Primary Cardiac Angiosarcoma Detected by Magnetic Resonance Imaging but Not by Computed Tomography

Moriaki Inoko, Kanji Iga, Katsuhide Kyo, Hirokazu Kondo, Toshihiro Tamura, Chisato Izumi, Shouji Kitaguchi, Toshiro Hirozane, Yoshihiro Himura, Hiromitsu Gen and Takashi Konishi

Abstract

A 51-year-old man with a primary angiosarcoma of the right atrium is reported. The angiosarcoma was not detected by transthoracic echocardiography or computed tomography, but magnetic resonance imaging and transesophageal echocardiography did show the tumor of the right atrial free wall. We performed a transvenous endomyocardial biopsy of the tumor under the guidance of transesophageal echocardiography and made the pathological diagnosis. This case demonstrates the advantage of magnetic resonance imaging and transesophageal echocardiography for tumor detection over transthoracic echocardiography and computed tomography and the usefulness of transesophageal echocardiography for guiding the right atrial endomyocardial biopsy procedure.

Key words: transesophageal echocardiography, transvenous endomyocardial biopsy

Introduction

We report a case of a primary angiosarcoma of the right atrium (RA), which was not detected by transthoracic echocardiography (TTE) or computed tomography (CT) but was demonstrated by magnetic resonance imaging (MRI) and transesophageal echocardiography (TEE).

Case Report

A 51-year-old man had been well until May 1998 when he developed dyspnea, abdominal distension, and facial and lower limb edema. Because TTE revealed a massive pericardial effusion, the patient was referred to Tenri Hospital on June 15. On admission, the pulse rate was 87/min, blood pressure was 120/90 mmHg, and a paradoxical pulse was found. Consciousness was clear and cyanosis was not observed. Heart sounds were normal with no murmurs. Coarse crackles in the basal lung fields and congestive hepatomegaly were noted. A chest X-ray examination revealed cardiomegaly, bilateral lung congestion and bilateral pleural effusion. TTE (model SSA-140A, Toshiba Medical Systems, Tokyo) showed massive pericardial effusion but no mass in the cardiac chambers or pericardial space. Electrocardiography on admission showed no abnormal findings. Blood tests showed an erythrocyte count of 420×10⁴/mm³; hemoglobin 13.2 g/dl; leucocyte count 6,900/mm³; platelet count 27.9×10⁴/mm³. Blood gas analysis revealed hypoxemia (PO₂ 78.2 mmHg, PCO₂ 38.4 mmHg, PH 7.454) in spite of inhalation of oxygen by a nasal oxygen cannula. Other blood chemistries showed liver injury (aspartate aminotransferase 71 IU (AST; normal range 11–32), alanine aminotransferase 180 IU (ALT; normal range 3–30), γ-glutamyl-transpeptidase 120 (γGTP; normal range 10–50) probably due to congestion. C-reactive protein was 2.6 mg/dl (normal range <0.2); prothrombin time 12.3 seconds (normal range 9.8–11.8); fibrin degradation products <0.5 μg/ml (normal range <0.5). The patient was severely ill, so we performed right heart catheterization and pericardiocentesis. The hemodynamics before the pericardiocentesis showed an elevation in the RA pressure (RAP; 21 mmHg), and a low cardiac index (CI; 1.7 l/min/m²). About 1,000 ml of bloody fluid was obtained by pericardiocentesis in which the hematocrit was 29.1% and protein was 5.9 g/dl, moreover the cytological examination was negative for malignant cells. After pericardiocentesis the patient felt better and his hemodynamic state improved (RAP, 12 mmHg: CI, 4.1 l/min/m²). Both plain and enhanced CT (X force, Toshiba Medical Systems, Tokyo, Japan; Contrast agent, bolus injection of 61.24 g of iopamidol) showed no evidence of malignancy either in or around the heart (Fig. 1), but T2-weighted MRI (1.5T Magnetom Vision, Siemens Medical Systems, Erlangen, Germany) by HASTE (Half-Fourier Acquisition Single-shot Turbo Spin Echo) revealed a high signal intensity mass in the anterior wall of the right atrium, and a bolus injection of 5.571 g of megilumine gadopentetate enhanced the signal intensity of the mass in T1-weighted turbo spin echo MRI.
Inoko et al

Figure 1. Axial images of computed tomography of the heart both with and without contrast enhancement. The cardiac tumor is undetectable.

(Fig. 2). TEE (model SSA-140A, Toshiba Medical Systems) demonstrated masses that extended from just above the tricuspid valve to just below the superior vena cava in the free wall of the right atrium (Fig. 3). The surface of the tumor was lobulated and the tissue characterization inside the tumor by TEE was homogenous. These characteristics both in MRI and in TEE were apparently different from cardiac thrombus. Angiographically, tufts of irregular vessels derived from the right coronary artery were observed. These findings suggested that the tumor was a malignancy of the heart. In a metastatic work-up, no metastasis was identified. To determine the pathological diagnosis, a TEE-guided biopsy of the tumor was performed with a transvenous catheter biopтомe introduced percutaneously from the right internal jugular vein. The biopтомe was easily positioned on the tumor surface by TEE guidance, and three biopsy samples of the tumor were immediately examined. These samples were reddish but were clearly different from a thrombus. A light microscopic examination revealed that the tissue was composed of immature blood vessels that contained erythrocytes and spindle-shaped cells with oval nuclei (Fig. 4). Mitosis was scattered throughout. The morphological appearance was compatible with the diagnosis of an angiosarcoma, and a subsequent surgical removal was undertaken. The sarcoma spread over the entire right atrial wall and into the endocardial side in some places. The sarcoma also invaded the epicardium, and the hemorrhage from the lesion was thought to have caused massive paracardial effusion (Fig. 5). Most of the right atrium was removed, and atrial reconstruction was performed with a Gore-Tex sheet under a total cardiopulmonary bypass. The postoperative course was uneventful. The pathological examination revealed sarcoma cells in the marginal zone of the removed tissues. Only 24Gy of irra-

plain

enhanced
Cardiac Angiosarcoma

Figure 2. Axial magnetic resonance imagings of the heart. T2-weighted images show multiple right atrial tumors with high signal intensity (arrows). Meglumine gadopentetate enhanced the tumor on T1-weighted images.

Figure 3. Transesophageal echocardiography showed lobulated masses in the free wall of the right atrium and the bioptome entering the right atrium from the superior vena cava and obtaining tumor tissue.

Figure 4. Light microscopic study of the biopsy specimen showing vascular structures with spindle-shaped cells and oval nuclei (HE stain, ×480).
radiation to the whole heart and mediastinum was performed because the patient suffered from bacterial pneumonia. The patient refused additional chemotherapy. Five months after the operation, the patient suffered from multiple metastases in the lumbar vertebrae, pelvis and both femurs. Additional radiation therapy was effective; however, twelve months after the surgery, the patient died due to metastases to the lung and subsequent pleural bleeding. An autopsy was not performed.

Discussion

The incidence of angiosarcomas of the heart is rare. However a review of the literature showed that the tumor is aggressive. In 88.4% of patients with a metastatic lesion at diagnosis, even in those with a primary angiosarcoma of the heart who were diagnosed intra vitam and underwent appropriate treatment, the mean survival time was 11.9 months (1). Although the antemortem diagnosis of a cardiac angiosarcoma is difficult and has been achieved in only 38.9% of all cases (1), it is becoming more frequent with the increasing use of modern imaging techniques. This patient was administered a cardiac tamponade, and the pericardiocentesis obtained a hemorrhagic effusion. We suspected malignant diseases and performed a work-up by transthoracic echocardiography and conventional CT. However, we were unable to obtain any evidence of malignancies. This was due to the following technical difficulties: the tumor was so thin that conventional CT could not show a clear image due to moving artifacts, and the angiosarcoma contained too much blood to make distinctions between the tumor and the blood in the RA cavity by both plain and enhanced CT. On the other hand, MRI did demonstrate clear images of the thin tumor by ECG-gating and clear distinctions between the tumor and the blood. Although it has been reported that MRI has more advantages for diagnosing a cardiac tumor by tissue characterization (2), the tumor was too thin to evaluate the heterogeneity. Nevertheless, some characteristics from the MRI images were recognized in this patient. T1-weighted images showed intermediate signal intensity, T2-weighted images showed increased signal intensity, and Gd-enhanced T1-weighted images revealed a notable increment in the signal intensity of the tumor. These findings are characteristic in both hemangiomas and angiosarcomas. It is not possible to distinguish between benign and malignant, or primary and secondary tumors by an MRI examination alone, but a careful analysis of the MRI images and a metastatic work-up could contribute by minimizing the possible number of differential diagnoses.

In most of the previous reports, the pathological diagnosis of a cardiac angiosarcoma intra vitam had been achieved by resecting the specimen or by a biopsy with thoracotomy (1). It is difficult to distinguish primary cardiac tumors and metastatic cardiac tumors without surgical procedures. In cases of metastatic cardiac tumor or cardiac involvement of malignant lymphoma, surgical resection should not be performed. A
transvenous endomyocardial biopsy is less invasive than an open biopsy or surgical resection and is a useful and safe diagnostic procedure under TEE guidance (3–6). However, some reports have stated that the specimens obtained from a transvenous biopsy show only false-negative findings of a cardiac angiosarcoma. Nitta et al reported a patient undiagnosed after a transvenous biopsy of a cardiac angiosarcoma under TEE guidance (7). Their autopsy findings showed that the tumor originated from the outer portion of the right atrium or the pericardium, and no tumor cells were detected in the endocardium. While we performed an endomyocardial biopsy under TEE guidance in only one patient, we made sure that this method was effective and safe enough to yield adequate tissue from the tumor segments. Because there is no evidence that a tumor resection improves the survival time in patients with a cardiac angiosarcoma, an endomyocardial biopsy might be an important diagnostic procedure to avoid an unnecessary operation despite such limitations. In the present case, after much discussion on the indication of tumor resection, we finally decided to resect the tumor for the following reasons: 1) no patients have reportedly survived without a tumor resection but some patients have survived after a tumor resection with supplemental treatment such as radiation therapy and chemotherapy, 2) the tumor seemed to be localized in RA free wall and it was thought to be totally resected, and 3) the recurrence of cardiac tamponade due to the hemorrhage from the tumor would be fatal to the patient. On the other hand, the immunological suppression, caused by a total cardiopulmonary bypass, might promote the dissemination of the angiosarcoma.

In summary, we present a case of a primary angiosarcoma of the heart, which demonstrated the advantages of MRI and TEE over TTE and CT for tumor detection.

References