Primary chondrosarcoma of the heart

Chirag Parmar*, Annie Jojo, Krishna C. Vachhani, Seethalekshmy N. Vijayan

Department of Pathology, Amrita Institute of Medical Sciences and Research Centre, Cochin 682026, Kerala, India

Received 10 October 2007; received in revised form 27 November 2007; accepted 28 November 2007

Abstract

Primary cardiac chondrosarcoma is extremely rare with very few cases reported in the literature. Most cardiac chondrosarcomas described in the literature are secondary. We report herein a case of 22-year-old man who presented with easy fatigability, which the general practitioner diagnosed as mitral stenosis clinically. Echocardiography showed left atrial mass, possibly myxoma. The histopathology of the excised mass revealed mesenchymal chondrosarcoma, confirmed by immunohistochemistry. Clinical and radiological studies did not reveal any other neoplasm.

Keywords: Cardiac chondrosarcoma; Mesenchymal chondrosarcoma

1. Introduction

Primary malignant tumours of the heart are uncommon comprising mainly of primary sarcomas, predominantly angiosarcoma, fibrosarcoma, and rhabdomyosarcoma [1,2]. Chondrosarcomas of the heart are very rare and the majority of them are secondary, wherein the primary site can be identified. Metastatic cases are usually located in right atrium [3]. Primary chondrosarcoma of the heart has not been described in WHO classification of cardiac tumours [4].

2. Case report

A 22-year-old student presented with complaints of easy fatigability and dyspnoea on exertion of 4 months duration. Patient was evaluated at AIMS and on examination pulse rate was 108/min, blood pressure of 110/84 mmHg and normal S1, loud S2 and P2. Presystolic murmur was detected at apex. Abdominal examination revealed tender hepatomegaly of 2 cm. Other systems of the heart were within normal limits. Echocardiography showed a mass in left atrium, possibly myxoma. After 2 weeks the patient developed secondary mitral regurgitation. Imaging modalities did not reveal any tumour other than in the heart. Surgical resection of the tumour was done through median sternotomy approach. Perioperatively the mass was sessile, located on the posterosuperior wall of left atrium, just above the mitral valve annulus and seemed to infiltrate the posterior wall of left atrium. The tumour was shaved off from the left atrial wall.

3. Materials and methods

The tissues were fixed with 10% formalin, 4–5 μm thick paraffin-embedded tissue sections obtained and stained with Harris’ hematoxylin and eosin stain. They were immunostained with the (pre-diluted) antibodies (Dako) for vimentin, smooth muscle actin, desmin, S-100 protein and CD-99 with positive control for each antibody and a negative control, using a streptavidin–biotin kit and the avidin–biotin–peroxidase method. MIB-1 (Ki-67, Dako, 1:50 dilution) labeling index was determined as a percentage of positive-staining tumour cell nuclei. The labeling index was determined in the area of the tumour with maximum immunopositivity (Fig. 1).

4. Histopathology

The resected mass consisted of greyish-white irregular bits, firm in consistency and measuring 6 cm × 3 cm × 2 cm in aggregate. Cut section showed lobulation and pearly white glistening appearance. Histopathologically, it was composed of two components: the first one consisted of islands of cartilage, with features of low-grade malignancy and the second being sheets of undifferentiated, small, round to spindle shaped cells exhibiting high nucleocytoplasmic ratio, variable nuclear density and mitotic figures (8–10/10 high

* Corresponding author. Tel.: +91 484 2801234x6308.
E-mail addresses: drchiragp@yahoo.com (C. Parmar), krishnacv@aims.amrita.edu (K.C. Vachhani).
power fields). These cells were seen in haphazard and vague fascicular arrangement in a myxoid background. The cartilaginous areas were sharply demarcated and merged gradually into surrounding small cell areas. Characteristic hemangiopericytoma-like vessels were found in the cellular areas. Necrosis, areas of haemorrhage or osteoid formation were not seen. Periodic acid-Schiff (PAS) stain was negative in areas with undifferentiated cells, whereas the neoplastic chondroid cells showed PAS positive cytoplasm. On immunohistochemistry, spindle and round undifferentiated (mesenchymal) cells expressed uniform positivity for Vimentin, CD-99 and SMA antibodies; whereas the chondroid cells were S-100, Vimentin and Desmin positive (Fig. 2).

On follow-up, the neoplasm recurred after 2 months, infiltrating the majority of mitral valve and occupying almost all of the left atrium. The patient died of the disease at 3 months of the presentation.

5. Discussion

Primary cardiac tumours are rare and usually benign [5]. Of malignant cardiac tumours, metastases from elsewhere are 30–40 times more common than primary malignancies. The most common subtypes are angiosarcoma and malignant fibrous histiocytoma, with primary cardiac leiomyosarcomas representing only 8–9% of all cardiac sarcomas [5].

Chondrosarcoma is a malignant tumour of cartilaginous tissues that has been exceptionally described in the heart. Most chondrosarcomas of the heart described in the literature are secondary [3] and in such cases the metastatic tumour is found in right-sided heart [6]. Primary chondrosarcoma of the heart is presumed to arise from multipotent mesenchymal stem cells that undergo malignant cartilaginous differentiation [7]. The tumour frequently originates from the endocardium, grows into the atrial or ventricular cavity and then, infiltrates into myocardial wall and extends to mediastinal structures [7].

In our case, imaging modalities, including skeletal survey did not reveal any tumour elsewhere in the body. Thus, it is indisputable that the tumour presented here is cardiac in origin. As for histology, the tumour was essentially a mesenchymal chondrosarcoma. This is a rare variant of chondrosarcoma, characteristically bimorphic in appearance, composed of sheets of primitive mesenchymal cells and islands of hyaline cartilage. It usually affects young adults, while conventional chondrosarcoma has a peak incidence in the fifth to seventh decade of life. The majority of cases arise in the axial skeleton or cranial bones, although 20–30% are located in extraskeletal sites such as somatic soft tissues and the meninges [7].

The prognosis of patients with cardiac chondrosarcoma is poor; survival is measured in weeks or months [7,8]. Radical surgery is the mainstay of treatment. A variety of primary chondrosarcomas of the bone are low- to intermediate-grade tumours with indolent clinical behaviour and show 72.7% of the overall 5-year survival rate [9]. Irradiation and chemotherapy are indicated for infiltrating lesions, which are not suitable for ablative surgery. However, these latter therapies are usually ineffective [10]. Mesenchymal chondrosarcoma has a high risk of local recurrence and seems to metastasize in unusual sites (i.e. liver, lymph nodes, kidneys, skin and brain) more frequently and more rapidly than other chondrosarcomas [10,7]. These characteristics imply careful follow-up of the patient.

In our patient, the clinical course was aggressive with a 3-months survival following the development of symptoms.
References


