Epicardial cavernous hemangioma: a two-case report.

Atef Ben Youssef, Sarra Zairi, Sonia Ouerghi, Aida Ayadi*, Mouna Bousnina, Ferdaous Berraies, Tarak Kilani.

Department of Thoracic and Cardio-Vascular Surgery. A. Mami Hospital.Ariana. Medical School of Tunis. Tunisia.
** Department of Pathology. A. Mami Hospital. Ariana. Medical School of Tunis. Tunisia.*

A. Ben Youssef, S. Zairi, S. Ouerghi, A. Ayadi, M. Bousnina, F. Berraies, T. Kilani.

L'hémangiome caverneux épicardique: à propos de deux cas.

LA TUNISIE MEDICALE - 2014 ; Vol 92 (n°04) : 268-271

R É S U M É

But : Nous rapportons deux cas d'hémangiomes caverneux prenant naissance au niveau de l'épicarde chez deux femmes âgées respectivement de 24 et 79 ans.
Observations : La première patiente était symptomatique et avait consulté pour des palpitations. La deuxième patiente nous a été adressée après une découverte fortuite à l'échographie cardiaque. Une exploration par TDM thoracique complétée par une IRM a été effectuée dans les deux cas, et avait montré une masse située dans la cavité péricardique. Un complément d'exploration par un coro-scanner était nécessaire dans le premier cas, pour déterminer les rapports avec l'artère coronaire en regard. Les deux patientes ont eu une résection chirurgicale sous circulation extracorporelle, avec des suites simples pour la première. La deuxième est décédée en post-opératoire suite à une pneumopathie.
Conclusion : Les hémangiomes caverneux de localisation cardiaque, bien que rares et bien tolérés nécessitent une prise en charge rapide et une exérèse chirurgicale dès leur découverte.

M o t s - c l é s

Caverneux, hémangiome, épicarde, chirurgie.

A. Ben Youssef, S. Zairi, S. Ouerghi, A. Ayadi, M. Bousnina, F. Berraies, T. Kilani.

Epicardial cavernous hemangioma: a two-case report.

LA TUNISIE MEDICALE - 2014 ; Vol 92 (n°04) : 268-271

S U M M A R Y

Aims: We report two-cases of cavernous hemangiomas arising from the epicardium in two women aged respectively 24 and 79 years old. The first patient was symptomatic and presented with palpitations. The second patient was referred after a random discovery at echocardiography. Chest CT and MRI were performed in the two cases and showed a mass located in the pericardial cavity. Coronary CT was necessary in the first case to ascertain the degree of coronary artery involvement. Both of our patients underwent surgical resection under cardiopulmonary bypass with an uneventful postoperative course for the first case. The second one, died postoperatively from pneumonia.
Conclusion: Cardiac cavernous hemangiomas, although rare and well tolerated require prompt management and surgery at discovery to avoid further complications which may put at risk the patient's life prognosis.

K e y - w o r d s

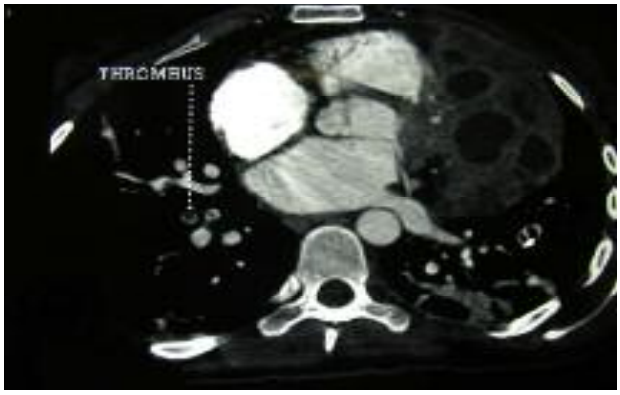
Cavernous, hemangioma, epicardium, surgery.

Cardiac cavernous hemangioma is an uncommon benign primary tumor of the heart. It may originate in any part but less commonly from the pericardium or epicardium. We report herein two cases of epicardial cavernous hemangioma managed during the past 5 years in the cardio-vascular surgery department of A. Mami Hospital.

Case n°1

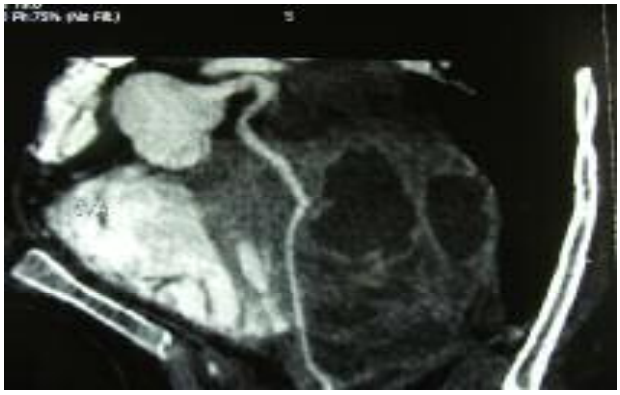
A 24-year-old woman, presented with a two-month history of chest pain and palpitations. Physical examination was unremarkable. Electrocardiogram showed a normal sinus rhythm with normal axis and T-wave inversion in leads V4 through V6. Chest X-ray showed a bulging of the left middle heart segment without pulmonary vascular redistribution or cardiomegaly. Tumor markers (CEA, AFP, HCG) were within normal limit. Transthoracic echocardiography revealed an anterior intrapericardial polycystic mass of 50cm2. It was in front of the pulmonary infundibulum and extended from the apex to the base of the heart, without compression of the cardiac cavities. Magnetic resonance imaging (MRI) revealed a 9×7cm pericardial tissue mass which was isointense on T1-weighted imaging and hyperintense on T2-weighted imaging. The mass showed a delayed, inhomogeneous enhancement after gadolinium administration because of interspersed septa within the mass. Coronary CT showed close contact with the myocardium and heterogeneous density with a tissue component. There was delayed enhancement in the post contrast imaging, with hypo-intense, unenhanced areas within the mass (Fig.1).

Figure1 : The mass showing delayed enhancement areas in the post contrast imaging.



The tumor surrounded completely the segments 2 and 3 of the left anterior descending coronary artery (LAD) and was associated to pericardial effusion (Fig.2). Patient was explored via sternotomy and after pericardotomy, the tumor was found on the left face of the heart, extended to the left and behind the pulmonary artery. Tumor infiltrated into the myocardium at the anterior wall of the right ventricle and lateral wall of the left ventricle. After cardiopulmonary bypass and under cardioplegic arrest, the tumor was incompletely resected, leaving a remnant close to the left anterior descending coronary artery, without opening of heart cavities. One diagonal branch running into the tumor has been sacrificed. The cardiac defect was closed in two layers with Teflon felt pledgets. Histological examination showed large vascular channels within the tumor, lined by flattened endothelial cells lining a fibrous stroma suggestive of cavernous hemangioma. The lining cells expressed endothelial markers CD34 and CD31 at immunohistochemistry studies. The postoperative clinical course was uneventful and the patient was discharged 10 days after surgery. Transthoracic echocardiography eight months later showed a remnant of hemangioma with a-69% left ventricular ejection fraction. Coronary CT 10 months later showed the small remnant of the tumor with normal cardiac cavities. The patient is currently asymptomatic 4 years after surgery.

Figure 2 : The tumor surrounding completely the segments 2 and 3 of the left anterior descending coronary artery.

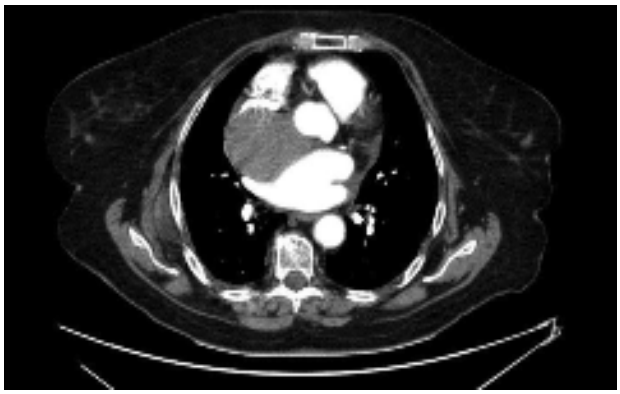


Case n°2

A 79-year-old woman with hypertension, a history of external electrical cardio-version for atrial fibrillation 4 years ago as well as a prior hysterectomy for a uterine cancer 12 years ago, was referred to our department for a random discovery of an intra-pericardial tumor with a large contact with the right atrium. The patient had a good general condition. Physical examination revealed an 80 beats/min cardiac arrhythmia with normal heart sounds. Chest X-ray showed a cardiomegaly with a bulging left middle arc. Electrocardiogram showed an atrial arrhythmia. Trans-thoracic echocardiography showed a 3x4cm round sessile fixed mass developing in the right atrium next to the superior vena cava contracting a large contact to the inter-atrial septum with a bi-atrial dilatation. Trans-esophageal echography showed a soft tissue mass of 65x45mm, with compression on the right atrium developing in the space bounded in front by the aorta and in below by the right atrium and the superior vena cava. A recent clot was visualized in the left atrium of 11x7mm. CT scan showed that the mass was adherent to the cardiac cavity and to the pericardium, presenting a homogeneous density without extension to the cardiac cavities or vascular compression (Fig.3). MRI showed a well-defined mass which was infiltrating the right atrial wall without

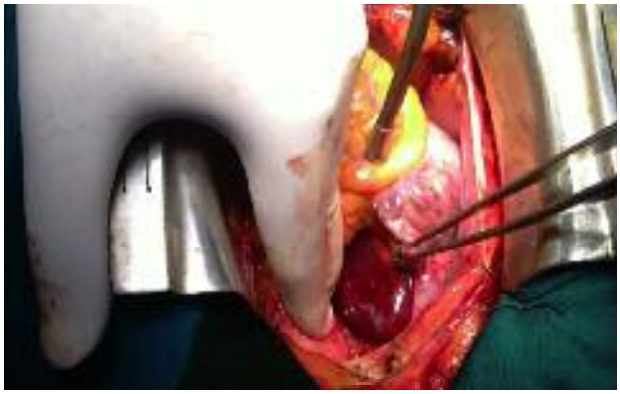
intraluminal extension. It was adherent to the right atrial heart walls and infiltrating the base of the aorta and both sides of the upper and lower cave system. An invasive cardiac tumor was highly suspected and a surgical approach was decided. A right thoracoscopy with pericardotomy was performed.

Figure 3 : The mass adherent to the cardiac cavity and to the pericardium, presenting a homogeneous density.



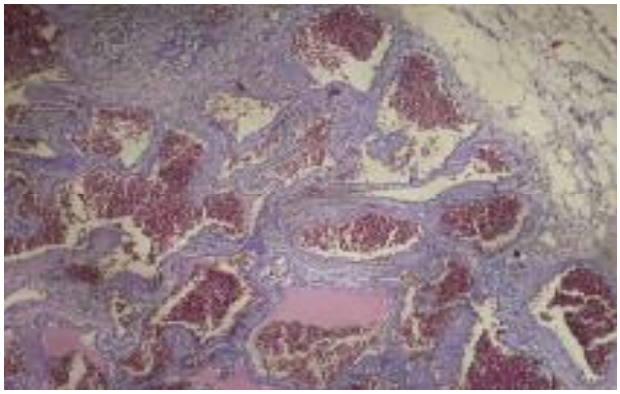
The tumor was sessile and tightly adherent to the right atrium. A conversion to sternotomy was decided. The tumor was developing at the postero-lateral face of the right atrium, behind the superior vena cava, extending to the aortico-caval space in contact with the roof of the left atrium and the pulmonary artery from which it was easily dissected (Fig.4).

Figure 4 : The tumor developing at the postero-lateral face of the right atrium.



Given the tight adherence to the right atrium, a cardiopulmonary bypass was decided. Complete dissection of the tumor from the right atrium was achieved without atriotomy and a total tumorectomy was possible. Frozen section was in favor of an hemangioma. Histological examination showed a 6x5x1.5 cm tender encapsulated mass with micro-cystic areas and important hemorrhagic reshuffle. There was a benign well limited vascular proliferation with vessels of various widths. These were lined by regular endothelial cells with enlarged lumen and areas of intercommunications filled with red blood cells (Fig.5). The diagnosis of intra-pericardial cavernous hemangioma was confirmed. The post-operative course was uneventful at the beginning. At the 7th day, the patient presented an ischemic stroke with peripheral signs of clotting. The laboratory tests revealed an immuno-induced thrombopenia. A respiratory distress secondary to nosocomial pneumonia was also diagnosed and the patient required mechanical ventilation and intra-venous antibiotics. The patient developed a septic shock two weeks later and died.

Figure 5 : TA vascular proliferation vessels with vessels of various widths, lined by regular endothelial cells with enlarged lumen.



DISCUSSION

Primary cardiac tumors are very rare and accounted for 0.056% among 12 485 consecutive autopsies [1]. Up to 75% of primary cardiac tumors are benign and cardiac hemangioma accounts for 1 to 2% of all benign tumors. We report two rare cases of cardiac hemangioma, which is a non-malignant vascular tumor that consists of closely packed capillary structures or widely dilated vascular channels lined by flattened endothelial cells and with focal connective tissue in the walls. It can be classified as capillary, arteriovenous or cavernous like in both of our cases [2]. It may originate from any part of the heart [3], but a previous review of 56 cases of cardiac hemangiomas, revealed a predilection for ventricles at 70% (36% right, 34% left ventricle) and 23% in the right atrium [4]. The pericardium is a rarely reported location [5] with hemangiomas arising from visceral pericardium, and most of the cavernous type [6]. Both of our patients illustrate this fact.

Hemangiomas may be encountered at any age and a slightly female predominance has been reported [7]. Cardiac hemangiomas are frequently well tolerated and asymptomatic in most of the patients with incidental discovery at imaging. For our cases, the presenting symptoms were chest pain and palpitations in the first case and an imaging finding in the second case.

Echocardiography is an efficient tool in determining the characteristics of any cardiac tumor with an accuracy rate of 81% [9]. Hemangiomas appear at echocardiography as hyperechoic lesions. Further examinations are recommended in

case of symptoms of coronary artery disease or a doubtful echography. CT and MRI are highly contributive in determining the extension of the tumor to the neighboring vessels and myocardium [9].

Chest CT shows a heterogeneous mass at unenhanced sequences and intensely enhanced after contrast material administration [10]. On MRI, hemangioma appears heterogeneous isotense or hypointense on T1-weighted, and usually hyperintense on T2-weighted images [11]. Cardiac catheterization studies reveal an intra-cavitary filling defect which may help the diagnosis of a cardiac tumor in 40% of cases. Coronary arteriography helps to establish the diagnosis with the classic finding of a vascular blush, which reflects the vascular nature of the tumor [10, 12]. Coronary CT is indicated in some patients, such our first case, in which an accurate determining of the tumor extent to the coronary vessels was mandatory. The different images modalities, even if accurate, can't certify the diagnosis which remains histological. The natural course of a hemangioma is not well known: spontaneous regression has been reported as well as various complications and even sudden death [13, 14]. For these reasons, hemangioma (such as any cardiac tumor) when resectable, has to be removed. Complete excision is the mainstay for treatment: it eases the symptoms and prevents complications. The long-term prognosis for operated patients is favorable, without relapses for completely removed hemangiomas. However, a regular screening with different imaging modalities is highly recommended in all cases, especially for incompletely resected tumors.

CONCLUSION

Cavernous epicardial hemangiomas are benign tumors but require prompt surgery when discovered, even for asymptomatic patients, to confirm the diagnosis and prevent farther complications.

References

References:

1- Lam KY, Dickens P, Chan AC. Tumors of the heart. A 20-year experience with a review of 12 485 consecutive autopsies. Arch Pathol Lab Med 1993; 117: 1027-31.

2- Chiappini B, Gregorini R, Vecchio L, Petrella L, Di Pietrantonio F, Giancola R. Cardiac hemangioma of the left atrial appendage: a case report and discussion. J Card Surg 2009; 24:522-3.

3-Yaganti V, Patel S, Yaganti S, Victor M. Cavernous hemangioma of the mitral valve: a case report and review of literature. J Cardiovasc Med (Hagerstown) 2009; 10:420-2.

4-Kojima S, Sumiyoshi M, Suwa S et al. Cardiac hemangioma: a report of two cases and review of the literature. Heart Vessels 2003;18: 153e6.

5- Ramasubbu K, Wheeler TM, Reardon MJ, Dokainish H. Visceral pericardial hemangioma: unusual location for a rare cardiac tumor. J Am Soc Echocardiogr 2005;18: 981.

6- Brodwater B, Erasmus J, McAdams HP, Dodd L. Case report. Pericardial hemangioma. J Comput Assist Tomogr 1996; 20: 954-6.

7-Matsumoto Y, Watanabe G, Endo M, Sasaki H. Surgical treatment of a cavernous hemangioma of the left atrial roof. European Journal of Cardio-thoracic Surgery 2001; 20: 633-5.

8-González López MT, Aranda Granados PJ, Delange Segura L, Gutiérrez de Loma J. An unexpected left atrial cavernous hemangioma: the cardiac surgeon needs an optimal preoperative study! Interactive CardioVascular and Thoracic Surgery. 2011; 13 :529-31.

9- Oshima H, Hara M, Kono T, Shibamoto Y, Mishima A, Akita S. Cardiac hemangioma of the left atrial appendage: CT and MR findings. J Thorac Imaging 2003; 18: 204e6.

10- Grebenc ML, Rosado de Christenson ML, Burke AP, Green CE, Galvin JR. Primary cardiac and pericardial neoplasms: radiologic–pathologic correlation. Radiographics 2000; 20:1073-103, quiz 1110-1071.

11-Sparrow PJ, Kurian JB, Jones TR, Sivananthan MU. MR imaging of cardiac tumors. Radiographics, 2005; 25: 1255-76.

12-Esmaeilzadeh M, Jalalian R, Maleki M, Givtaj N, Mozaffari K, Parsaee M. Cardiac cavernous hemangioma. Eur J Echocardiography 2007; 8: 487-9.

13-Palmer TE, Tresch DD, Bonchek LI. Spontaneous resolution of a large, cavernous hemangioma of the heart. Am J Cardiol 1986; 58:184-5.

14-Abad C, Campo E, Estruch R et al. Cardiac hemangioma with papillary endothelial hyperplasia: Report of a resected case and review of the literature. Ann Thorac Surg 1990; 49: 305-8.