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ATRIAL HAEMANGIOMA – A CASE REPORT AND REVIEW OF THE LITERATURE

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Abstract: Primary cardiac tumours are rare entities and haemangiomas represent only 2.8% of these tumours. Haemangiomas are usually revealed by various symptoms depending on their size and location or can be incidentally discovered during routine examination or at autopsy. We report the case of a 70 years old Caucasian male referred for autopsy after being found deceased in his apartment. The main cause of death was considered to be haemorrhagic stroke secondary to rupture of an anterior communicating artery aneurysm with bronchopneumonia as a concurring cause. Heart dissection revealed the presence of a round, reddish, tumour adherent to the posterior wall of the left atrium and unrelated to the cause of death. Microscopic examination established the diagnosis of cavernous haemangioma. An interrogation of PubMed database for the time interval between 2008 and 2018 revealed only 11 cases of cardiac haemangiomas, 6 of them of cavernous type. In conclusion, cardiac haemangiomas are exceptional discoveries but cardiologists and cardiac surgeons should be aware of their existence as the symptoms and medical imaging aspects are non-specific and potentially mislead the diagnosis.

Key words: primary cardiac tumour, cavernous haemangioma, autopsy, histological examination.

INTRODUCTION

Primary cardiac tumours are rare entities whose incidence ranges from 0.001% to 0.3% [1-3]. Reynen quantified 157 (0.021%) cases in an analysis of 22 autopsy series including 731,309 cases [4]. Between 79-85% of these tumours are benign [5], and haemangioma represent only 2.8% of primary cardiac tumours [3]. Haemangiomas are more frequent in males with a mean age at diagnosis of 43 years, may develop in any heart cavity, some are asymptomatic or revealed by various symptoms depending on their location and size (arrhythmias, heart failure, valve or outflow tract obstruction, pericardial effusion) [6]. We report a case of atrial haemangioma discovered at autopsy in a 70 years old male patient. Furthermore, we performed a review of clinical signs and symptoms, diagnosis and treatment methods of this rare entity less known by clinicians.

CASE REPORT

A 70 years old Caucasian male was submitted to autopsy after being found deceased in his apartment by his son who entered his home because he has not been answering the mobile phone for 2 days. The medical file from his general practitioner indicated that he was diagnosed and treated for cardiovascular diseases (severe arterial hypertension, lower limbs venous insufficiency) and had a history of asthma and duodenal ulcer with no spirometry and echocardiography report in his personal medical registry.

Macroscopic evaluation

The autopsy was performed about 24 hours after death. Gross examination showed a hypersthenic

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constitution, lower limbs varices with varicose ulcers, liver congestion, pulmonary stasis and focal oedema in the right upper lobe of the left lung, fibrinous pleural fluid in the left pleural cavity, cardiomegaly (558 grams) with normal sized aorta, intracerebral haematoma in the left frontal lobe, subarachnoid haemorrhage and a ruptured aneurysm of the anterior communicating artery. Heart dissection revealed the presence of a reddish round mass with a diameter of approximately 3 cm in the left atrium, adherent to the posterior wall (Fig. 1). The mass was excised, and fragments were submitted for histopathological examination.

Microscopic evaluation

Tissue fragments collected at the autopsy have been fixed in formalin followed by paraffin embedding. Paraffin-embedded sections were mounted onto slides for haematoxylin- eosin (H&E) and Masson's trichrome staining. Microscopic examination of ventricular fragments revealed endocardial fibrosis associated with intramyocardial areas of fibrosis, focal myocyte hypertrophy and coronary atherosclerosis. When analysing left atrium fragments, a tumoral proliferation with large vascular spaces separated by conjunctive septa was identified on the epicardial aspect with infiltration of the adjacent myocardium up to the subendocardial layer (Fig. 2-4). The vascular spaces contained blood or thrombi and were lined by endothelial cells presenting no atypia.

Examination of lung sections revealed bronchopneumonia foci associated with fibrinous pleuritis thus confirming the macroscopic diagnosis. Examination of brain sections showed areas of haemorrhagic necrosis and ischemic lacunar spaces associated with oedema, subarachnoid haemorrhage and passive vascular congestion of intracerebral and

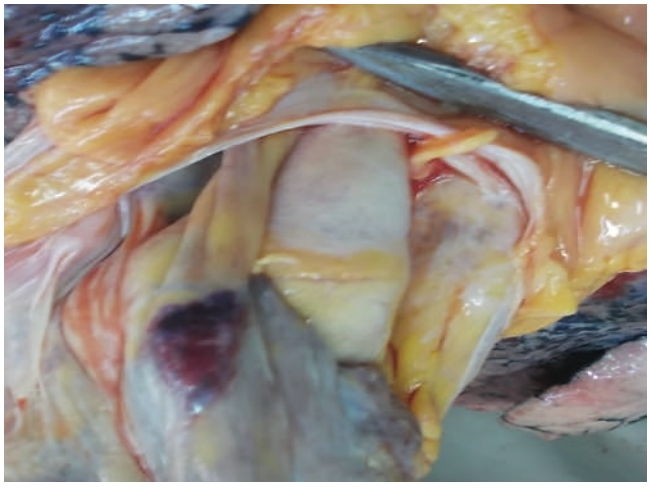

Figure 1. Gross aspect of the tumour.

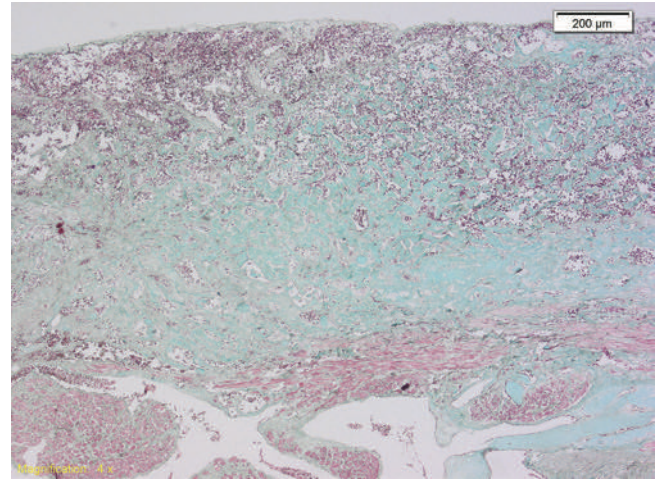

Figure 2. Overviewn of the tumoral proliferation (trichrome Masson staining x 40).

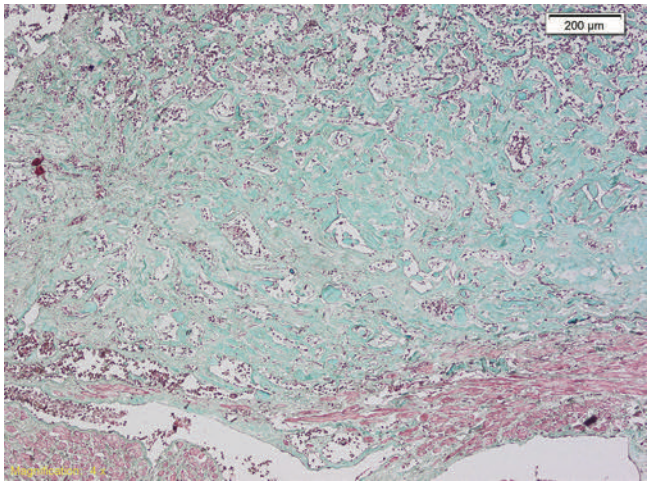

Figure 3. Vascular spaces separated by conjunctive septa located in the epicardium (trichrome Masson staining x 100).

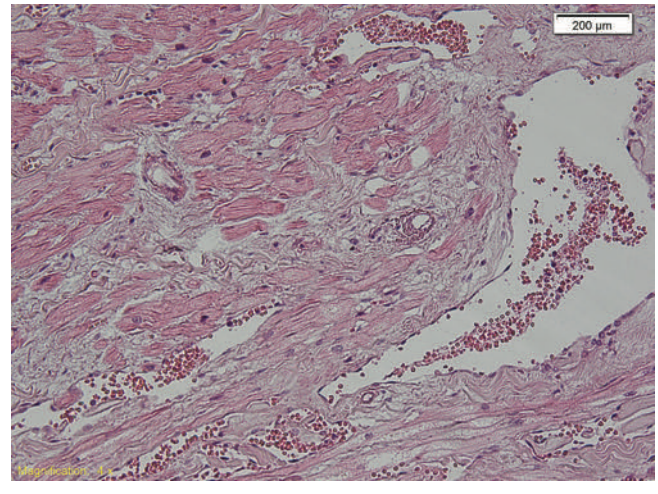

Figure 4. The tumour proliferation also invaded the left atrial myocardium (H&E staining x 200).

leptomeningeal vessels.

The main cause of death was considered to be haemorrhagic stroke secondary to rupture of an anterior communicating artery aneurysm with bronchopneumonia as a concurring cause.

DISCUSSIONS

The first cardiac cavernous haemangioma was described in 1893 by McAllister [7, 8]. Such lesions have been reported at all ages irrespective to sex, with a slightly male predominance [9, 10].

Haemangiomas can develop in any layer of the heart wall, the most interested being the endocardium [11], followed by the atrial and ventricular myocardium [8,12], interatrial [13] and interventricular septum [14,15], and lastly, the epicardium [16].

Most cases of cavernous haemangiomas involve the ventricles, both right and left. A review of 56 cases reported that 20 have interested the right ventricle, 19 the left ventricle, 13 the right atrium, 6 the interventricular septum, 6 the interatrial septum and only 4 the left atrium [17]. In the presented case, the tumour was adherent to the posterior wall of the left atrium.

The authors reviewed all articles reporting cases of haemangiomas with atrial localisation published between 2008-2018 and indexed in PubMed database. Only case reports written in English language, available in full text and well documented have been analysed. Review articles have been excluded. There were identified 11 cases of atrial haemangiomas, 5 involving the walls of the right atrium, 1 the interatrial septum and 5 the walls of the left atrium (Table 1). All cases but one were symptomatic. In one case, the tumour was discovered incidentally following routine tests. Histopathological analysis of resected fragments established the diagnosis of cavernous haemangioma in 5 cases, capillary haemangioma in 2 cases and visualised mixed aspects in 2 cases (Table 1).

The most reported symptoms were dyspnoea, atypical precordial pain, pericardial effusion, embolization related symptoms, cardiac failure and arrhythmias in case of involvement of the conduction

Table 1. Cases of cardiac haemangiomas identified in analysed articles

Case no.	Age (yrs)	Sex	Localization	Size (cm)	Histological type	Symptoms	Medical imaging	Treatment	Reference
1	67	F	left atrium	9.5 x 7.5 x 6.0	NA	dyspnoea and palpitations	CT, MRI	surgical resection	Abu Saleh *et al.*, 2016 [2]
2	49	F	right atrium	3.5×3.5×2.5	cavernous	dyspnoea and arrhythmias	echo, CT, MRI	surgical resection	De Filippo *et al.*, 2010 [5]
3	74	M	right atrium	5.5 x 5	mixed (capillary, cavernous, and arteriovenous)	asymptomatic	echo, RX, MRI	surgical resection	Sanchez-Enrique *et al.*, 2015 [21]
4	74	M	left atrium	2×1.5	capillary	mild dyspnoea chest discomfort	echo	surgical resection	Hong *et al.*, 2014 [22]
5	76	F	left atrium	1.7 × 1.4	capillary	progressive dyspnoea	echo	surgical resection	Abad *et al.*, 2008 [26]
6	60	M	right atrium	0.63 × 0. 6 × 0.57	cavernous	intermittent chest discomfort	echo, MRI	surgical resection	Yuan *et al.*, 2008 [27]
7	61	F	interatrial septum	NA	cavernous	asymptomatic	echo, CT. MRI	surgical resection	Hrabak-Paar *et al.*, 2011 [28]
8	54	M	right atrium	2.4 x 2.5	cavernous	dizziness for months	echo, CT	surgical resection	Turak *et al.*, 2012 [29]
9	44	M	right atrium	0.5 × 0.4 × 0.3	cavernous	precordial dull pain	echo, PET-CT	surgical resection	Li *et al.*, 2014 [30]
10	35	F	left atrium	not available	mixed (cavernous-capillary)	anasarca	echo, RX, CT, MRI	surgical resection	Nishio et l., 2015 [31]
11	82	F	left atrial roof	10.2 x 6.8 x 7.1	cavernous	dyspnoea, bilateral lower extremity oedema	echo, CT, MRI	surgical resection	Nakamura *et al.*, 2018 [32]
Our case	70	M	left atrium, posterior wall	3 x 3 x 2.7	cavernous	unknown	-	autopsy	-

system [16]. Severity of clinical symptoms depended on the location and extent (infiltration of the cardiac wall) of the tumour. Haemangiomas involving in the interatrial septum can be asymptomatic [13]. Two authors have signalled cases of previously asymptomatic atrial haemangiomas as cause of sudden death, the diagnosis being established secondary to autopsy findings [2, 3].

Modern medical imaging (echocardiography, computed tomography, magnetic resonance) can accurately identify, diagnose and locate the tumour and also characterise its invasive pattern [18].

At gross examination, cardiac haemangiomas occur most frequently as single/solitary lesions, with sessile or polypoid aspect and varying sizes between 2-3.5 cm [19].

According to microscopical examination, several types can be described: cavernous, capillary, arteriovenous or cirsoid aneurysm [13, 20]. Cavernous haemangioma is formed by multiple large vascular spaces, lined by endothelial cells displaying no atypia and separated by fibrous tissue. Capillary haemangioma is formed by a proliferation of capillary vessels in a conjunctive environment. Arteriovenous haemangioma is rare and formed by multiple dysplastic malformed arteries and veins [20].

In some cases, cardiac haemangiomas can display a combination in various proportions of the previously reported aspects together with fibrous and adipose areas like haemangiomas located in skeletal muscles [20, 21].

The evolution of cardiac haemangiomas is unpredictable [22], as they can extend or suddenly regress [23-25].

In order to prevent complications due to embolization, compression, infiltration of conduction system and valve dysfunction, surgical excision is advised in all cases [3, 26]. Complete excision is generally associated with a low recurrence rate [16,18]. Differential diagnosis is generally being made with angiosarcoma characterised by marked atypia, central necrosis and increased proliferation rate [1].

The association of cardiac cavernous haemangioma with a cerebral aneurysm has not been previously reported and a genetic cause can be suspected.

In conclusion, cardiac haemangiomas are exceptional discoveries, but cardiologists and cardiac surgeons should be aware of their existence as the symptoms and medical imaging aspects are non-specific and potentially mislead the diagnosis.

Conflict of interest

The authors declare that they have no conflict of interest.

References

1. Jain D, Maleszewski JJ, Halushka MK. Benign cardiac tumors and tumorlike conditions. Ann Diagn Pathol. 2010;14(3):215-230.
2. Abu Saleh WK, Al Jabbari O, Bruckner BA, Reardon MJ. Case Report: A Rare Case of Left Atrial Hemangioma: Surgical Resection and Reconstruction. Methodist Debakey Cardiovasc J. 2016;12(1):51–54.
3. Vargis RS, Phansalkar M, Padhi S, Phansalkar D, Nair SR. Pericardial Haemangioma: A Common Tumour in an Unusual Location: Case Report and Review of Literature. J Clin Diagn Res. 2017;11(5):ED15–ED17.
4. Reynen K. Frequency of primary tumors of the heart. Am J Cardiol. 1996;77(1):107.
5. De Filippo M, Corradi D, Nicolini F, Rastelli A, Ugo F, Araoz PA, Sverzellati N, Gherli T, Zompatori M. Hemangioma of the right atrium: imaging and pathology. Cardiovasc Pathol. 2010;19(2):121-124.
6. Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumours: diagnosis and management. Lancet Oncol. 2005;6(4):219-228.
7. Esmaeilzadeh M, Jalalian R, Maleki M, Givtaj N, Mozaffari K, Parsaee M. Cardiac cavernous hemangioma. Eur J Echocardiogr. 2007;8(6):487-489.
8. Ouerghi S, Youssef AB, Ouechtati W, Zidi A, Mezni F, Belhani A, Kilani T. Cardiac cavernous haemangioma. Heart Lung Circ. 2011;20(3):197-201.
9. Thomas JE, Eror AT, Kenney M, Caravalho J. Asymptomatic right atrial cavernous hemangioma: a case report and review of the literature. Cardiovasc Pathol. 2004;13:341-344.
10. Erdoğan İ, Sarıalioğlu F. Cardiac evaluation in children with hemangiomas. Turk Kardiyol Dern Ars. 2004;13(6):341-344.
11. 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(5):420-422.
12. Harada H, Hirata T, Utoh J, Kunitomo R, Hara M, Kitamura N. Cardiac hemangioma of the right ventricle-report of a case (Japanese). Jpn J Thorac Cardiovasc Surg. 1998;46(7):629-633.
13. Ayadi-Kaddour A, Abid M, Harrath Y, Smati B, Kilani T, El Mezni F. Asymptomatic hemangioma of the interatrial septum. J Thorac Cardiovasc Surg. 2006;132(3):691-692.
14. Tadros NB, Akl BF, Avasthi P, Crotta P, Wernly JA, Pett SB, Croke R, Newell JD 2nd, Frierson J. Arteriovenous and capillary hemangiomas of the interventricular septum. Ann Thorac Surg. 1988;46(2):236-238.
15. Han Y, Chen X, Wang X, Yang L, Zeng Y, Yang J. Cardiac capillary hemangioma: a case report and brief review of the literature. J Clin Ultrasound. 2014;42(1):53-56.
16. Shikata D, Nakagomi T, Yokoyama Y, Yamada Y, Nakajima M, Oyama T, Goto T. Debulking surgery for venous hemangioma arising from the epicardium: report of a case. World J Surg Oncol. 2017;15:81.
17. Kojima S, Sumiyoshi M, Suwa S, Tamura H, Sasaki A, Kojima T, Mineda Y, Ohta H, Matsumoto M, Nakata Y. Cardiac hemangioma: a report of two cases and review of the literature. Heart Vessels. 2003;18(3):153-156.
18. Eftychiou C, Antoniades L. Cardiac hemangioma in the left ventricle and brief review of the literature. J Cardiovasc Med (Hagerstown). 2009;10(7):565-567.
19. Chao JC, Reyes CV, Hwang MH. Cardiac hemangioma. South Med J. 1990;83(1):44–47.

20. Travis W, Brambilla E, Müller-Hermelink K, Harris C. Pathology and Genetics of Tumours of the Lung, Pleura, Thymus and Heart. Lyon:IARC Press;2004.
21. Sánchez-Enrique C, Vivas D, Carnero Alcázar M, Vilacosta I. Giant right atrial hemangioma: Correlation between magnetic resonance imaging and histopathologic evaluation. J Thorac Cardiovasc Surg. 2015;150(4):e51-53.
22. Hong SY, Park KT, Lee YH, Cho KH, Seo JS, Han IY. Cardiac hemangioma: a case report. Korean J Thorac Cardiovasc Surg. 2014;47(2):149-151.
23. Gupta N. Intrapericardial hemangioma: A case report. J Clin Diagnostic Res. 2013;7(1):169–170.
24. Ben Youssef A, Zairi S, Ouerghi S, Ayadi A, Bousnina M, Berraies F, Kilani T. Epicardial cavernous hemangioma: a two-case report. Tunis Med. 2014;92(4):268-271.
25. Vakili H, Khaheshi I, Foroughi M, Ghaderi H, Esmaeeli S, Jafari M. Cardiac Hemangioma of RVOT in a Patient with Atypical Chest Pain. Case Rep Med. 2014;2014:285479.
26. Abad C, de Varona S, Limeres MA, Morales J, Marrero J. Resection of a left atrial hemangioma. Report of a case and overview of the literature on resected cardiac hemangiomas. Tex Heart Inst J. 2008;35:69–72.
27. Yuan SM, Shinfeld A, Kuperstein R, Raanani E. Cavernous hemangioma of the right atrium. Kardiol Pol. 2008;66(9):974-976.
28. Hrabak-Paar M, Hübner M, Stern-Padovan R, Lušić M. Hemangioma of the interatrial septum: CT and MRI features. Cardiovasc Intervent Radiol. 2011;34 Suppl 2:S90-93.
29. Turak O, Ozcan F, Başar FN, Ozen A, Birincioğlu CL, Okten RS, Topaloğlu S. Cavernous hemangioma of the right atrium: a very rare case of complete atrioventricular block. J Am Coll Cardiol. 2012;60(13):1204.
30. Li ZY, Li XC, Wu Q. PET-CT diagnosis of cardiac cavernous hemangioma with large pericardial effusion. Eur Rev Med Pharmacol Sci. 2014;18(21):3256-3259.
31. Nishio S, Kosuga K, Miwa S, Fujiwara Y, Katsuyama K, Hata T, Okada M, Takeuchi Y, Takeda S, Inuzuka Y, Seki J, Takeuchi E, Terashima T, Ikeguchi S. Rare case of cardiac hemangioma causing massive pericardial effusion: can a left atrial tumor produce pericardial effusion? Circulation. 2015;131(3):e21-23.
32. Nakamura E, Nakamura K, Furukawa K, Ishii H, Kawagoe K, Ideguchi T, Oguri N. A Giant Cardiac Cavernous Hemangioma Involving the Left Atrial Roof in an Elderly Woman. Ann Thorac Cardiovasc Surg. 2019;25(1):60-63.