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**ARTERIOVENSKI HEMANGIOM DESNE KOMORE: PRIKAZ SLUČAJA I PREGLED LITERATURE**


**UDC:**

**DOI:** [https://doi.org/10.2298/VSP171108025M](https://doi.org/10.2298/VSP171108025M)

When the final article is assigned to volumes/issues of the Journal, the Article in Press version will be removed and the final version appear in the associated published volumes/issues of the Journal. The date the article was made available online first will be carried over.
THE ARTERIOVENOUS HEMANGIOMA OF THE RIGHT VENTRICLE: CASE REPORT AND LITERATURE REVIEW

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Abstrakt

Uvod: Hemangiomi desne komore srca su izuzetno retki i uglavnom asimptomatski benigni tumori. Hirurško lečenje je terapija izbora.


Zaključak: Konstatovano je da se ova procedura može bezbedno koristiti kao terapija izbora sa odličnim dugoročnim rezultatima.

Klučne reči: hemangioma srca, desna komora, hirurško lečenje

Abstract

Introduction: Cardiac hemangiomas of the right ventricle are very rare and mostly asymptomatic benign tumors. The surgical excision is the first line treatment.

Case report: We report a case of 69-year old woman with an asymptomatic arteriovenous hemangioma of the right ventricle. Compete surgical excision was performed with the use of cardiopulmonary bypass and the patient was discharged on postoperative day 6 with no relapse at the six months follow-up.

Conclusion: The review of the literature showed that this procedure can be performed safely with excellent long-term results.
Keywords: cardiac hemangioma, right ventricle, surgery

Introduction

Hemangiomas of the heart are exceptionally rare benign tumors constituting 1-2% of all cardiac tumors, which may occur in all cardiac layers: pericardium, myocardium or endocardium. Their location in the right ventricle is highly uncommon and usually without any symptoms. Cardiac hemangiomas are clinically classified into three subcategories: capillary, cavernous and arteriovenous type. This report accounts for a case of arteriovenous cardiac hemangioma, an extremely rare subtype of this tumor.

Case report

We report a 69-year old woman without any reported symptoms who was accidentally diagnosed with the tumor of the right ventricle during a routine echocardiography. She is a nonsmoker with previous history of hypertension, under control by means of therapy. Transthoracic echocardiography showed a mass in the right ventricle with no tricuspid regurgitation, normal right ventricle diameter and normal left ventricular function. Cardiac MRI showed an intermediate-density mass 20x25 cm fixed with a small pedicle to the anterior wall of the right ventricle. The coronary angiography did not show signs of coronary disease. Laboratory test results were all within normal ranges, as well as serum tumor markers.

Under general anesthesia, the median sternotomy was performed. After the institution of bicaval cardiopulmonary bypass, the heart was arrested with warm blood cardioplegia in normothermic conditions. The tumor was resected completely with a clear margin through right atriotomy, there was no involvement of the tricuspid valve. There was no need for ventricle wall reconstruction.
The histopathology exam revealed the mixture of arterial and venous vessels confirming the diagnosis of arteriovenous hemangioma. (Fig2.a,c) Endothelial markers CD 31 were positive on immunohistochemical staining. (Fig2b) The patient was discharged on postoperative day 6 with the uneventful postoperative course. On six months follow-up, the patient is alive and well with no relapse of the hemangioma showing at control echocardiography.

Discussion

Cardiac hemangiomas are extremely rare benign tumors of the heart. They can occur in all three layers of the myocardium and can be present anywhere in the heart cavities or pericardium. They consist of small arterial or venous vessels and cavernous vascular channels, leading to the division on three subtypes: capillary, cavernous and arteriovenous. They are very uncommon in the right ventricle, especially the arteriovenous type presented in our case. The disease can appear in patients of all ages, and the clinical presentation depends on the localization and the size of the tumor – it can vary from asymptomatic to the signs of right ventricle congestion, but they are usually asymptomatic. (2) Cases have been described resulting in sudden cardiac death, rhythm disturbances in hemangioma localized in the vicinity in AV node and tamponade caused by a ruptured hemangioma in the pericardium. Tumors localized in the valvular apparatus can cause orifice obstruction and distal embolization. (3) The indication for surgical procedure was made due to malignant localization of the tumor, even it was supposed to be benign as well as for definite histopathology exam confirmation of the tumor type. Especially because the right-sided heart tumor mass is always suspicious for malignancy because of high frequency of cardiac metastases originate from primary malignant tumor (bronchogenic carcinoma, breast, hepatic and renal carcinoma) via venous dissemination. (4)

The diagnosis is set by means of echocardiography followed by confirmation of contrast-enhanced CT or cardiac MRI. Coronary angiography is useful in determining the relationship with coronary arteries if necessary, or to exclude the concomitant coronary artery disease if suspected. (5)
MRI was performed in order to visualize the tissue structure and the possible invasiveness of the tumor mass. MRI in addition to echocardiography has been showed to have significantly more correct histopathological diagnosis than the echocardiography alone, especially for the right-heart localization due to difficulty in obtaining this view on standard echocardiography. (6) Contrast enhancement feature of the MRI is highly predictive for malignancy, as well as the ability of the MRI to show exact localization and tissue invasiveness, in addition of pleural effusion presentation, which are all predictive for cardiac malignancy. The T1 and T2 sequence contrast density alongside with presence contrast enhancement can differentiate between different types of tumor masses. Cardiac lipomas are hyperintense while fibromas and myxomas are hypointense. Cardiac hemangiomas homogeneous, intermediate-to-high signal on T1-weighted, and diffusely hyperintense on T2-weighted images. (7)

The treatment of choice is surgical resection with a clear margin, with ventricle wall reconstruction if needed. (8)

The first hemangioma of the right ventricle was described by Hochberg in 1950 and up to date 35 cases are found in literature, including the one presented here. Median sternotomy was used predominately and the thoracotomy was used only in two cases. Total resection with a clear margin was performed in 31 patients, while four patients had the biopsy done without resection. Over 90 % of surgeries were performed with the use of CPB mainly, with the aortic cross clamping and the use of cardioplegic arrest of the heart. The follow-up data was available for 80% of patients (from 6 – 24 months) and there were no relapses or fatal outcomes. The main localization was the anterior wall (63%) of the right ventricle, while the right ventricle outflow tract (35%) had the most dramatic clinical presentation. Three tumors had an apical ventricular localization and these hemangiomas may require the RV wall reconstruction similar to the aneurysmectomy. Jiang and al. reported this procedure with a satisfactory postoperative cardiac function. (9)

Conclusion
Cardiac hemangiomas of the right ventricle are very rare and mostly asymptomatic benign tumors. The surgical excision is the first line treatment. The procedure can be safely done
with the use of cardiopulmonary bypass while the complete excision is mainly achieved with a low rate of recurrence and excellent long-term survival.

Conflict of interest: none declared.

Appendix

Figure 1. MRI imaging (a,b) and intraoperative image (c) showing the hemangioma in the right ventricle
Figure 2. Microscopic view of the arteriovenous hemangioma of the right ventricle (a,c) and the CD 31 positive marker staining (b)

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Received on November 08, 2017.
Accepted on February 06, 2018.
Online First February, 2018.