

# Multimodality imaging of a rare right ventricular angioleiomyoma

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## Abstract

We report a rare case of right ventricular angioleiomyoma in a 35-years old man diagnosed by multimodality imaging, including echocardiography, cardiac magnetic resonance imaging (CMR), Positron emissions tomography (PET) and Computer tomography angiography (CTA). The imaging results were finally confirmed by surgery. We present the clinical value of multimodality imaging in diagnosing cardiac angioleiomyoma.

## Title Page

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【Abstract】

We report a rare case of right ventricular angioleiomyoma in a 35-years old man diagnosed by multimodality imaging, including echocardiography, cardiac magnetic resonance imaging (CMR), Positron emissions tomography (PET) and Computer tomography angiography (CTA). The imaging results were finally confirmed by surgery. We present the clinical value of multimodality imaging in diagnosing cardiac angioleiomyoma.

## 【Introduction】

Angioleiomyoma is a benign tumor with clear perimeter, composed of bundles of mature smooth muscle and thick-walled blood vessels<sup>1</sup>, described mainly occurs in the subcutaneous or deep dermis, rarely happens in the heart. Although very rare, it can cause serious consequences, even the most benign and smallest tumors can lead to significant morbidity and mortality<sup>2</sup>. Therefore, early diagnosis and treatment is very important. Multimodality imaging, including cardiac magnetic resonance imaging (CMR), Positron emissions tomography (PET) and Computer tomography angiography (CTA) can make the diagnosis more accurate. We report a rare case of right ventricular angioleiomyoma, presenting the significant role of multimodality imaging in diagnosing.

## 【Case Presentation】

A 35-year-old male suffered from emaciation with no obvious cause for more than five months. An unexpected right ventricle (RV) mass was revealed by transthoracic echocardiography (TEE) in a physical check-up of the local hospital, so he came to our hospital for the further treatment. His vital signs and physical examination showed no special.

Our TTE showed an oval medium-high echo RV mass, measured 1.4 cm\*1.1 cm, with a rough surface at the basal level of the interventricular septum (IVS), which swung slightly and seemed to be connected to the IVS by a pedicle about 0.3 cm wide, and the attachment point was about 1.6 cm from the tricuspid annulus (Fig 1A). Two-dimensional and three-dimensional transesophageal echocardiography (TEE) indicated that the attachment of the RV mass seemed to be closely connected with the tricuspid tendinous cord and adjacent to the aortic root (Fig1B~D). Cardiac magnetic resonance imaging (CMR) showed the soft tissue signal nodule presenting slightly T1 hypo-intensity and slightly T2 hyper-intensity which suggested high possibility of benign neoplasm, but the attachment point was unclear (Fig 1E). Positron emissions tomography (PET) showed that there was no obvious abnormal increase of metabolism in the RV (Fig 1F). Computer tomography angiography (CTA) also showed a filling defect measured about 1.0 cm in the RV (Fig 1G). The patient then underwent video-assisted thoracoscopic cardiac mass resection and tricuspid valvuloplasty. Thoracoscopy revealed the attachment of RV mass was approximately 1.5 cm from the anterior-septal junction of tricuspid valve and adhered to tendinous cord of tricuspid septal valve (Fig 1H). The resected tumor was a slightly hard spherical mass with a capsule and had a gray-white and red surface, measuring 1.2 cm\*1.3 cm\*1.2 cm (Fig 2A). Further histopathological examination confirmed a right ventricular angioleiomyoma with stromal mucinous change, the immunohistochemical stain showed h-caldesmon and smooth muscle actin positive (Fig 2 B~F).

## 【Discussion】

Angioleiomyoma is a benign tumor with clear perimeter, composed of mature smooth muscle cells with a prominent vascular component<sup>3</sup>. The tumor tends to occur most commonly within the 30–50 years age range and is approximately twice as prevalent in women than in men<sup>4</sup>. It appears most frequently on the extremities, especially the leg. Other localizations such as uterus, ear, lip, nose, neck, and larynx have been also reported. Heart localization is extremely rare. In fact, we found only one previous report of cardiac angioleiomyoma<sup>5</sup>. Histogenesis of cardiac angioleiomyoma remains unclear, it is reported that subendocardial multipotential mesenchymal cells, persistent tissue of the endocardial cushion, or valvular interstitial cells may be the origin<sup>6</sup>. Histologically angioleiomyomas were classified as solid, venous, and cavernous, of which the solid type being most frequently observed. Myxoid or hyaline change may occur due to flow disturbances within the tumor<sup>5</sup>. Although primary ventricular angioleiomyomas are exceedingly rare in human, but not uncommon in cattle. A study of cardiac angioleiomyoma in 44 cattle suggested that angioleiomyomas are usually in subendocardial tissue and swing slightly with the cardiac cycle but not within the myocardium<sup>6</sup>.

Also, the position of the tumors is highly characteristic: all involved the valve complex (valves, chordae tendineae, or papillary muscles). The overlying endocardium of the tumor is smooth, without thrombosis and the cut surface of tumors is white or red, or both. These findings are almost consistent with those of this case and another patient reported in the literature<sup>5</sup>.

In addition to the high specificity of tumor location, angioleiomyoma also has its imaging characteristics, and the advent of multimodality imaging has made the diagnosis more accurate. Angioleiomyoma is usually a well-circumscribed oval mass with a homogeneous echo texture on US<sup>7</sup>. In this study, cardiac angioleiomyoma can swing slightly with the cardiac cycle in TTE or TEE. Computer Tomography(CT) or contrast-enhanced CT often show a soft density tissue<sup>8</sup>. The tumor presents as a filling defect in CTA. In MRI, it has been described in previous studies as a well-circumscribed mass with an isointense or slightly hyperintense signal relative to the muscle on T1-weighted images and a heterogeneous and hyperintense signal on T2-weighted and STIR images<sup>7</sup>. In addition, as a benign tumor, angioleiomyoma usually do not cause increased metabolism in PET. The imaging techniques described above can provide anatomical and hemodynamic information of angioleiomyoma, which is conducive to surgical planning.

In conclusion, we have reported a right ventricular angioleiomyoma, a rare type of heart tumor. It should be considered in the differential diagnosis of ventricular tumors.

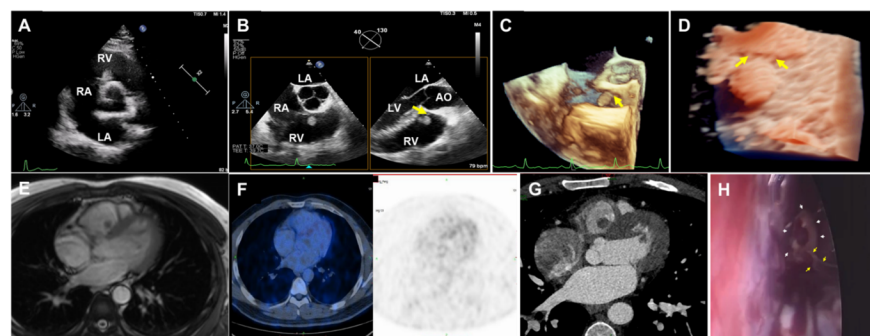


Fig 1. (A) Transthoracic echocardiogram showed a RV mass at the basal level of the interventricular septum; (B~D) Transesophageal echocardiography showed the attachment closely connected with the tricuspid tendinous cord (yellow arrows) and adjacent to the aortic root; (E) Cardiac magnetic resonance imaging showed a soft tissue signal nodule; (F) PET showed no obvious abnormal increase of metabolism in the right ventricle; (G) CTA showed a RV filling defect; (H) Thoracoscopy revealed the mass (the white arrows indicated its outline) and its attachment adhered to tendinous cord (little yellow arrows) of tricuspid septal valve.

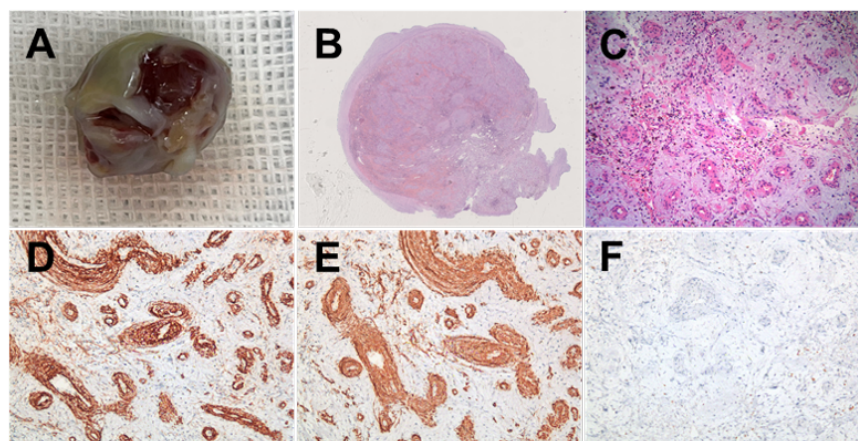


Fig 2. (A) Resected mass presented a spherical shape with a capsule and had a gray-white and red surface; (B and C) HE stain showed a well-circumscribed nodular tumor composed of small muscular blood vessels with mucoid degeneration in it; (D and E) Smooth muscle cells showed immunoreactivity for h-caldesmon, smooth-muscle-actin; (F) Immunohistochemical stain showed HMB45 negative.

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