Extraskeletal mesenchymal chondrosarcoma involving the heart: report of a case

Gabriella Nesi, Elena Pedemonte, Franca Gori

Department of Pathology, University of Florence, Florence, Italy

Key words:
Heart neoplasms;
Chondrosarcoma.

Extraskeletal mesenchymal chondrosarcoma is a rare tumor frequently arising in the meninges and lower limbs. We describe a case of mesenchymal chondrosarcoma involving the heart in a 39-year-old man who presented with fever, chest pain and shortness of breath. His clinical course was rapid, leaving insufficient time for a complete diagnostic work-up, and the patient died 2 months after the onset of symptoms.

(Ital Heart J 2000; 1 (6): 435-437)

Received April 5, 2000,
accepted May 11, 2000.

Address:

Prof.ssa Franca Gori

Dipartimento di Patologia Umana
Università degli Studi
Viale Morgagni, 85
50134 Firenze

Introduction

Cardiac involvement by either primary or secondary chondrosarcoma is a rare event[1,2]. We report a case with such an aggressive clinical course that the correct diagnosis could only be made postmortem.

Case report

A 39-year-old man was admitted with a diagnosis of viral pneumonia. Medical management was undertaken and, on his satisfactory recovery, he was discharged after 3 weeks. One week later he suffered from an episode of severe right lumbar pain. Bone survey was negative. One month after discharge the patient was readmitted for prolonged chest pain. An electrocardiogram showed sinus tachycardia and ST segment elevation. Echocardiography and ventriculography revealed a filling defect in the right ventricular chamber extending to the pulmonary outflow tract (Fig. 1). A computed tomography scan of the chest confirmed these findings and detected the presence of mediastinal lymphadenopathy, suggesting a diagnosis of lymphoma. During this diagnostic work-up, small, hard lumps appeared under the skin of his trunk and scalp. A skin biopsy was carried out, but before histological diagnosis could be made, the patient's clinical course rapidly deteriorated and he died of respiratory failure 2 months after onset of pulmonary symptoms.

Pathological findings. At autopsy, the heart weighed 650 g. A large tumor of the anterior right ventricular wall was found protruding into the cavity and obstructing the right outflow tract (Fig. 2). Numerous neoplastic nodules were present on the skin, in the lungs, the right kidney and the spleen. A large mass, measuring 10 u 7 u 4 cm, was located in the retroperitoneum, next to the right iliac bone. Multiple bilateral pulmonary infarcts due to neoplastic emboli, foci of pneumonia and acute pulmonary edema were also found. On cut sections, all tumor masses were pearly-white and firm, with a glistening surface.

Histologically, the lesions were characterized by the presence of two main components: islands of cartilage having features of low-grade malignancy and showing occasional calcifications, and sheets of undifferentiated, small, round- to spindle-shaped cells exhibiting a high nucleo-cytoplasmic ratio and variable nuclear density (Figs. 3 and 4). Mitotic figures were frequently seen only in the small cell component. The cartilaginous areas were sometimes sharply demarcated and sometimes they merged more gradually into the surrounding small cell areas. Necrosis was a prominent feature. The undifferentiated cells displayed vimentin positivity and periodic acid-Schiff (PAS) stains failed to demonstrate any significant amounts of intracellular glycogen. The neoplastic chondroid cells had PAS positive cytoplasms and were reactive for S-100 protein. In

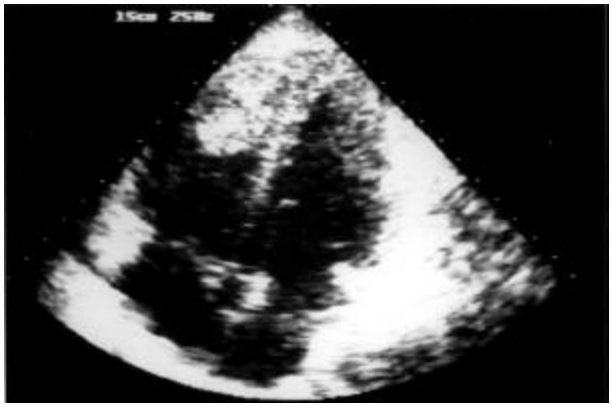

Figure 1. Echocardiogram (apical 4-chamber view optimized for the right sections) showing a large echogenic mass in the left ventricle.

Figure 2. A section through the right ventricular outflow tract shows the tumor mass reaching the pulmonary valve.

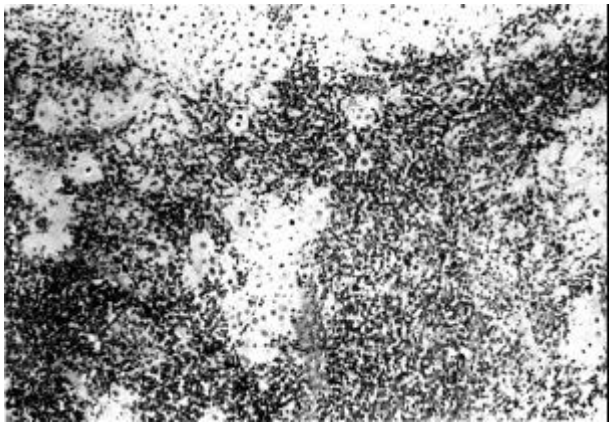

Figure 3. Typical bimorphic proliferation in mesenchymal chondrosarcoma: sheets of small cells and islands of well-differentiated cartilage (H&E, 100u).

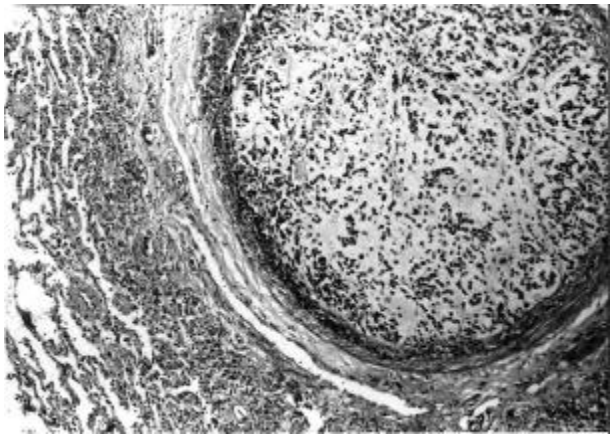

Figure 4. Neoplastic emboli obstructing some branches of the pulmonary artery (H&E, 100u).

some sections, the bimorphic proliferation was spotted by foci of small cells, arranged in short anastomozing strands separated by mucoid material, and of aggregates of large, markedly atypical, sarcomatous cells. The diagnosis of mesenchymal chondrosarcoma was made.

Discussion

Chondrosarcoma is a malignant tumor of cartilage-forming tissues that has been exceptionally described in the heart. In a review of the literature published in 1998, Fichaux et al.[2] found 21 cases of chondrosarcoma of the bone with myocardial metastases, to which they added one more case. From the overall cases reported to date, heart involvement occurs late in the clinical course, in the presence of widespread disease, and affects the right atrium[3].

Chondrosarcoma is even more rarely encountered as a primary tumor of the heart and great vessels. In such instances, it is presumed to arise from multipotent mesenchymal stem cells that undergo malignant differentiation into cartilage. Only 6 cases of primary cardiac chondrosarcoma have been documented so far[1,4-8]. The tumor frequently originates from the endocardium and grows into the atrial or ventricular cavity. Then, it progresses to myocardial wall infiltration and to mediastinal tissue invasion[7].

The prognosis of patients with cardiac chondrosarcoma, both primary and secondary, is poor; survival is measured in weeks or months. Radical surgery is the mainstay of treatment, while irradiation and chemotherapy are indicated for infiltrating lesions not suitable for ablative surgery. However, these latter therapies are ineffective[1,3].

In our case, the bone survey and autopsy findings ruled out the presence of a primary bone neoplasm. Although a single large mass was found in the retroperitoneum, the remarkable involvement of the right ventricle with obstruction of both the outflow tract and the pulmonary valve strongly suggests the cardiac origin of the tumor. As for histology, the tumor was essentially a mesenchymal chondrosarcoma[9]. This is a rare variant of chondrosarcoma, characteristically bimorphic in appearance, composed of sheets of primitive mesenchy-

mal 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 to 30% are located in extraskeletal sites, such as somatic soft tissues and the meninges. Mesenchymal chondrosarcoma has a high risk of local recurrence and seems to metastasize to unusual sites (i.e. liver, lymph nodes, kidneys, skin and brain) more frequently and more rapidly than other chondrosarcomas.

To the best of our knowledge, the present case represents the first instance of mesenchymal chondrosarcoma of the heart to be described in the English literature. Pneumonia was the first clinical sign of the tumor and was related to pulmonary neoplastic embolism, whereas lumbar pain was presumably due to the retroperitoneal mass next to the iliac bone. The clinical course was aggressive with a 2-month survival following the development of symptoms.

References

1. Miwa S, Konishi Y, Matsumoto M, Minakata K. Primary cardiac chondrosarcoma: a case report. Jpn Circ J 1997; 61: 795-7.
2. Fichaux O, de Muret A, Dessenne X, et al. Cardiac metastasis of chondrosarcoma: a case report. Ann Cardiol Angeiol (Paris) 1998; 47: 165-8.
3. Leung CY, Cummings RG, Reimer KA, Lowe JE. Chondrosarcoma metastatic to the heart. Ann Thorac Surg 1988; 45: 291-5.
4. Winer HE, Kronzon I, Fox A, et al. Primary cardiac chondromyxosarcoma - clinical and echocardiographic manifestations. A case report. J Thorac Cardiovasc Surg 1977; 74: 567-70.
5. Bashi VV, Walter A, John S. Primary chondrosarcoma of heart. Indian J Chest Dis Allied Sci 1989; 31: 203-6.
6. Murakami A, Hamanaka H, Ueyama T, Kitagawa M, Asanoi H, Sumitani T. Primary cardiac malignant mesenchymal tumor containing chondrosarcomatous component: a case report. Nippon Kyobu Geka Gakkai Zasshi 1991; 39: 361-5.
7. Ng SH, Ko SF, Yang TS, Wong HF, Wan YL, Ho YS. Primary cardiac chondrosarcoma. Am J Emerg Med 1996; 14: 285-7.
8. Tsai FC, Lin PJ, Wu WJ, Kuo TT, Chang CH. Primary chondrosarcoma of the heart: a case report. Chang Keng I Hsueh 1996; 19: 348-51.
9. Enzinger FM, Weiss SW. Soft tissue tumors. 3rd edition. St Louis, MO: Mosby, 1995: 1004-8.