Primary cardiac hemangioendothelioma of the mitral valve

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ABSTRACT

Primary cardiac hemangioendothelioma is a very rare tumor. Up to now, <20 cases have been reported worldwide; involvement of mitral valve by the tumor is extremely rare. In this report, a case of hemangioendothelioma arose from the mitral valve and was successfully resected, and after few months the tumor recurred and infiltrated the heart.

1. Introduction

Primary cardiac hemangioendothelioma is a very rare tumor. Up to now, <20 cases have been reported worldwide; involvement of mitral valve by the tumor is extremely rare. In this report, a case of hemangioendothelioma arose from the mitral valve and was successfully resected, and after few months the tumor recurred and infiltrated the heart.

2. Case report

A 35-year-old man presented by progressive shortness of breath for about 6 months. He previously had been healthy, not diabetic nor hypertensive. On physical examination, patient looks ill, restless and dyspneic, cardiac examination revealed pansystolic murmur over the apex with accentuated second heart sound. Transesophageal echocardiography displayed that anterior mitral valve leaflet was infiltrated by a large tumor which extended to the interatrial septum about 5 x 3 cm, with moderate to severe mitral stenosis and severe mitral regurgitation, dilated right ventricle and right atrium with severe pulmonary hypertension (Fig. 1).

Transthoracic echocardiography displayed that anterior mitral valve leaflet was infiltrated by a large tumor which extended to the interatrial septum about 5 x 3 cm, with moderate to severe mitral stenosis and severe mitral regurgitation, dilated right ventricle and right atrium with severe pulmonary hypertension (Fig. 1).

The patient was sent for cardiac magnetic resonance imaging, but he developed panic attack and refused to continue examination. At the operation, the cardiac tumor was infiltrating the mitral valve, fibrous trigone, and interatrial septum, the tumor mass was resected with the mitral valve, interatrial septum with safety margin with repair of the interatrial septum by autopericardium and mitral valve metallic prosthesis was inserted, and postoperative echocardiography revealed well-functioning prosthetic valve and intact interatrial septum (Fig. 3).

Pathological examination revealed that the tumor was composed of short strands, cords of round to slightly spindle-shaped cells with round nuclei, and prominent cytoplasmic vacuolization consistent with hemangioendothelioma (Fig. 4).

The postoperative course was uneventful with clinical improvement. The patient was discharged from the hospital after 1 week. The patient was asymptomatic and recovered well (Fig. 5). After 9 months, the symptoms recurred again with severe dyspnea and orthopnea, transesophageal echocardiography revealed recurrence of the tumor which infiltrated the prosthetic mitral valve and aortic valve and extended to the interatrial septum (Fig. 6). The patient condition deteriorated rapidly. The decision was to redo the operation, but the patient refused to undergo another surgery, and he died after 1 month from heart failure.

3. Discussion

Cardiac neoplasms may be primary or secondary. Metastatic tumors are about 30 times more common than the primary ones; most primary tumors are benign, and only about 30% of them are malignant. Among the primary cardiac tumors, myxoma is the most common neoplasm. Hemangioendothelioma is an uncommon vascular tumor which often presents in soft tissues and is known to occasionally metastasize.
arising from the heart is very rare; to our knowledge, <20 cases have been previously reported in the literature with only two cases of involvement of mitral valve. Cardiac hemangioendothelioma is classified as a low-grade malignant neoplasm. Up to now, the pathogenesis of this rare disorder is unknown, but this tumor usually originates from the subendocardium and may occur at any part of the heart. In hemangioendothelioma, vascular differentiation proceeds through the formation of multicellular, canalized vascular channels. Vascular differentiation in these tumors is more primitive and is expressed primarily at the cellular level. It has been reported that most of patients with cardiac hemangioendothelioma after resection have a favourable outcome, whereas radiation therapy and chemotherapy have only limited effect on the tumor. In the present case, the tumor was infiltrating the mitral valve and extending to the interatrial septum; it was successfully resected with no other additional therapy. The patient was discharged in a good condition, but after a few months, the symptoms of heart failure recurred again and echocardiography revealed recurrence of the tumor with aggressive infiltration of the prosthetic mitral valve, aortic valve, and interatrial septum. The patient condition deteriorated rapidly and the patient refused to undergo another surgery and the patient died after few days.
Fig. 3. Transesophageal echocardiography showing the tumor.

Fig. 4. Pathological examination with short strands, cords of round to slightly spindle-shaped cells.

Fig. 5. Postoperative echocardiography showing well-functioning valve.
4. Conclusion

Surgical resection alone may be not sufficient in management of hemangioendothelioma; the role of adjuvant chemotherapy and radiotherapy should be addressed, although the rarity of these cases may make that difficult.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Appendix A. Supplementary material

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ehj.2018.06.002.

References

1. Bonow RO, Mann DL, Zipes DP, Libby P. Braunwald’s heart disease E-book: a textbook of cardiovascular medicine. Elsevier Health Sciences; 2011.
2. Weiss SW, Ishak KG, Dail DH, Sweet DE, Enzinger FM. Epithelioid hemangioendothelioma and related lesions. Semin Diagn Pathol. 1986;3(4):259–287.
3. Sugimoto T, Yamamoto K, Yoshii S. A primary epithelioid hemangioendothelioma of the right atrium: report of a case and literature review. Open J Thorac Surg. 2013;3:63.
4. Enzinger F, Weiss SW. Benign tumors and tumorlike lesions of blood vessels. Soft Tissue Tumors. 1988;489–532.
5. Wang LF, Liu M, Zhu H, et al.. Primary cardiac hemangioendothelioma: a case report. Chin Med J-Beijing-English Ed. 2006;119(11):966.