

Suspected adult cellular rhabdomyoma in a 24-year old woman: diagnostic and therapeutic challenges

Clinical Case Portal

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Case Report

The patient presented with dyspnea (NYHA II). Physical examination was unremarkable. Transthoracic echocardiography revealed an oval shaped solid tumor, measuring 50x27 mm, originating from the distal two-thirds of the interventricular septum. Transesophageal echocardiography and computerized tomography showed similar findings. No endomyocardial biopsy was performed because it was felt that this would be too risky for the patient. It was concluded that the mass is probably an adult cellular rhabdomyoma.

Ten months from the initial examination the patient is still being followed without medical therapy. The mass did not increase in size and the patient did not report any symptomatic deterioration. It is not clear whether this patient should be followed with no intervention or operated.

Patient history prior to current observation :24-year-old woman was admitted to our hospital for the detailed delineation of left ventricular mass that was noted on an outpatient echocardiogram. History revealed that for the last 3-4 years the patient was complaining about dyspnea on heavy-to moderate exertion (NYHA II). She had two near-syncope, but she never fainted. Other details of personal and family history were unremarkable.

Clinical findings on admission, evolution and outcome :

Physical examination was unremarkable. Blood pressure measured 90/60 mmHg. Her chest X-ray was also unremarkable. EKG showed normal sinus rhythm of 65 beats/min, normal axis, and negative T wave in leads D2, D3, aVF. Routine laboratory tests were within normal range.

Transthoracic echocardiography revealed an oval shaped solid tumor, measuring 50x27 mm, originating from the distal two-thirds of the interventricular septum protruding into the left ventricle and to a lesser extent into the right ventricle (fig. 1, fig. 2). **The tumor was contracting simultaneously** with the rest of the left ventricle, and was well demarcated. EF was visually estimated at 60%. Other findings were unremarkable. Transesophageal echocardiography showed similar findings. **CT scan** of the heart and mediastinum showed left ventricular mass originating from the interventricular septum with no signs of extracardiac extension or other mediastinal masses fig. 3.

24h Holter monitoring revealed normal sinus rhythm with frequent episodes of sinus arrhythmia during sleep. There was a total of 6 ventricular extrasystoles in two morphological forms, and one episode of idioventricular rhythm. Electrophysiology testing demonstrated normal function of SA and AV nodes, and ventricular stimulation did not produce ventricular tachycardia or fibrillation.

Metabolic testing using Bruce treadmill protocol was stopped at the stage 3 (7.8 METS) due to fatigue. Peak oxygen consumption was 27.54 ml/kg/min, which was 52% of her maximal predicted value. No complex ventricular arrhythmias were noted on exertion.

No endomyocardial biopsy was performed because it was felt that this would be too risky for the patient. It was concluded, based on echocardiographic characteristics, that the left ventricular mass is probably tumor originating from cardiac striated muscle cells, possibly an adult cellular rhabdomyoma.

Ten months from the initial examination the patient is still being followed. The mass did not increase in size and the patient did not report any symptoms except dyspnea on exertion (NYHA II). However, it is still not clear whether this patient should be followed or referred to surgery.

Discussion

Diagnostic challenges. Echocardiographic features of intracardiac mass in this patient suggested that the mass is consistent with primary cardiac tumor. Lack of any sign of systemic deterioration over the period of 3-4 years, when the first symptoms were noted, further indicated that the mass is probably a benign tumor. Site of origin and fact that the mass was contracting simultaneously with the rest of the left ventricle suggested that the mass most probably originated from the cardiac striated muscle cells.

As recently suggested by Burke and colleagues, (1) tumors originating from cardiac striated muscle can be either rhabdomyoma, hamartoma, adult cellular rhabdomyoma or rhabdomyosarcoma. Only biopsy can provide a definite diagnosis, but clinical and morphological features of the intracardiac mass indicated that the mass may be an adult cellular rhabdomyoma, since it was diagnosed in adulthood and was well demarcated. Adult cellular rhabdomyomas are true neoplasms and have probably benign biologic behaviour due to low-to-moderate propensity toward cell proliferation. Adult rhabdomyomas usually occur in the head and neck region, (2) and only four cases of intracardiac location have been described so far. (1, 3) Diagnosis of a rhabdomyoma is unlikely as these tumors rarely occur after the age of 10 and are frequently associated with tuberous sclerosis. (4) Similarly, diagnosis of a hamartoma can also be ruled out with reasonable certainty as hamartomas are poorly demarcated.

Although the exact pathohistologic diagnosis remains highly speculative, echocardiography helped us to considerably narrow differential diagnosis and plan further therapeutic options.

Therapeutic challenges. Even if they are histologically benign, intracardiac masses should be considered to have malignant behavior by virtue of their location. Therefore surgical removal of the tumor in our patient should be regarded as the most logical therapeutic option. Although successful surgical removal of the intracardiac rhabdomyomas has been described (1, 5) these tumors were not nearly as large as the tumor in our patient. Surgical removal of the tumor would lead to severe mutilation of the septum that would probably require implantation of the patch. If this would be the case, left ventricular systolic function would be seriously impaired, and there would be no improvement in patient's functional status. Moreover, it is not clear what is the mortality and morbidity rate for the operation of large intracardiac rhabdomyomas. And last, but not least, surgeons in our institution do not have experience with similar operations. But then, if the tumor continues to grow it would be even more difficult to do surgery in the future.

Conclusion

Since the patient is oligosymptomatic, another line of therapeutic reasoning is to wait whether the tumor will further continue to grow and/or whether the patient will develop more severe symptoms. Since functional impairment is probably due to left ventricular diastolic dysfunction caused by the tumor and electrophysiologic testing was unremarkable, it is not clear whether the patient would benefit for any medical treatment.

For the time being we decided to follow the patient without any medical therapy. She is being followed for ten months now, did not experience any symptomatic deterioration and the tumor remained exactly the same size. Although this decision may be argued, we opted for this modality because of the uncertainties that surgeons in our institution expressed about the surgical removal of the tumor.

References

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Fig. 1 :

[Suspected adult cellular rhabdomyoma Transthoracic short axis view](#)



Fig. 2 :

[Suspected adult cellular rhabdomyoma Transthoracic subcostal four-chamber view](#)



Fig. 3 :

[Suspected adult cellular rhabdomyoma CT scan](#)

