An 18-Year-Old Man with Cardiomegaly and Pericardial Effusion

Kambiz Mozaffari MD[1], Farzad Yazdani Biucki MD[2], Ahmad Amin MD[3] and Katayoun Rahmatabadi MD[4]

Abstract

We present an 18-year-old male who sought medical attention due to exertional dyspnea of a few weeks' duration. His physical exam revealed an elevated jugular venous pressure, facial puffiness, muffled heart sounds, and mild lower extremity pitting edema.

Chest X-ray showed cardiomegaly; and in echocardiography, huge intrapericardial masses with massive pericardial effusion were noted. The only noteworthy finding on abdominal ultrasonography and CT scan was the presence of ascites. The only abnormal laboratory results consisted of a hemoglobin level of 10.8 g/dl, a 1-hr ESR 77, CRP 34 mg/dl and LDH 771. Some 1500cc pericardial fluid was aspirated, and two multilobated creamy-brown masses with foci of necrosis and hemorrhage were excised.

Microscopically, hypercellular sheets of malignant round cells were seen. Based on morphology, a diagnosis of high-grade round cell sarcoma was made. Immunohistochemical markers were negative for cytokeratin, CD 34, desmin, and smooth muscle actin, while positive reactivity was noted only for vimentin. Therefore, the cells were mesenchymal in origin with no vascular, skeletal, or smooth muscle differentiation and the final diagnosis was undifferentiated sarcoma.

The patient was discharged in good clinical condition and underwent chemoradiation therapy***(Iranian Heart Journal 2009; 10 (4):45 -48).***

Key words: cardiac tumor ■ undifferentiated sarcoma ■ cardiomegaly

Primary cardiac tumors are rare. Tumors of mesenchymal tissue are the most diverse and are further subdivided by tissue type and may be benign or malignant.[1] Primary cardiac sarcoma (PSC) is a rare and aggressive malignancy that is usually diagnosed late due to its nonspecific symptoms.[4] They are similar histologically to those arising in extracardiac soft tissue. The largest group demonstrates fibroblastic or myofibroblastic differentiation. Other categories include angiosarcomas, leiomyosarcomas, and rhabdomyosarcomas, but a high proportion defy classification and are, therefore, dealt with as undifferentiated sarcomas.[1]

We present an 18-year-old boy with exertional dyspnea of a few weeks' duration who had cardiomegaly with massive pericardial effusion. Based on our findings, a high grade round cell sarcoma was noted. Most immunohistochemical markers were negative, and positive reactivity was only noted for vimentin.

Case report

We present an 18-year-old man who came to seek medical attention due to exertional dyspnea.

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[1]Assistant Professor of Pathology, Shaheed Rajaie Cardiovascular Medical and Research Center (Corresponding author)
[2]Assistant Professor of Pathology, Tehran University of Medical Sciences
[3]Assistant Professor of Cardiology, Shaheed Rajaie Cardiovascular Medical and Research Center
[4]Clinical and Anatomical Pathologist, Surgical Pathology and IHC Dept, Iranian Blood Transfusion Organization

Also among his complaints were facial swelling and edema in the lower extremities, which had developed over the previous few weeks. His past medical history, however, was unremarkable except for a recent upper respiratory tract infection.

His vital signs included a blood pressure of 120/80 mmHg and PR: 105/min, but no fever was detected. On physical examination, the remarkable findings consisted of an elevated jugular venous pressure, facial puffiness, muffled heart sounds, and a mild lower extremity pitting edema.

Chest X-ray films revealed cardiomegaly and a normal pulmonary vasculature. Transthoracic echocardiography showed a huge intra-pericardial mass located posteriorly to the ventricles with probable intra-atrial extension, accompanied by a massive compressive pericardial effusion. The abdominal ultrasonography and the CT scan of the abdominopelvic cavities were unremarkable except for the presence of ascites.

The patient's laboratory findings were all within the normal range, except for a hemoglobin level of 10.8 g/dl, a 1 hr ESR of 77 mm, CRP 34 mg/dl (normal range up to 10 mg/dl), and his serum LDH level was 771 IU/l (normal range up to 480 IU/L).

The patient underwent surgery, during which some 1500mL sero-sanguineous fluid was first aspirated and then two masses were removed: the first one measured 20x6x5 cm, located within the pericardial space with attachment to the inferior surface of the right atrium and the second mass measured 4x5x6 cm, originating from the right atrium. Cytological study of the pericardial fluid showed a bloody effusion that was negative for malignancy.

Grossly, the specimens consisted of multilobated creamy-brown masses with a variegated appearance and firm-to-solid consistency (Fig. 1).

Fig. 1. Multilobated creamy-brown masses with a variegated appearance

The cut surface was predominantly solid and creamy with foci of necrosis and hemorrhage.

Microscopically, hypercellular sheets of malignant round cells were seen (Fig. 2) that had vesicular nuclei or more often they looked like plump oval cells (Fig. 3) with perivascular cellular accentuation and slit-like vessels in between.

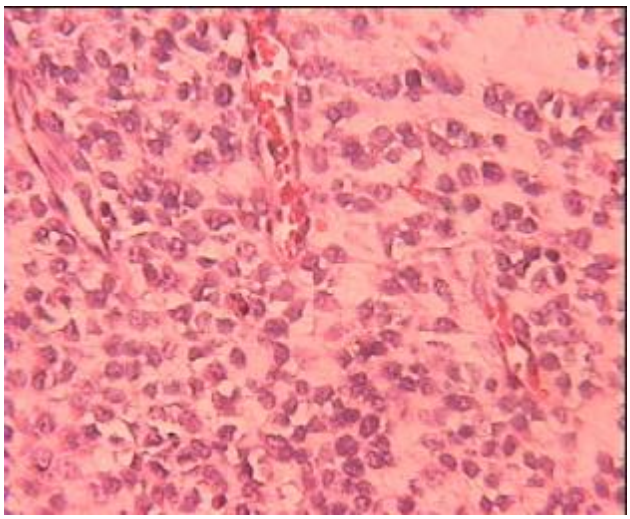

Fig. 2. Hypercellular sheets of malignant round cells (H&E, X400).

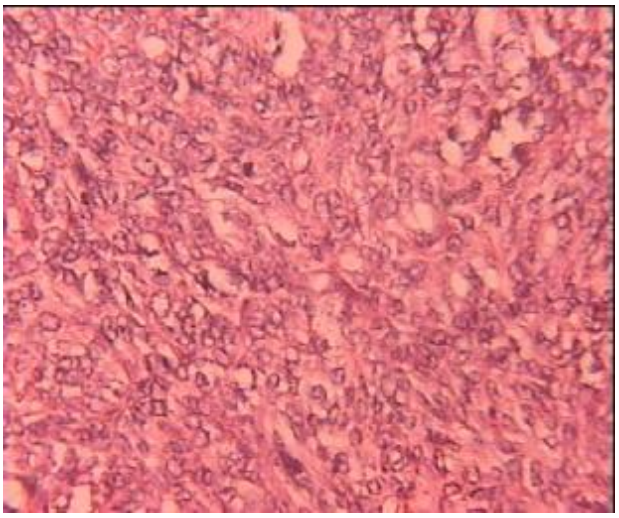

Fig. 3. Areas of tumor with plump oval cells and vesicular nuclei (H&E, X 400)

Areas of tumoral necrosis and hemorrhage were readily evident. Based on the histopathological findings, a diagnosis of high grade round cell sarcoma was made.

The histopathological differential diagnosis of a malignant round cell tumor in the heart lies between the following: angiosarcoma, leiomyosarcoma, rhabdomyosarcoma, synovial sarcoma (monophasic type), liposarcoma and an unclassified sarcoma. To further assist the diagnostic approach, a panel of immunohistochemical markers (IHC) was requested which showed negative results for cytokeratin (CK), CD 34, desmin, and smooth muscle actin (SMA). The tumoral cells revealed reactivity only for vimentin. Based on the IHC findings, the neoplastic cells were mesenchymal in origin with no vascular, skeletal, or smooth muscle differentiation and the final diagnosis was undifferentiated sarcoma.

The patient was discharged in good clinical condition and was referred to receive multiple chemoradiotherapy courses.

Discussion

Primary cardiac tumors are rare. The most common is the cardiac myxoma, which constitutes nearly 80% of surgically-excised masses. Tumors of mesenchymal tissue are the most diverse and are further subdivided by tissue type and may be benign or malignant.[1]

Primary cardiac sarcoma (PCS) is a rare and aggressive malignancy that is usually diagnosed late due to its nonspecific symptoms.[4] Primary sarcomas of the heart are similar histologically to those arising in extracardiac soft tissue. The largest group of primary cardiac sarcomas demonstrates fibroblastic or myofibroblastic differentiation. Another category is angiosarcoma with malignant endothelial cells that form vascular channels.

Leiomyosarcomas are composed of fascicles of spindle cells that intersect one another at right angles. Rhabdomyosarcomas constitute 5% of cardiac sarcomas and the majority are of the embryonal type with numerous tadpole-shaped rhabdomyoblasts.

A high proportion of cardiac sarcomas defies classification and is, therefore, dealt with as undifferentiated sarcomas. The majority of these tumors occur in the atria, and their differential diagnosis primarily consists of metastatic tumors. Other rare sarcomas include liposarcoma, malignant peripheral nerve sheath tumor, and synovial sarcoma.[1]

Most patients with primary heart sarcomas present with intractable congestive heart failure, arrhythmias, or signs of superior vena cava obstruction. In rare cases, a metastatic lesion is the first manifestation of the disease.

It has been pointed out that malignant tumors are more frequently found in the right side of the heart and that benign neoplasms are more common on the left side.[2]

Other modes of presentation for cardiac sarcoma include pericardial tamponade, embolic phenomena, chest pain, syncope, fever of unknown origin, and peripheral edema.

The histological classification of cardiac sarcomas is currently of little clinical use. The treatment and survival are not apparently affected by the type of sarcoma. It has been shown that there is especially poor survival in patients with tumors that have a high mitotic

rate and areas of necrosis. For this reason, the surgical pathologist should note the presence of these features, which are indicators of high grade sarcomas.[3]

Cytology and cardiac biopsy may be negative, and suspicion for the tumor is warranted in recurrent pericardial effusion.[4]

Angiosarcomas arise mainly from the right heart and they have a preference for the atria.

They appear in two forms: 1) a large obstructing mass and 2) a locally infiltrative tumor.

Pericardial lesions simulate pericarditis. Well-differentiated areas of angiosarcoma show neoplastic blood vessels; nevertheless, in poorly-differentiated lesions, a solid growth pattern is seen with spindle-shaped and anaplastic cells. No vasoformative pattern is, therefore, noted and factor VIII related Ag is usually negative, but CD 31 and CD 34 are usually positive.[1-4]

Rhabdomyosarcoma has a polypoid appearance and may involve the pericardium.

They are positive for desmin and myogenin.

Synovial sarcoma is another rare differential diagnosis. The typical biphasic pattern is exceptional, but one may find the epithelial glandular structures as well as a cellular spindle-celled stroma. Keratin and epithelial membrane antigen (EMA) tend to be positive in this neoplasm.[1-4]

Last but not least, undifferentiated sarcoma is our final diagnosis when most helpful markers remain negative. The tumor may be positive for smooth muscle actin and vimentin, yet occasional cells may also be positive for cytokeratin. The differential diagnosis of this malignancy includes round cell liposarcoma, embryonal rhabdomyosarcoma, and primitive neuroectodermal tumor (PNET), which is positive for CD 99.[1-4]

Echocardiography is the major diagnostic tool and aids pericardiocentesis. Pericardial window may be useful for recurrent pericardial effusion but does not preclude its reaccumulation. There is no proven effective treatment for PCS, and treatments include surgical resection, cardiac transplant, chemotherapy, and radiotherapy. Despite its poor prognosis, symptomatic relief is important and attainable.[4]

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