

## Case Report

# Rare pleomorphic undifferentiated sarcoma in right atrium with lung and pericardial metastasis in a 36 year-old woman: a case report

Bo Liu, Jing Xu, Jun-Kui Wang, Shang-Jian Li, Yong Zhang, Shuo Pan

First Department of Cardiology, People's Hospital of Shaanxi Province, No. 256 of West Youyi Road, Xi'an 710068, Shaanxi, People's Republic of China

Received December 20, 2015; Accepted March 5, 2016; Epub March 15, 2016; Published March 30, 2016

**Abstract:** The primary cardiac tumors were rare and the pleomorphic undifferentiated sarcomas were only accounted for a small amount of them with an incidence of 0.0004% to 0.005%. The pleomorphic undifferentiated sarcoma in right atrium with lung and pericardial metastasis was not reported before. A 36-year-old Chinese Han woman suffered 1 weeks of dyspnea on exertion, chest tightness. Transthoracic echocardiogram, cardiac nuclear magnetic resonance and positron emission tomography/computed tomogram scan and pathological biopsy were preformed were performed to make the diagnosis. Transthoracic echocardiogram showed large pericardial effusion, after the drainage of 500 ml pericardial fluid, 26 mm × 17 mm irregular mass revealed in the right atrium. Cardiac nuclear magnetic resonance and positron emission tomography/computed tomogram scan suggested multiple lung and pericardial metastasis. The final pathological diagnosis of mass in right lower lung was confirmed to be undifferentiated pleomorphic sarcoma. The patient denied further treatment and died 47 days after the first visit in our department. The pleomorphic undifferentiated sarcoma of right atrium may lead to metastasis of pericardium and lung and can be extremely lethal if left untreated.

**Keywords:** Pleomorphic undifferentiated sarcoma, right atrium, pericardium, lung

## Background

Primary cardiac tumors are rare with an incidence of 0.0017% to 0.019%, 25% of these tumors are malignant, the pleomorphic undifferentiated sarcomas are very rare [1]. It is known to primarily occupy the left atrium [2-4]. The metastasis of primary cardiac undifferentiated pleomorphic sarcoma was also rare, only a few cases reported the metastasis to maxilla [5], cerebra [6], skin [7] and spinal column [8]. In this case, we reported the natural progression of pleomorphic undifferentiated sarcoma in right atrium with lung and pericardial metastasis in a 36 year-old woman.

## Case report

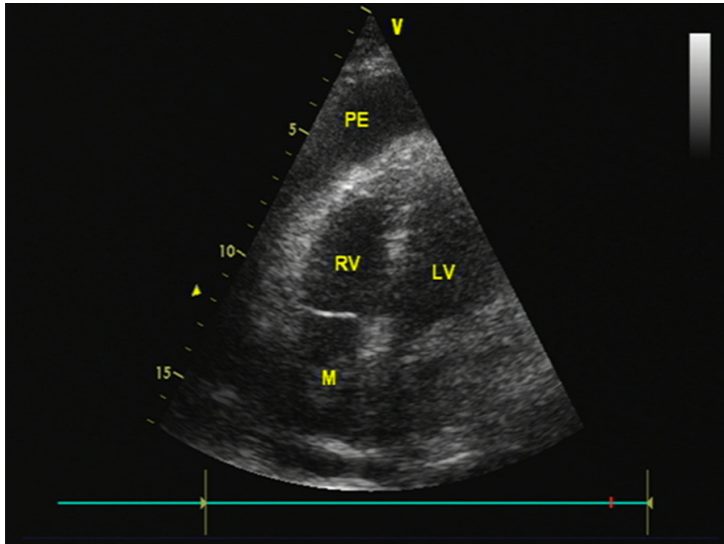
A 36-year-old Chinese Han woman with no past medical history presented to Cardiology Department of our hospital with 1 weeks of gradual onset of dyspnea on exertion, chest tightness. She denied any symptom of fever,

cough, expectoration, chest pain, abdominal pain and unintentional weight loss. She was not taking any medications or using tobacco, alcohol, or illicit drugs.

On physical examination, she had a temperature of 36.5°C, a heart rate of 112 bpm, respiratory rate of 24 breaths per minute, blood pressure of 90/60 mmHg, and oxygen saturation of 90% with 2-3 L/m oxygen by oxygen catheter. Her physical examination was notable for jugular venous distension, bilateral enlargement of the heart, and mild bilateral lower-extremity edema. Pathological murmurs were not heard on each auscultation area of heart.

Her electrocardiograph (ECG) showed sinus tachycardia with significant ST-T abnormalities on lead II, III, aVF and V1-V3. A transthoracic echocardiogram showed large pericardial effusion, intracardiac structures can't be presented clearly till pericardial effusion was removed. A chest radiograph showed enlargement of the

## Rare sarcoma in right atrium with metastasis



**Figure 1.** Transthoracic echocardiogram showing a 26 mm × 17 mm irregular mass in the right atrium.



**Figure 2.** Cardiac nuclear magnetic resonance scan showing abnormal signal intensity in inner side of superior vena cava, right side of ascending aorta and anterior and front space of right atrium. The mass was about 6.6 cm × 5.7 cm × 5.9 cm and was closely connected to anterior wall of right atrium and atrial septal and the superior vena cava was compressed and part of the auliflower-like mass protruded into right atrium.

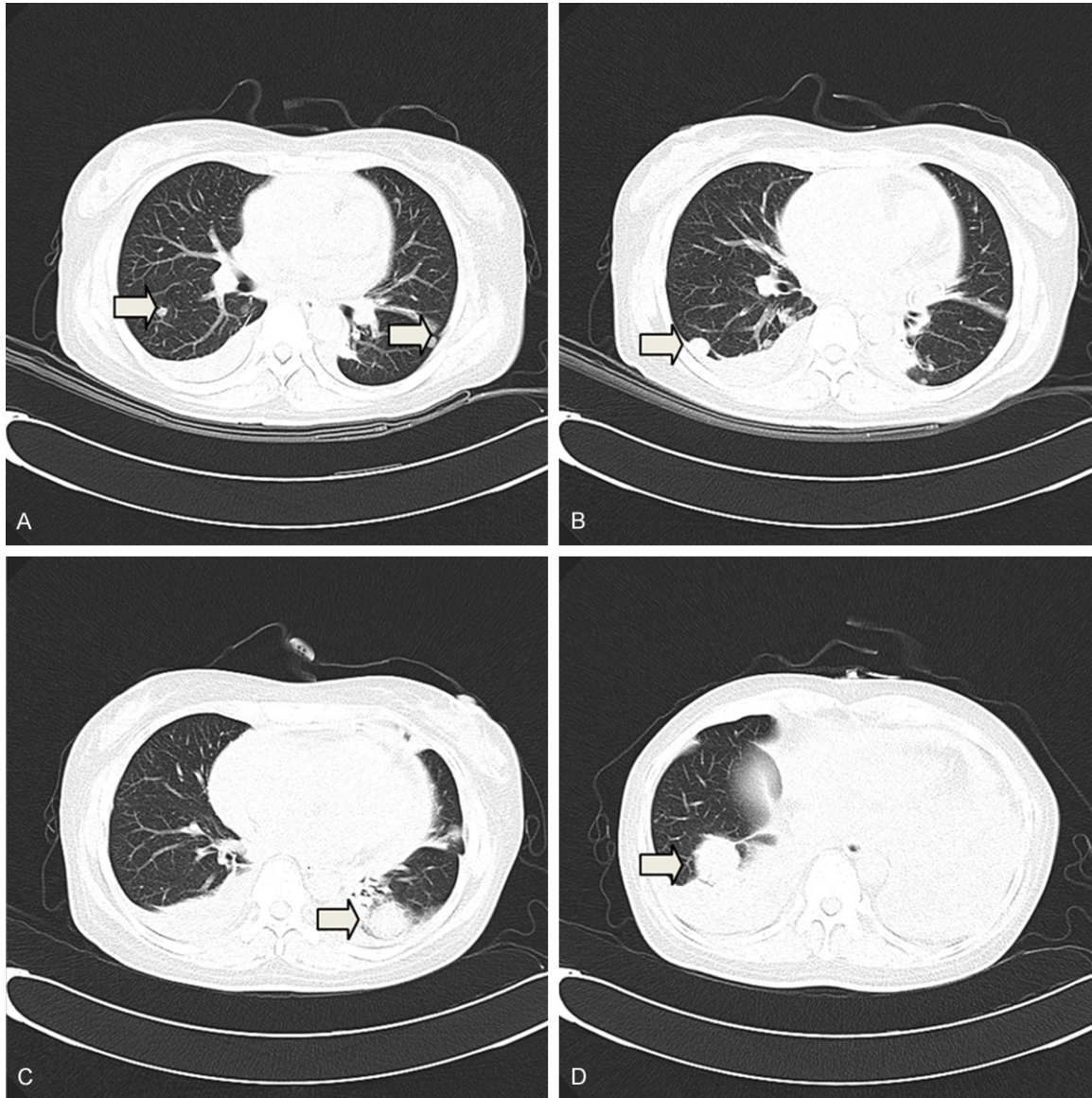
cardiac silhouette, bilateral air space opacities, and small bilateral pleural effusions.

back segment nodular mass was seen. The lesion tissues were wedge removed. Pathology

A pericardial puncture was performed and 500 ml of hemorrhagic fluid was extracted. Repeated transthoracic echocardiogram showed normal left and right ventricular systolic function. A 2.6 cm × 1.7 cm irregular mass in the right atrium was seen and no abnormal heart reflux was seen (**Figure 1**). Cardiac nuclear magnetic resonance imaging (NMRI) suggested that abnormal signal intensity was in inner side of superior vena cava, right side of ascending aorta and anterior and front space of right atrium. The size of the mass was about 6.6 cm × 5.7 cm × 5.9 cm and was closely connected to anterior wall of right atrium and atrial septal. The superior vena cava was severely compressed (**Figure 2**). Chest computed tomogram (CT) revealed multiple nodules in both lungs (**Figures 3, 4**). The following positron emission tomography/computed tomogram (PET/CT) showed intrapericardial mass with multiple lung metastasis and pericardial metastasis, no other metastatic lesion was seen throughout the body. Serum levels of the tumor markers carcinoembryonic antigen-125 (255.30 U/mL) and neuron-specific enzyme (26.90 U/mL) were elevated.

Since the increase of pericardial effusion was very fast with draining volume of 400-600 ml hemorrhagic fluid daily, the cardiac function and general status showed progressive deterioration, the weight loss was obvious. 14 days after the admission, the patient received the thoracoscopic wedge resection of right lower lung under general anesthesia. In the surgery, 3 cm × 3 cm basal segment mass of right lung lower lobe and 1.5 cm × 1.0 cm

## Rare sarcoma in right atrium with metastasis



**Figure 3.** Chest computed tomogram scan of pulmonary window showing multiple nodules (the arrow) in both lungs (From A to D: from upper level to the lower level).

showed that tumor cells were spindle-shaped or oval, diffusely arranged, homogeneous and dark stained under microscope, mitosis and pathological mitosis of the nuclear can be seen indicating that the tumor cells had the low degree of differentiation and a high degree of malignancy (**Figure 5**). Immunohistochemistry showed that Vim (+), CK (-), CK7 (-), SMA (-), des (-), CD34 (-), NapsinA (-), P63 (-), P40 (-). The final pathological diagnosis was undifferentiated pleomorphic sarcoma.

The patient and family members denied the further heart surgery and chemotherapy we had suggested, then the patient was discharged.

Two weeks later, the patient presented progressive paroxysmal nocturnal dyspnea, generalized edema, eating difficulties and was sent to Oncology Department. The chest radiograph showed increased number of nodular masses in both lungs (**Figure 6**). The transthoracic echocardiogram showed 48 mm × 47 mm mass in the right atrium (**Figure 7**). The patient died 47 days after the first visit in the Cardiology Department.

### Comment

In this case, the cardiac sarcoma was located in the right atrium while the majority of the pleo-

## Rare sarcoma in right atrium with metastasis



**Figure 4.** Chest computed tomogram scan of mediastinal window showing multiple nodules (the arrow) in both lungs (From A to D: from upper level to the lower level).

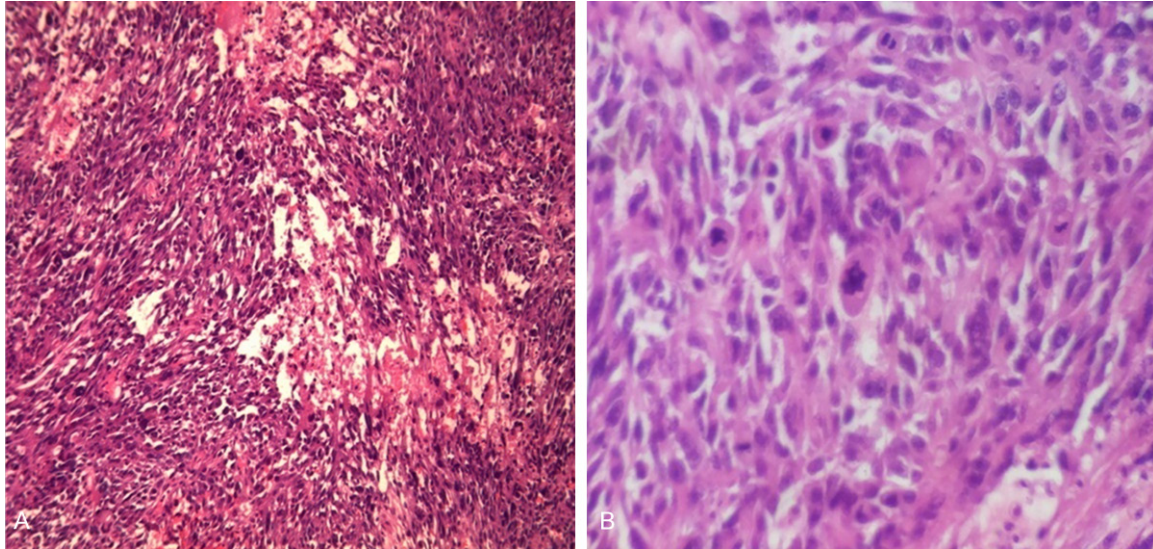
morphic undifferentiated sarcomas were located in the left atrium. Meanwhile, the metastasis of pleomorphic undifferentiated sarcoma was very rare, the metastasis to pericardium and lung were not documented elsewhere.

Some case reports had suggested the resection of sarcomas [3, 4, 9], however, the mean survival time after excision is still very short because of high potential for recurrence and metastasis [1, 10]. In this case, pericardial metastasis was observed and pericardial effusion increased dramatically, the patient endured 12.5 kg of weight loss from first visit till death. The benefit of the surgery may be limited

and the benefit of adjuvant chemotherapy and radiotherapy is still unknown for the lack of randomized trials [11]. After communication with the patient and relatives about the prognosis, the patient denied further heart surgery and chemotherapy.

The natural progression of the diseases was fast, only less than 20% of patients were alive 1 year after diagnosis despite the treatment [12]. In this case, because of increased size of mass in the right atrium, the rapid growth of pericardial effusion and increased number of nodular masses in both lungs, the general condition of the patient deteriorated dramatically, the

## Rare sarcoma in right atrium with metastasis



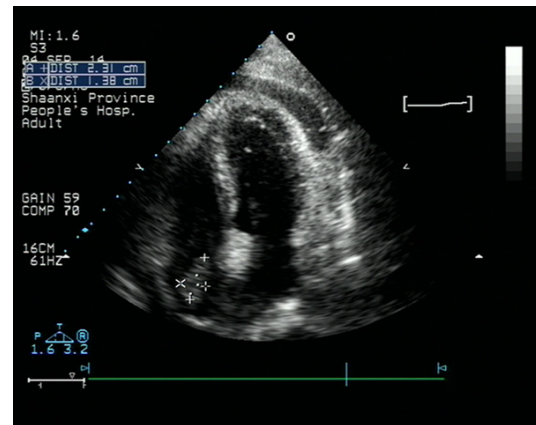
**Figure 5.** Pathology of right lung lower lobe mass showing spindle-shaped or oval, diffusely arranged, homogeneous and dark stained cells under microscope (A), mitosis and pathological mitosis of the nucleus can be seen indicating that the tumor cells (B) had the low degree of differentiation and a high degree of malignancy.



**Figure 6.** Chest radiograph showing increased number of nodular masses in both lungs.

patient died 47 days after the first visit in our department.

This case is unique because it is the only reported case of pleomorphic undifferentiated sarcoma in right atrium with lung and pericardial metastasis, the natural progression of the diseases could be extremely fast if left untreated.



**Figure 7.** Transthoracic echocardiogram showing 48 mm × 47 mm mass in the right atrium.

### Acknowledgements

This paper was funded by National Natural Science Funds of China (Grant No. 81500308). Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images.

### Disclosure of conflict of interest

None.

### Authors' contribution

BL, SP obtained the radiographic images and drafted the manuscript. JX, J-KW, S-JL, YZ

## Rare sarcoma in right atrium with metastasis

helped to draft the manuscript. BL, SP conceived of the case report, participated in coordination. All authors read and approved the final manuscript.

**Address correspondence to:** Dr. Shuo Pan, First Department of Cardiology, People's Hospital of Shaanxi Province, Xi'an 710068, People's Republic of China. Tel: +86 29 85251331; E-mail: parker1985@sina.com

### References

- [1] Yang EH, Gabrielian V, Ji P, Omari B, Thomas JL, Chung J. Left atrial undifferentiated pleomorphic sarcoma causing mitral valve obstruction. *Circulation* 2011; 123: e11-14.
- [2] Shanmugam G. Primary cardiac sarcoma. *Eur J Cardiothorac Surg* 2006; 29: 925-932.
- [3] Bégué C, Barreda E, Hammoudi N, Fouret P, Toledano D, Isnard R, Leprince P, Montalescot G, Barthélémy O. Left atrial high-grade undifferentiated pleomorphic sarcoma protruding through the mitral valve. *Ann Thorac Surg* 2014; 98: 2227-2229.
- [4] Abe D, Sato A, Takeyasu N, Tokunaga C, Akishima S, Iijima T, Sakakibara Y, Aonuma K. Life-threatening acute heart failure due to primary cardiac undifferentiated pleomorphic sarcoma. *Intern Med* 2014; 53: 1775-1777.
- [5] Eisenberg E, Natarajan E, Yamase H, Hatzigiannis G. Undifferentiated cardiac sarcoma metastatic to the maxilla: report of a case. *J Oral Maxillofac Surg* 2007; 65: 148-151.
- [6] Regel JP, Pospiech J, Baume B, van de Nes JA. Cerebral metastasis from an undifferentiated sarcoma of the left atrium. *Acta Neurochir* 2006; 148: 595-596.
- [7] Val-Bernal JF, Figols J, Arce FP, Sanz-Ortiz J. Cardiac epithelioid angiosarcoma presenting as cutaneous metastases. *J Cutan Pathol* 2001; 28: 265-270.
- [8] McLoughlin GS, Sciubba DM, Ali SK, Weinkauff JG, Fourny DR. An unusual case of spinal column metastasis after orthotopic transplantation for cardiac sarcoma. *J Neurosurg Spine* 2008; 9: 377-381.
- [9] Nowacka A, Muller O, Hurni M, Prêtre R, Niclauss L. Obstructive pulmonary artery sarcoma. *Heart Lung Circ* 2015; 24: e63-64.
- [10] Agaimy A, Rösch J, Weyand M, Strecker T. Primary and metastatic cardiac sarcomas: a 12-year experience at a German heart center. *Int J Clin Exp Pathol* 2012; 5: 928-938.
- [11] Llombart-Cussac A, Pivot X, Contesso G, Rhor-Alvarado A, Delord JP, Spielmann M, Türsz T, Le Cesne A. Adjuvant chemotherapy for primary cardiac sarcomas: the IGR experience. *Br J Cancer* 1998; 78: 1624-1628.
- [12] Barreiro M, Renilla A, Jimenez JM, Martin M, Al Musa T, Garcia L, Barriales V. Primary cardiac tumors: 32 years of experience from a Spanish tertiary surgical center. *Cardiovasc Pathol* 2013; 22: 424-427.