Exertional Dyspnea and Left Atrial Mass

Kambiz Mozaffari*, Farzad Yazdani Biucki,** Gholamreza Omrani,*** Jafar Hashemi,† and Ahmad Amini††

Abstract

We present a 56-year-old woman who came to the emergency department with an exertional dyspnea of recent onset. A transthoracic echocardiographic examination revealed two round masses in her left atrium with mild mitral stenosis and mild-to-moderate mitral valve regurgitation.

Her laboratory data were unremarkable except for a mild anemia. Surgical excision of the masses was performed, and two creamy-white fleshy tumors were removed. On cross section, they were solid and creamy-brown with gritty areas. Histopathological examination showed extensive sheets of round to oval cells, and hemangiopericytoma-like patterns. Also, multiple lobules of well-differentiated hyaline cartilage were present. An immunohistochemistry (IHC) panel revealed that the chondroid areas were reactive for S100 protein. The round cells expressed CD99 with focal positivity for NSE (neuron-specific enolase), but were negative for the following cytokeratin, CD34, factor VIII, actin, and desmin. Therefore, the cells were mesenchymal in origin with chondroid differentiation, and the final diagnosis was a mesenchymal chondrosarcoma.

A full-scale investigation into the source of the tumor was unrevealing. Shortly after her discharge from the hospital, she developed an embolic cerebrovascular accident ***(Iranian Heart Journal 2010; 11 (3):47-49).***

Key words: heart tumor ■ sarcoma ■ dyspnea

rimary cardiac tumors are rare. Tumors of mesenchymal tissue are the most diverse and are further subdivided by tissue type. They are histologically similar to those arising in extracardiac soft tissues.[1] We present a 56-year-old woman who came to the emergency department with exertional dyspnea of recent onset, with two round masses in her left atrium.

Case report

A 56-year-old woman came to the emergency department with an exertional dyspnea of recent onset. In light of the abnormal heart examination, a transthoracic echocardiographic study was performed, which revealed two round masses in her left atrium.

The first one was attached to the inter-atrial septum and the other to the posterior leaflet of the mitral valve, with resultant mild stenosis and mild-to-moderate valve regurgitation. Transesophageal echocardiography confirmed these findings and showed a suspected mass density with a turbulent flow in the right lower pulmonary vein.

Angiography revealed normal epicardial coronary arteries and contrast-enhanced tumor vascularity. Her laboratory data were unremarkable except for a mild normochromic normocytic anemia (Hb 11g/dL). Surgical excision of the lesions was performed, and acceptable free surgical margin. The right lower pulmonary vein seemed to be uninvolved.

The specimen consisted of brownish pieces that measured 6×6×3 cm. On cutting, they were solid and creamy-brown in appearance with areas gritty in consistency.

Histopathological evaluation showed extensive sheets of plump, round to oval cells with moderate pleomorphism. A few mitotic figures and areas with a hemangiopericytoma-like pattern were noted (Fig. 1).

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*Assistant Professor of Pathology, Shaheed Rajaie Cardiovascular, Medical and Research Center ,**Assistant Professor of Pathology, Tehran University of Medical Sciences, ***Associate Professor of Cardiac Surgery, †Associate Professor of Cardiology, and ††Assistant Professor of Cardiology, all at Shaheed Rajaie Cardiovascular, Medical and Research Center.

Correspondence to: Kambiz Mozaffari MD, Surgical Pathology Laboratory, Shaheed Rajaie Cardiovascular, Medical and Research Center, Meliat

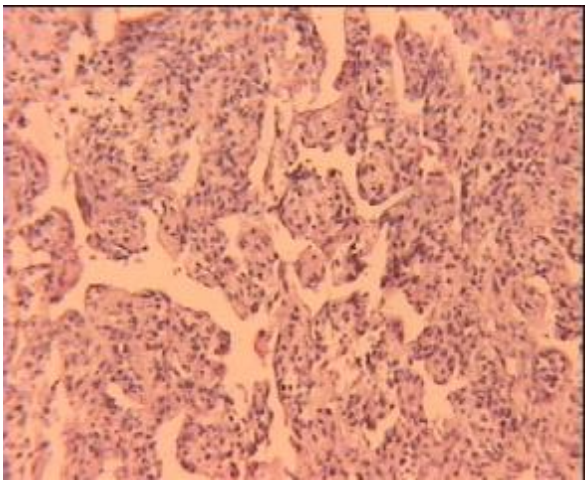

Fig. 1. Photomicrograph showing plump, round to oval cells with a hemangiopericytoma-like pattern (H&E, X 400).

No areas of tumoral necrosis were seen. Also, multiple lobules of well-differentiated hyaline cartilage were present (Fig. 2).

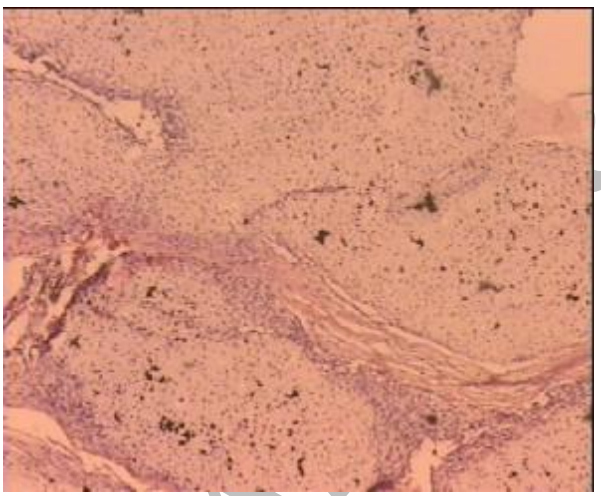

Fig. 2. Photomicrograph of the tumor depicting lobules of well-differentiated hyaline cartilage (H&E, X 400).

Based on histomorphology, our diagnosis was a bimorphic malignant cartilage-forming tumor, namely an extra-skeletal mesenchymal chondrosarcoma. Malignant mesenchymoma could also be a differential diagnosis.

Therefore, an immunohistochemistry (IHC) panel was requested for the confirmation of our diagnosis. Immunohistochemically, the chondroid areas were reactive for S100 protein. The plump, round cells expressed CD99 and showed focal positivity for NSE (neuron-specific enolase), but were negative for cytokeratin, CD34, factor VIII, actin, and desmin.

A full-scale investigation into the source of the tumor was also carried out (i.e. whole body scan, thoracoabdominal CT scan) as well as a complete workup regarding the thyroid gland, breasts, and uterine adnexae. All the studies were negative. Shortly after her discharge from the hospital, she developed a cerebrovascular accident, which was found to be embolic in nature.

The patient's deteriorating condition precluded the use of chemoradiation.

Discussion

Primary sarcomas of the heart are histologically similar to those arising in extracardiac soft tissue.[1] They are rare and aggressive malignancies, usually diagnosed late due to their non-specific symptoms.[4] Those of mesenchymal origin are the most diverse and are further subdivided by tissue type. The largest group of such primary mesenchymal sarcomas demonstrates fibroblastic or myofibroblastic differentiation, but angiosarcomas, leiomyosarcomas, rhabdomyosarcomas, and liposarcomas are also the other types of neoplasms encountered in this organ. It is also possible for the heart to harbor those lesions with malignant osteoid and chondrosarcoma.[1] In fact, malignant osteoid and areas of chondrosarcoma are not rare in left atrial sarcomas. More than half the cardiac sarcomas are located in the left atrium.[3]

Here, we are dealing with a malignant sarcoma that is biphasic in appearance, namely the lobules of hyaline cartilage as well as the round cells that are arranged as extensive sheets, some of which have formed discrete hemangiopericytoma-like patterns.[2]

This biomorphic pattern, consisting of undifferentiated round to oval cells that look like embryonal mesenchyme as well as the well-differentiated cartilaginous areas, establishes a characteristic finding.

IHC results are positive for S100 protein in the cartilage-forming areas and negative for cytokeratin, actin, desmin, CD34, and factor

VIII.[5] As was mentioned in our case, the IHC findings were confirmatory.

Malignant cartilaginous tumors not connected to bone are named extraskeletal chondrosarcomas.[5] They predominately occur in the head and neck region, as well as in the lower extremities.[2, 6] Nevertheless, fewer than twenty cases of cardiac mesenchymal chondrosarcoma have so far been reported.[6]

Extraskeletal chondrosarcoma is a rare malignancy and represents approximately 2% of all soft tissue sarcomas.[7]

Although many predisposing factors have been listed including genetic diseases, radiation exposure, chemicals, lymphedema, and trauma, no specific etiology has hitherto been found.

Lightenstein and Bernstein were the ones who first described this form of chondrosarcoma in 1959, and Dowling reported the first extraskeletal mesenchymal chondrosarcoma in 1964.[5, 8]

Mesenchymoma, which was considered a differential diagnosis, is a tumor that may differentiate along several cell lines. This tumor may contain cartilage, bone, small cells, and skeletal muscle tissues. To prevent further confusion, it is even believed that this term will eventually be abandoned by surgical pathologists.[3]

Despite the fact that our patient could not benefit from chemoradiation, complete excision with adjunctive systemic chemotherapy and local radiotherapy is the recommended treatment. Frequent and thorough follow-up examinations are needed because of the highly malignant nature of the tumor.[5]

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