

1           **Primary cardiac hemangioendothelioma of the right ventricle**

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17**Abstract**

18Primary cardiac hemangioendothelioma is a very rare low-grade malignancy. We

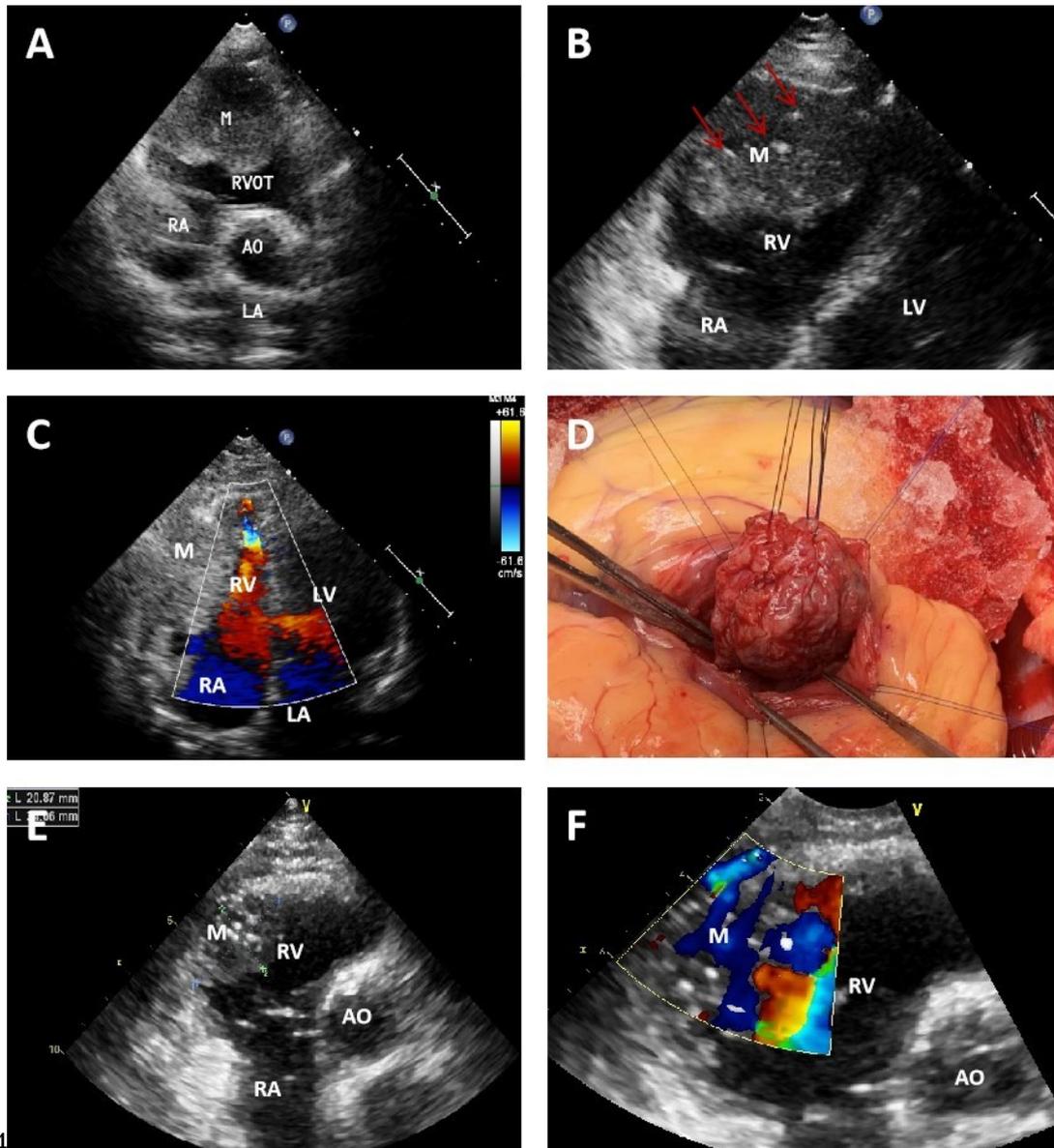
19present a case of a 41-year-old female patient with a huge primary cardiac

20hemangioendothelioma that arose from the right ventricle was recurred after being

21removed, and the diagnostic images given by echocardiography.

22**Keywords:** heart neoplasms, hemangioendothelioma, ventricle, right,

23echocardiography



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25Figure. (A) Ultrasound images showing a solid mass attached to the myocardium of  
 26the right ventricular free wall. (B) Point-like calcification within the mass (red arrow).  
 27(C) Absence of flow signals within the tumor. (D) Intra-operative photograph showing  
 28a soft red mass in the right ventricular wall. (E) The tumor recurred in the original  
 29location. (F) It has more blood flow signals than before the operation.  
 30M, mass; RV, right ventricle; RA, right atrium; RVOT, right ventricular outflow tract;  
 31AO, aorta; LA, left atrium; LV, left ventricle.

### **32Introduction**

33Primary cardiac tumors are rare, and the estimated frequency at autopsy is only  
340.001%-0.300% [1], of which only about 30% are malignant tumors. Primary cardiac  
35hemangioendothelioma is extremely rare. So far, fewer than 20 cases have been  
36reported worldwide [2]. Echocardiography is the most useful diagnostic technique for  
37cardiac tumors, and its diagnostic rate reached 94% with regarding non-necropsy  
38cardiac tumors [3]. Of course, the final diagnosis still depends on pathology.

### **39Case presentation**

40A 41-year-old woman presented to our hospital with atypical chest discomfort of 2  
41months duration. She was previously healthy and denied hypertension, coronary heart  
42disease, and other cardiovascular diseases. Physical examination showed mmHg of  
43120 / 80mmHg (1mmHg = 0.113kPa) blood pressure, 72 beats per minute of pulse,  
44and pre-cardiac area with II/VI grade contraction murmur. Electrocardiogram showed  
45sinus rhythm and normal electrocardiogram. Chest X-ray identified normal.  
46Electrocardiogram showed sinus rhythm and was normal. Echocardiography showed a  
47solid  $5.1 \times 3.5 \times 5.3$  cm mass with a wide base attached to the myocardium of the  
48right ventricular free wall (Fig. A). The boundary between the tumor and the  
49myocardium was indistinct. The mass oscillated slightly with the cardiac cycle.  
50Internal echogenicity was medium and heterogeneous, and point-like calcification was  
51noted (Fig. B). No blood flow signals were detected within the tumor, and the right  
52ventricular outflow tract was slightly obstructed, there was slight tricuspid valve

53regurgitation (Fig. C). We diagnosed a right ventricular tumor.

54 The mass was successfully removed. It was found to be located in the right  
55ventricular wall, consistent with the preoperative echocardiography findings. The  
56mass appeared red and soft, and the boundary between the tumor and myocardium  
57was indistinct (Fig. D). The histopathological examination of the tumor section  
58showed that it was an angiogenic tumor with dense proliferation of focal endothelial  
59cells, which was considered composite hemangioendothelioma. The patient's  
60echocardiography was reviewed three months after the operation, and the results  
61showed that the tumor recurred at the original location (Fig. E). It measured  $3.4 \times 2.0$   
62 $\times 1.6$  cm with more blood flow signals than before the operation (Fig. F).

63

#### 64**Discussion**

65Hemangioendothelioma is a vascular tumor that exhibits borderline biological  
66behavior, intermediate between completely benign hemangioma and high-grade  
67angiosarcoma [4]. Hemangioendothelioma originating in the heart is extremely  
68rare[2], which is generally regarded as a low-grade malignant tumor. It usually  
69originates from the subendocardium and may occur at any part of the heart.

70 Imaging examination can provide valuable information for the diagnosis of cardiac  
71hemangioendothelioma, of which echocardiography is the best diagnostic technique.  
72Echocardiography can provide information about the size, shape, location of the heart  
73tumor, its relationship with surrounding structures and its effect on cardiac

74hemangioendothelioma, and is helpful for preoperative diagnosis. We performed  
75echocardiography on the patient in this case, and imaging shows: 1. a solid mass, ill-  
76defined border, soft-texture, and with a wide base attached to the right ventricular  
77myocardium. 2. medium-echo, heterogeneous echopattern with calcification and 3.  
78No blood flow signal in the tumor.

79 However, since hemangioendothelioma is classified as an intermediate malignancy,  
80so that sometimes metastasizes and recurs[5]. The echocardiogram of this patient was  
81reviewed three months after the operation. The results showed that the tumor recurred  
82in the original position of the right ventricular wall and its blood supply was more  
83abundant than before the operation. This should be vigilant, this tumor has a stronger  
84biological behavior.

### 85**Conclusion**

86In this rare case, we have described a huge primary cardiac hemangioendothelioma  
87was recurred after being removed, and the diagnostic images given by  
88echocardiography, and to enrich the imaging information of cardiac  
89hemangioendothelioma.

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