixoma de átrio direito. A paciente foi submetida à cirurgia, observando-se massa tumoral de aspecto muco-gelatinoso friável. A massa foi submetida a congelação para exame anatomopatológico, com laudo sugestivo de tumor maligno mesenquimal. Foi realizado exame imunohistoquímico compatível com condrossarcoma mixóide. A paciente evoluiu com remissão espontânea do quadro após ressecção completa da neoplasia. Realizou acompanhamento ambulatorial por 14 meses, sem apresentar sinais de recidiva do tumor.

Descritores: **Neoplasias cardíacas. Átrios do coração/ cirurgia. Condrossarcoma.**

INTRODUCTION

Primary cardiac tumors present epidemiological patterns that are practically established, although the symptomatology is not specific. The symptoms of the patients, when presented, are imprecise, suggesting myocardial, pericardial or valvular diseases [1]. These tumors are rare, with a serial incidence of necropsy range of 0.0017% to 0.28%, being about 20 times less frequent than heart metastatic tumors [1]. The differentiation between primary and metastatic tumors must be established according to the clinical context of the patient [2].

Concerning the benign or malignant aspect, metastatic tumors can show invasive and penetrating images and involvement of more than one cavity and mediastinal invasion. However, the definitive distinction often occurs during surgery or necropsy.

CASE REPORT

A 46-year-old female patient, Caucasian, with over 20 years of hypertension, arrived at the Hospital das Clínicas of the Federal University of Uberlândia with a primary diagnosis of infectious endocarditis due to a transthoracic echocardiographic image suggestive of tricuspid valve vegetation. Another transthoracic echocardiogram was performed, which suggested the existence of right atrium myxoma. During her period of stay in the hospital, the patient developed severe thrombocytopenia (14 to 49 thousand) and microcytic hypochromic anemia. The hematologial presentation of the patient was investigated through iliac crest myelogram with discreet results of megakaryocytic hyperplasia and absence of medullary iron clusters, maintaining a non-conclusive platelet presentation. The patient underwent surgery with the establishment of hypothermia at 28ºC with myocardial protection through infusion of cold blood cardioplegia solution at 4ºC with warm reperfusion.

After right atrium incision, a tumorous mass with friable gelatinous mucus was observed (Figure 1), measuring approximately 8cm (Figure 2). It was submitted to lyophilization for anatomopathological examination. The report suggested malignant mesenchymal tumor.

An immunohistochemical examination was performed with an avidin-biotin-peroxidase technique compatible with myxoid chondrosarcoma. The patient developed spontaneous remission of the thrombocytopenic presentation after complete resection of the neoplasia. An outpatient follow-up was performed after 14 months without signals of tumor recurrence.

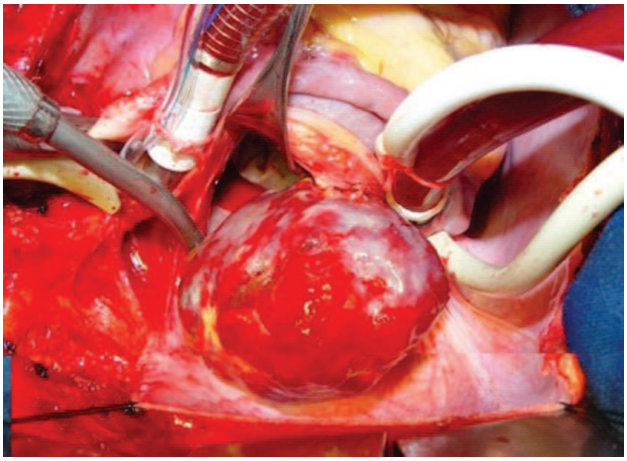

Fig. 1 – Intraoperatory aspect of neoplasia after atrial opening

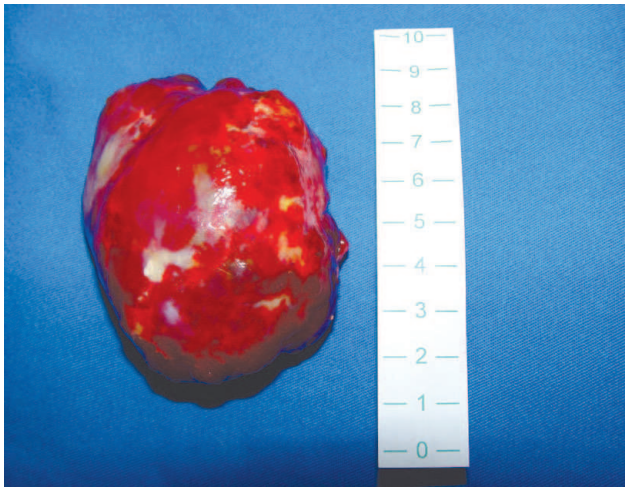

Fig. 2 – Aspect and measurement of neoplasia after complete resection

DISCUSSION

Primary heart sarcomas are rare, constituting less than 25% of all cardiac neoplasias; however, among the malignant tumors, sarcomas are the more frequent histological type, and they present dissemination potential and local invasion [3]. The right chambers are the most common places of origin of the sarcomas [4]. Lynch et al. [5] have shown that cardiac tumors are predominant in women, being less frequent in the ventricles and more common in the right atrium. When they begin their growth on the atrium's external side, they are usually malignant tumors.

Sarcomas are derived from mesenchymal tissue, and they have variable morphology, being that all histological types can be found in the cardiac site [6]. Difficulty in receiving treatment and rapid progression to death are characteristics of sarcomas [7].

Extraskeletal myxoid chondrosarcoma affects adults in the 3th and 5th decade of life, with a predominance in men. The majority of lesions are located in a deep site, consistently intramuscular and mainly in lower limbs [8].

Primary cardiac chondrosarcoma is extremely rare and possibly derived from multipotent mesenchymal stem cells that underwent a malignant differentiation process in cartilage [9].

There are few cases in the medical literature about primary cardiac chondrosarcoma [10].

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