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Review Article

Primary Cardiac Hemangioendothelioma in the Adult and Cardiac Involvement in Patient with Non-Cardiac Hemangioendothelioma. Review of the Literature

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Abstract

Background: Cardiac hemangioendothelioma (CHE) in the adult and cardiac involvement in adult patient with non cardiac-hemangioendothelioma (NC-HE) were rare and uncommon in daily clinical experience.

Methods: We researched in PubMed, Web of Science, Scopus, Cochrane library, and Medline for identifying relevant studies, case series, review and case reports.

Results: We found 33 cases of CHE in the adult and 7 cases of cardiac involvement in NC-HE patients. According to scientific data, the most of CHE patients were female (52%), histological type epithelioid (64%), with main localization in right atrium (41%), and the main clinical diagnosis was incidentally discovered on echocardiogram. The main therapeutic choice was surgical. Otherwise the cardiac metastasis in NC-HE patients (7 cases reported) were most frequent in the hepatic hemangioendothelioma primary localization (57%) than others (spleen, bone, limb, intracranial). The most of patients were female (85.7%) and the main clinical presentation were heart failure (85%).

Conclusion: CHE and cardiac metastasis of NC-HE patients in the adult were rare and unusual neoplasms. The early diagnosis and surgical treatment in each case demonstrated an increase of patients' survival and quality of life. We explained the scientific data to increase physician knowledge about this rare adulthood vascular neoplasm.

Keywords: *Hemangioendothelioma; Primary Cardiac Tumor; Metastatic cardiac tumor; cardiac tumor; cardiac neoplasm; cardiac metastasis.*

Introduction

Historical background: Hemangioendothelioma (HE) was described for the first time by Mallori in the 1908 and it was defined as a vascular neoplasms characterized by intermediate features between hemangiomas and angiosarcomas [1, 2]. With the technological evolutions, Stout and colleagues [3] studied firstly in deep the microscopic appearances in the 1943 and explained the likeness of HE to the blood vessels. In the 1967, Fernholz edited one of the first article on "Contribution of History of HE". Only in the 1988, Enzinger and colleague defined the HE features in intermediate, borderline, or low grade malignancy.

Classification and Clinical presentation: HE was a vascular neoplasm with typical prevalent proliferation of endothelial cells and is was distinguished according to its histological characteristics as [2, 4]:

- papillary intralymphatic,
- retiform hemangioendothelioma,
- kaposiform hemangioendothelioma,
- epithelioid hemangioendothelioma,
- pseudomyogenic hemangioendothelioma
- and composite hemangioendothelioma

HE was characterised by suggestive histological feature as intracytoplasmic vacuoles, nuclear cytoplasmic inclusion, myxoid stroma, hyaline stroma and chondroid stroma (Figures 1 a and b). The most of patient had expression of vascular markers (CD 31, CD 34, ERG, FL1I) and cytokeratins (CAM 5.2, CK7, CK 18) [5].

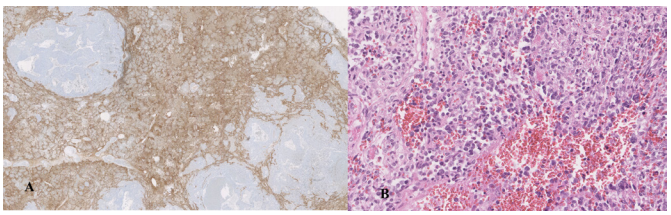

Figure 1: Kaposiform hemangioendothelioma of the right atrium associated to KAsabach-Merritt syndrome: a) image of the vascular neoplasm with epithelial membrane antigen (EMA) antibodies and b) eosin and hematoxylin view.

HE involved mainly liver, lung, skin, soft tissue, bone and spleen and occurring firstly in infancy and early childhood, in the adult it was very uncommon [2].

Each of the histological types of HE had own typical presentations and patterns, such as association with lymphatic vessel proliferation as well as Kasabach-Merritt syndrome (KMS) [6-9]. KMS included thrombocytopenia, microangiopathic hemolytic anemia and mild consumptive coagulopathy, and developed often in KHE, Kaposiform lymphatic anomaly (KLA) and tufted angioma (TA) [7, 8].

HE would have different grades of aggression like:

- finite
- low aggression
- mild aggression
- moderate aggression
- severe aggression, like Kaposiform HE
  - Local to the adjoining structures
  - Metastatic

Singularly, epithelioid HE was defined in two histological grade according to its aggressiveness: classic (low-grade) and malignant (intermediate-grade) [5-55].

The progression of HE was unpredictable: sometimes it grew slowly and sometimes the tumour was more active and spread quickly [2]. The characteristics of the patients, the different clinical and spreading of HE influenced the therapeutic choices: surgical, medical or palliative.

Finally, also the mortality depended to histological type and grade of aggression and clinical correlations.

Methods

We researched english article on PubMed, Web of Science, Scopus, Cochrane library, and Medline for identifying relevant english studies, reviews, case series and case reports in the adult with the following keywords: 1) cardiac hemangioendothelioma, 2) primary cardiac hemangioendothelioma, 3) cardiac metastasis in hemangioendothelioma, 4) Cardiac Involvement in Hemangioendothelioma in the adult, and 5) Hemangioendothelioma/ Heart/ Adult. We found 272 articles with those criteria (Figure 2) and we selected and chosen 33 case reports and 7 reviews that satisfied our research criteria (Figure 2).

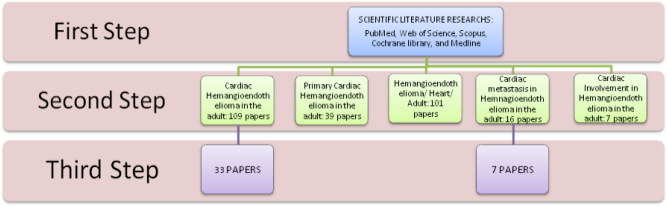

Figure 2: Description of scientific literature review process: first step we had researched all article in the literature

Results

According to our scientific research, the primary cardiac hemangioendothelioma was 33 reported cases. The patient age range was from 19 to 77 year-old, the average age was 45.1 ± 16.5 year-old (Table 1). The most of patients were female (n=17, 52%, Figure 3). The most common primary cardiac localization was right atrium (n=14, 41%) then the cardiac valve (n=5, 15%), left atrium (n=4, 12%), right ventricle (n=3, 9%), coronaric sinus (n=2, 6%), superior vena cava/ right atrium (n=2, 6%), right appendage (n=1, 3%), left ventricle (n=1, 3%), and not defined (n=2, 6%) (Figure 4). The histopathological feature of the HE in the case reported was predominantly ephiteloid (n=21, 66%), kaposiform (n=1, 3%) and defined malignant (n=3, 9%) and not defined (n=8, 24%) (Figure 4). The clinical presentations of these patients were characterised by incidentally discovered on echocardiogram (n=11, 34.4%), dyspnoea (n=2, 6.3%), chest pain (n=4, 12.5%), alveolar hemorrhage (n=1, 3.1%), cardiac tamponade (n=3, 9.4%), incidentally discovered an heart murmur (n=3, 9.4%), arrhythmia (n=1, 3.1%), cerebrovascular event like stroke (n=1, 3.1%), widened superior mediastinum noticed on a routine chest radiograph (n=1, 3.1%), thromboembolism (n=1, 3.1%), and incidentally discovered by autopsy (n=1, 3.1%) (Figures 5 and 6). The most of patients were CD31 and CD34 positive (30.3%) and CD 99 positive (18.1%). The main therapy were the surgery (n=21, 65.6%). In the case of cardiac metastasis of hemangioendothelioma (seven reported cases in literature), the age of patients were 61.7± 12.7 year-old and they were female (85.7%). The first localization of the tumour was indicated in the Table 2. In all the cases, the patient suffered of heart failure and the most of them were undergone to surgery.

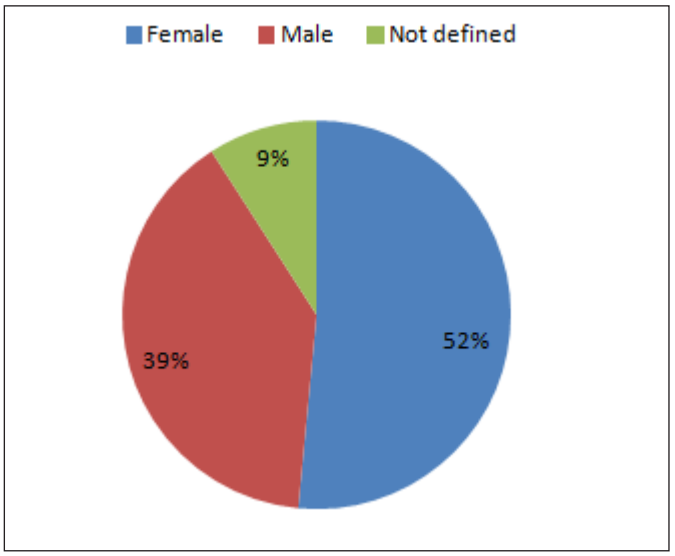

Figure 3: The percentage of cardiac hemangioendothelioma localization in the reported literature cases.

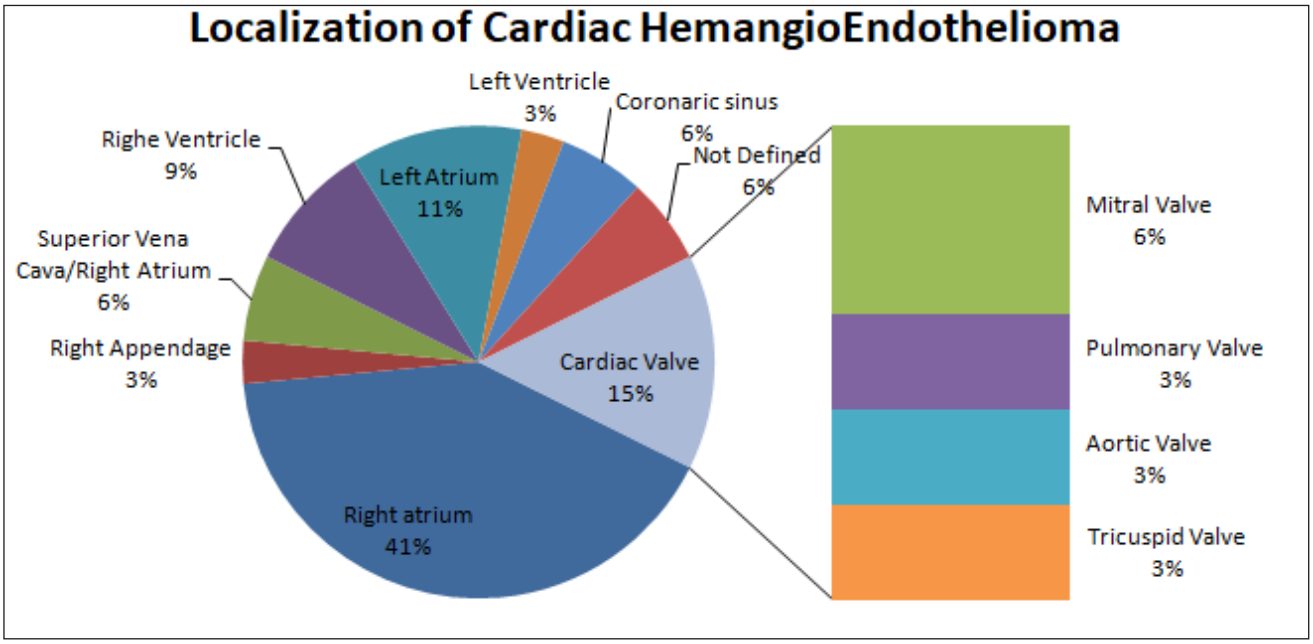

Figure 4: The percentage of cardiac hemangioendothelioma histological type in the reported literature cases.

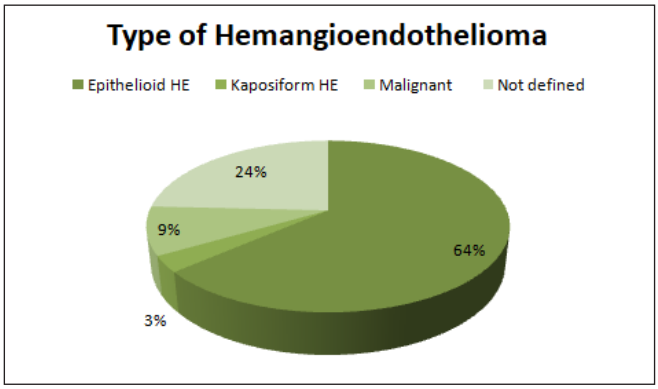

Figure 5: Type of hemangioendothelioma

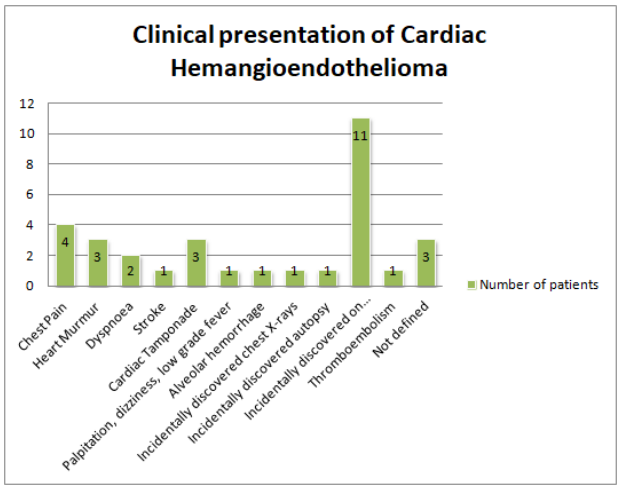

Figure 6: The clinical presentation of cardiac hemangioendothelioma in the reported literature cases were absolutely unpredictable in the adult with a major incidence of chest pain or dyspnea but wide variability in the clinical first manifestation due to patient's health background, histological type of hemangioendothelioma and grade of aggression.

Discussion

The cardiac hemangioendothelioma was predominantly epithelioid type (66% of the cases), with a peculiar tumour site in right atrium (40% of the cases). The most of these patients were female and they had good outcome (follow up positive after 6 months in the 46.9%). In particularly, the patients undergone to aggressive and rapid surgery, were better outcome than delay diagnosis or late surgical procedure [15-45]. As reported by scientific literature, cardiac malignant tumours were rare and uncommon, especially metastatic cardiac involvement [53, 54]. Cardiac hemangioendothelioma were considered an atypical unexpected malignant cardiac tumours with high local aggressiveness and metastasizing potential [30-32, 34, 36-37].

On the other hand, we had researched the reported articled of cardiac metastasis in patient with non cardiac hemangioendothelioma reported in Table 2 [48-52]. Bisesi and colleagues [14], had already firstly documented the possibility of cardiac involvement in multifocal epithelioid hemangioendothelioma in 1996, hence we had studied in deep the scientific literature to look for other cases like this and to understand the clinical presentation and evolution of the cardiac metastasis of a hemangioendothelioma.

In our research, we had found 6 cases of cardiac metastasis of hemangioendothelioma. All of the cases were in female (100%) with a primary HE in liver (50%), one ovarian HE, one bone HE and the last intracranial HE. The most of them showed heart failure and worsening hemodynamic parameters in association with coagulative disorders and hence end--stage of multi-organ failure. The prognosis of them was unfavourable specially in the presence of coagulative disorders.

Conforming in Table 2 and our review of the literature, cardiac metastasis in patients with non-cardiac hemangioendothelioma were out of ordinary and exceptional but remarkable for the prognosis of the patients [14, 48-52].

Table 1. Cases of cardiac hemangioendothelioma in the adult reported in scientific literature. M: male, F: female, ND: non defined, RA: right atrium, MV: mitral valve, LA: left atrium, RV: right ventricle, LV: left ventricle, PV: pulmonary valve, AV: aortic valve, TV: tricuspid valve, CS: coronaric sinus, SCV: superior vena cava, F-up: follow up.

	Reference	Year	Age	Sex	Site	HE Type	Presentation	Therapy	Follow up
1	Blanchard et al[15]	1952	29	M	Right Appendage	ND	Chest pain	ND	ND
2	Crenshaw et al[16]	1959	36	F	RA	ND	Chest pain	ND	ND
3	Allaire et al[17]	1964	ND	ND	ND	ND	ND	ND	ND
4	Hayward et al[18]	1979	49	F	MV	Epithelioid	Diastolic murmur typical of mitral stenosis	Surgical	Alive after 4 years
5	Di Biasi et al[20]	1988	ND	M	RV	Epithelioid	Incidentally discovered on echocardiogram	Surgical	ND
6	Singal et al[19]	1987	19	F	LA	Epithelioid	Heart murmur, clubbed finger	Resection	Dead
7	Gegenbach et al[21]	1991	ND	ND	LV	Epithelioid	Incidentally discovered on echocardiogram	ND	Alive after 4 months
8	Montes Orbe et al	1991	56	F	PV	Epithelioid	Thromboembolism	Surgical and medical	ND
9	Marchiano et al[22]	1993	71	F	RA	Epithelioid	Palpitation, dizziness, low grade fever	Surgical resection	Alive after surgery
10	Billé-Turc et al[23]	1993	59	M	AV	Epithelioid	Stroke	Surgical	Alive (F-up 1 year)
11	Di Biasi et al[24]	1995	35	M	RV	Epithelioid	Incidentally discovered on echocardiogram	Surgical	Alive (F-up 1 year)
12	Ichikawa et al[25]	1997	32	F	CS	Malignant	Incidentally discovered on echocardiogram	Surgical	Alive
13	Hongquan et al[26]	1998	42	F	RA	ND	Incidentally discovered on echocardiogram	Surgical	Alive
14	Yoshida et al[27]	1999	ND	M	RA	Malignant	Cardiac tamponade	Surgical	Dead
15	Di Biasi et al[24]	1995	35	M	RV	ND	Dyspnoea		
16	Kamiyoshihara M[28]	2001	32	M	ND	Malignant	ND	Surgical Excision	Dead after 41 months
17	Agaimy et al[29]	2002	68	M	RV/TV	Epithelioid	Incidentally discovered antopsy	ND	Dead
18	Kitamura et al[30]	2005	36	F	RA	Epithelioid	Cardiac tamponade	Surgical resection	Alive (F-up 3 years)
19	Vall Bernal et al[31]	2005	69	F	MV	Epithelioid	Edema, exertional dyspnea	Surgical resection	Alive (F-up 6 months)
20	Wang et al[32]	2006	36	F	RA	ND	Dyspnoea	ND	ND
21	Moulai et al[33]	2006	53	M	CS	Epithelioid	Incidentally discovered on echocardiogram	Heart transplant	Alive (F-up 10 years)
22	Lisy et al[34]	2006	61	M	LA	Epithelioid	Incidentally discovered on echocardiogram	Surgical resection	ND
23	Safirstein et al[35]	2007	51	F	RA	Epithelioid	Incidentally discovered on echocardiogram	Surgical resection	Alive (F-up 8 years)
24	Zhang et al[36]	2008	66	F	LA	Epithelioid	Incidentally discovered on echocardiogram	Heart transplantation	Alive
25	Messias et al[37]	2008	21	F	LA	Epithelioid	Chest pain	Surgical resection	Alive
26	Kahlout M et al[38]	2009	44	F	RA	Epithelioid	Cardiac tamponade	ND	ND
27	Guray et al[39]	2010	25	M	RA	ND	Heart failure	Medical	ND
27	Lahon et al[40]	2012	29	F	SVC	Epithelioid	widened superior mediastinum noticed on a routine chest radiograph	Surgical resection	Alive

28	Sugimoto T et al[41]	2013	77	F	RA	Epithelioid	Incidentally discovered on echocardiogram	ND	ND
29	Allain et al[42]	2014	ND	ND	SVC/RA	Epithelioid	ND	ND	ND
30	Ellouze et al[43]	2015	53	M	RA	Epithelioid	Incidentally discovered on echocardiogram	Surgical resection	Alive
31	Gaytán-Cortés et al[44]	2016	23	M	RA	ND	chest pain, intermittent paroxysmal nocturnal dyspnea and malaise	Surgical resection	Alive
32	Lappa et al[45] (article in press)	2017	36	F	RA	Kaposiform	Alveolar Hemorrhage	Surgical and drug supports	Dead

Table 2. Cases of cardiac metastasis in adult patients with non-cardiac primary hemangioendothelioma, reported in scientific literature. M: male, F: female, ND: non defined, MTs: metastasis; RA: right atrium; HE; hemangioendothelioma; IL: interleuchin; CT: chemotherapy; Pt: patient.

Principal Reported Case of metastatic cardiac hemangioendothelioma					
Author	**Age Pt**	**Gender**	**Primary Tumor**	**Complications**	**Therapy**
Myauchi et al (1987)[46]	ND	F	Ovarian Hemangiomatosis	MTS pericardium and RA	Medical
Dubois et al (1987)[47]	64	F	hepato-splenoganglionic hemangioendothelioma	MTs at the heart → heart failure	Surgical and medical
Hurley et al (1994)[48]	ND	F	Intracranial epithelioid HE	Seizures, Liver and heart multifocal locliation	Aggressive surgical treatment
Bhutto et al (1995)[49]	ND	M	hepatic epithelioid hemangioendothelioma	Heart MTs	irradiation, IL-2 injection, and/or surgery
Bellmunt et al (1999)[50]	48	F	hepatic epithelioid hemangioendothelioma	Heart metastasis Portal hypertension	CT → adriamycin
Ilasi et al (1999)[51]	ND	F	HE of the bone	Heart failure MTs in RA	Chemotherapy
Hsu CY (2014)[52]	73	F	hepatic epithelioid hemangioendothelioma	Heart failure	embolization of the abnormal neoangiogenic vessels

Conclusion

Our literature review would like to be a resume of the cardiac hemangioendothelioma and cardiac involvement in patients with non-cardiac hemangioendothelioma in daily clinical experience. In line with our analysis, the cardiac-HE was vascular aggressive neoplasm with variable histopathological pattern and outcome, principally in female patient and right atrium. Its outcome would be influenced to the early diagnosis and prompt surgical and medical treatments. On the contrary, the cardiac involvement in patients with non-cardiac hemangioendothelioma was weird and life-threatening: only seven cases described and with worsening evolution due to coagulative disorders, heart failure and end-stage multi-organ disease. In case of suspicion of CHE or cardiac involvement of NC-HE were immediately undergone to appropriate diagnostic exams (lab test, echocardiogram, computed tomography, and so on) and rapid surgical and medical therapies in order to avoid the life-threatening evolution of these malignant pathologies.

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Conflicts of interest: All the authors declare non conflict of interest.

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