IHJ Cardiovascular Case Reports (CVCR) xxx (2016) xxx–xxx

Contents lists available at ScienceDirect

IHJ Cardiovascular Case Reports (CVCR)

journal homepage: www.elsevier.com/locate/ihjccr

Case Report

Right atrial angiosarcoma presenting as giant pseudoaneurysm with impending rupture

J.S.N. Murthy[a,*], Rajani Gorantla[b], T. Periyasamy[c], Arunan Murali[b], Mahesh Vakamudi[d], B. Ranjith Karthekeyan[d], Rajendiran Swaminathan[e], Leena Dennis Joseph[e]

[a] Department of Cardiology, Sri Ramachandra University, Porur, Chennai 600116, Tamil Nadu, India
[b] Department of Radiology, Sri Ramachandra University, Porur, Chennai 600116, Tamil Nadu, India
[c] Department of Cardio Thoracic & Vascular Surgery, Sri Ramachandra University, Porur, Chennai 600116, Tamil Nadu, India
[d] Department of Anesthesiology, Sri Ramachandra University, Porur, Chennai 600116, Tamil Nadu, India
[e] Department of Pathology, Sri Ramachandra University, Porur, Chennai 600116, Tamil Nadu, India

ARTICLE INFO

Article history:
Available online xxx

Keywords:
Cardiac angiosarcoma
Cardiac pseudoaneurysm
Cardiac tumors

ABSTRACT

A 35-year-old female, who is a known case of right atrial angiosarcoma with giant pseudoaneurysm had been referred to us. CT angiogram was done and it showed signs of impending rupture. The diagnosis was confirmed by surgery and histopathology. Very few cases of cardiac rupture were reported in literature.

1. Introduction

Cardiac angiosarcoma is a rare aggressive primary malignant tumor of the heart and often difficult to diagnose clinically due to nonspecific signs and symptoms. Echocardiography is the first line of imaging; however, the advent of cross-sectional imaging with CT & MRI allows earlier detection of tumor and its complications along with distant metastasis.

2. Case report

A 35-year-old female was evaluated for dyspnea and chest pain, 5 months back with chest X-ray, contrast-enhanced CT (CECT) thorax, and cardiac MRI. CT thorax showed thin-walled partially thrombosed right atrial aneurysm (Fig. 1a) with multiple nodules in the lung. Cardiac MRI also showed a thin-walled aneurysm sac from the right atrium (Fig. 1b). There was no soft tissue infiltration of the right atrium and no pericardial effusion. The diagnosis of giant right atrial aneurysm was made. Her workup for connective tissue disorders came negative. She was started on medication and advised for regular follow-up.

She came to our institute with progressive worsening of dyspnea and increased chest pain. In our hospital, routine blood investigations, ECG, chest X-ray and 2D echocardiogram were done. Blood investigations revealed anemia. ECG showed normal sinus rhythm. Chest X-ray showed huge cardiomegaly with mediastinal widening (Fig. 2). Echo correlation revealed large collection communicating with anterolateral margin of right atrium with to and from color flow. Minimal pericardial effusion was also seen. However there was no cardiac tamponade. No echogenic mass was noted with in the cardiac chambers.

In view of continuous breathing difficulty and pericardial effusion, CECT chest was done, which showed a large aneurysmal sac near the right atrial appendage medially and extending up to the lateral chest wall (Fig. 3a and b). Luminal thrombus was seen on the lateral aspect of the aneurysm sac. Focal areas of wall irregularity and defects were observed on the lateral aspect of the sac, a diagnostic sign of aneurysmal rupture (Fig. 3a). There was also extraneous compression upon the superior vena cava with thrombosis at veno-atrial confluence (Fig. 3c). Large pericardial hematoma (Fig. 3d) was seen. The rest of cardiac chambers appear normal. In the lung window multiple hypodense nodules of various sizes (Fig. 4) are seen in both lung fields.

On comparison with previous CT & MRI (Fig. 1a and b) done 5 months back, there was significant increase in size of aneurysmal sac with features of impending rupture and also increase in the size and number of lung nodules. The possibility of the underlying primary malignant cardiac tumor causing spontaneous right atrial rupture and pseudoaneurysm formation with lung metastasis

* Corresponding author.
E-mail addresses: drjsnm@gmail.com (J.S.N. Murthy), drraaji@gmail.com (R. Gorantla), drsamy79@yahoo.com (T. Periyasamy), dr.arunan@gmail.com (A. Murali), vakamudi@gmail.com (M. Vakamudi), ranjithb73@gmail.com (B. Ranjith Karthekeyan), srajendiran@sriramachandra.edu.in (R. Swaminathan), leenadj@gmail.com (L.D. Joseph).

http://dx.doi.org/10.1016/j.ihjccr.2016.09.002

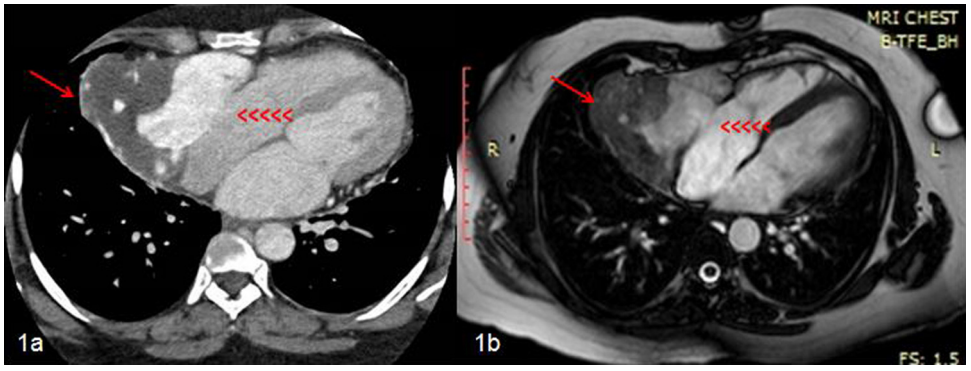

Fig. 1. Selective axial sections from CECT (a) and four-chamber view of cardiac MRI (b) showing defect in the right atrium (arrow heads) and partially thrombosed aneurysm (long arrows).

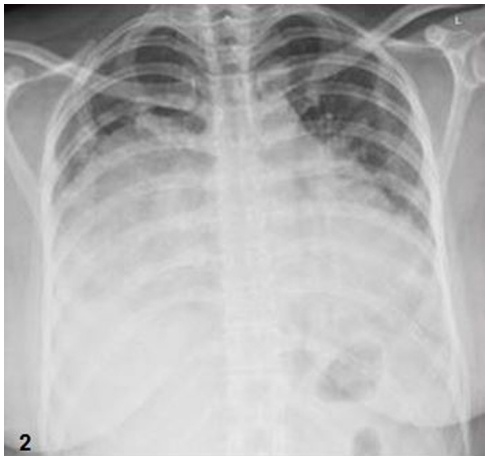

Fig. 2. Chest X-ray PA view demonstrating huge cardiomegaly and mediastinal widening.

was suspected. Based on CT findings of impending aneurysmal rupture she was taken up for surgical intervention with high-risk consent.

Surgery was performed under cardiopulmonary bypass with femoro femoral cannulation. Right atrium was found very friable and fenestrated. Blood in the aneurysmal sac was drained and large thrombus was evacuated (Fig. 5a). Biopsy was taken from the lung nodule (Fig. 5b). Right atrium was reconstructed with bovine pericardium and hemostasis secured. Patient was weaned off from cardiopulmonary bypass.

Decannulation was done, but the patient continued to have diffuse mediastinal ooze with deranged coagulation parameters. She received multiple blood transfusions and was shifted to intensive care unit with chest open and ventilator support. On the next day, she went into low cardiac output syndrome and could not be revived.

Histopathological examination showed a tumor with endothelial cell pleomorphism and mitosis. The pleomorphic tumor cells were seen surrounding vascular lamina, suggesting a high-grade malignancy of vascular origin (Fig. 6a). This was confirmed by

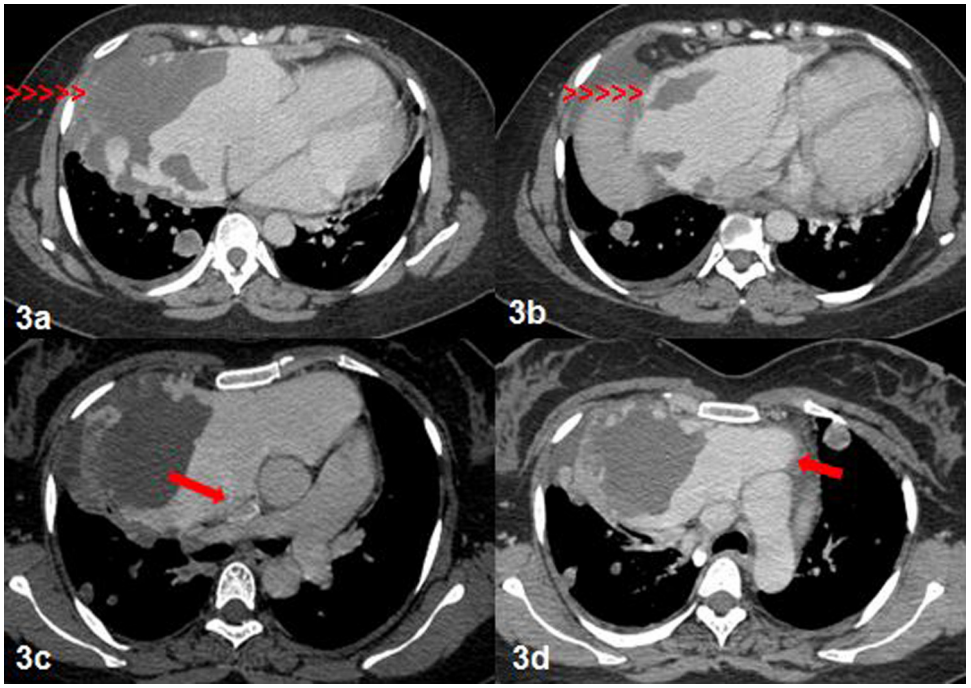

Fig. 3. Axial sections of the CECT thorax showing extent of aneurysmal sac up to the lateral chest wall with partial thrombosis (a and b). Contour of sac demonstrating focal defects and irregularities (arrow heads in a and b). Distal SVC thrombosis at veno atrial confluence (arrow in c). Large pericardial hematoma was also seen (arrow in d).

Please cite this article in press as: Murthy JSN, et al, Right atrial angiosarcoma presenting as giant pseudoaneurysm with impending rupture, *IHJ Cardiovasc Case Rep*. (2016), http://dx.doi.org/10.1016/j.ihjccr.2016.09.002