Letter to the Editor

Giant cavernous hemangioma of the left atrial appendage: a case description and literature review

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Introduction

In 2015, the World Health Organization (WHO) proposed a new histological classification of cardiac tumors. Cardiac tumors are divided into the following: (I) benign tumors and tumor-like conditions, (II) tumors of uncertain biological behavior, (III) germ cell tumors, (IV) malignant tumors, and (V) tumors of the pericardium (1,2). Clinically, primary cardiac tumors (PCTs) are rare entities, and cardiac hemangiomas (CHs) are particularly rare benign vascular lesions, accounting for approximately 5–10% of benign cardiac tumors and less than 2% of all heart neoplasms (3,4). CHs occur as a result of abnormal dilation or hyperplasia of small arteries, veins, or capillaries (5), and are histologically divided into capillary hemangioma, cavernous hemangioma, and arteriovenous malformation (1,2).

In children, the most common site of CHs is the right atrium (RA), whereas in adults, they are mostly located in the left ventricle (LV) and cardiac valves (1). Cavernous hemangioma of the left atrial appendage is especially rare. Herein, we report a case of cavernous hemangioma of the left atrial appendage. We also investigated the recent correlative literature to summarize its clinical characteristics, imaging manifestations, histopathology, and therapeutic methods.

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was provided by the patient for the publication of this case report and the accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

A 60-year-old woman was admitted with a mediastinal mass that was discovered incidentally on 24 December 2019. In the course of the disease, she had no known history of nausea, vomiting, palpitations, or dyspnea. After admission to the hospital, physical examination revealed normal cardiac sounds with no murmur. Laboratory examination results revealed that all the parameters were within the normal reference range. It was noted that no abnormalities

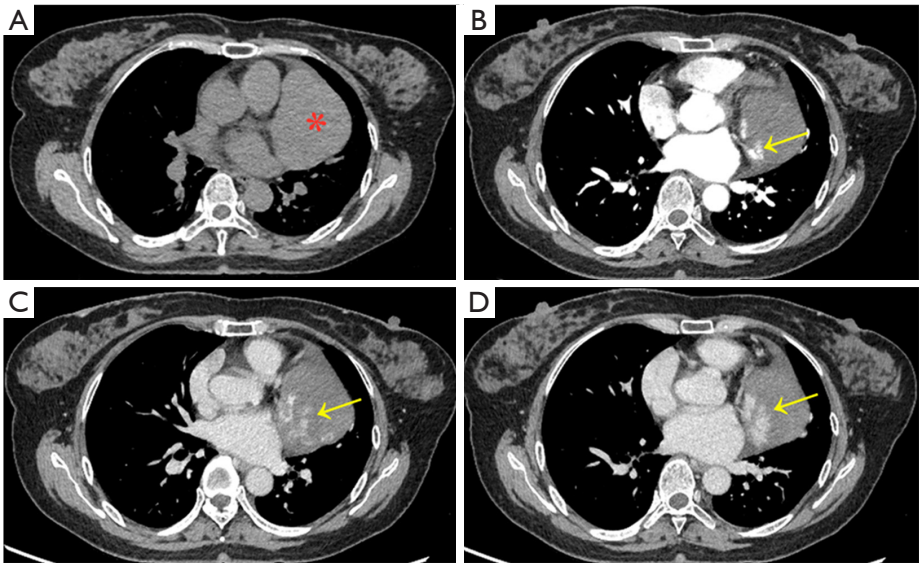

Figure 1 A 60-year-old woman was diagnosed with a cavernous hemangioma of the left atrial appendage. (A) On NECT, the mass is on the left side of the patient's heart (red star). (B-D) At the arterial (B) and venous phases (C,D) of the CECT, it shows a pattern of "progressive" or "prolonged" enhancement (yellow arrows), which are typical findings of a hemangioma. NECT, non-contrast-enhanced computed tomography; CECT, contrast-enhanced computed tomography.

were illustrated on the echocardiogram. Contrast-enhanced computed tomography (CECT) of the chest showed a heterogeneous and well-defined lesion on the left side of the mediastinum measuring 7.0×8.5×6.0 cm (*Figure 1A*). The mass was well demarcated from the left atrium (LA), LV, and pulmonary artery. In addition, this mass showed nodular and patchy enhancement in the arterial phase (*Figure 1B*), and the range of enhancement increased in the venous phase (*Figure 1C*,*1D*). Our radiologists tried to perform cardiac magnetic resonance (CMR) imaging on this patient to gain a comprehensive understanding of the lesion. However, the CMR examination was unsuccessful because of the patient's claustrophobia.

Subsequently, surgical excision of the lesion using a midsternal thoracotomy was scheduled for both definitive diagnosis and therapeutic purposes (*Figure 2A*,*2B*). Histopathological examination of the mass confirmed the diagnosis of cavernous hemangioma originating from the left atrial appendage (*Figure 3*). At 6 months after the surgery, non-contrast-enhanced computed tomography (NECT) examination depicted the absence of the tumor (*Figure 4*). After discharge, the patient was followed up by an echocardiogram, which showed no recurrence of this tumor.

Discussion

Clinical features

CHs are rare benign vascular tumors originating from the heart, whereas myxomas and papillary fibroelastomas are more common in clinical diagnosis. CHs can be found in any cavity of the heart, and neonatal hemangioma commonly occurs in the RA. However, most CHs are in the ventricle, especially in the lateral wall of the LV (6). A CH rooted in the left atrial appendage is particularly rare, and only several cases have been identified in previous studies (*Table 1*) (7-14). As a result, our case can be added to this body of literature.

Patients of all ages can develop CH. Based on the related literature, CH patients are usually approximately 40 years old, with a small male predominance (15). However, our patient was 67 years old and had a left atrial appendage hemangioma. Most affected patients are asymptomatic, but some may show atypical chest pain, abnormal conduction, coronary artery insufficiency, congestive heart failure, pericarditis, pericardial effusion, tamponade, and thromboembolism, among others (3). The patient's clinical presentations can be vastly different due to the location, size, activity, infiltration, or the speed of growth.

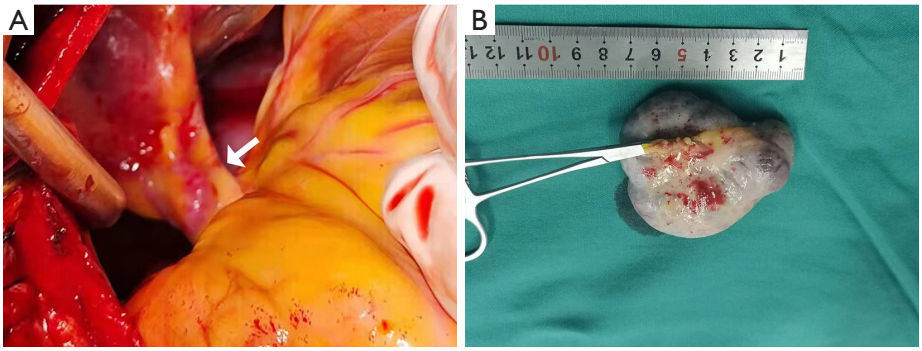

Figure 2 A 60-year-old woman was diagnosed with a cavernous hemangioma of the left atrial appendage. (A) The lesion attached to the left atrial appendage with a pedicle (white arrow). (B) Gross view of the tumor after resection.

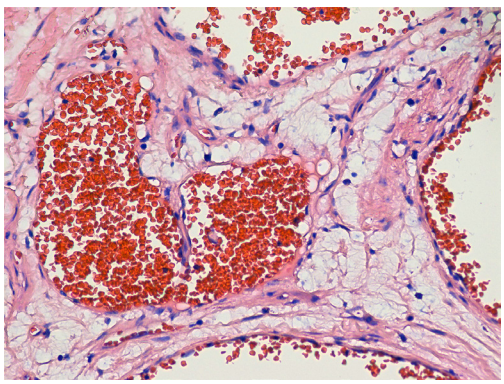

Figure 3 A 60-year-old woman with cavernous hemangioma of the left atrial appendage. Dilated vascular cavities with a large number of red blood cells in the lumen. These cavities have thick walls (HE, ×200). HE, hematoxylin and eosin.

Endocardial hemangiomas often present as soft tissue masses, which may contain mucoid components. They often protrude into the heart cavities and lead to hemodynamic changes, thrombosis, obstruction of the outflow tract, valve regurgitation, arrhythmia, or embolism. Furthermore, CHs may also present as Kasabach-Merritt syndrome, which is characterized by multisystemic hemangiomas, thrombocytopenia, and consumptive clotting disorder (3).

Histologic and pathologic features

Histologically, CHs are classified as capillary (numerous capillaries without red blood cells in the lumen), cavernous (thin-walled dilated capillaries with multiple red blood cells in the lumen), or arteriovenous (composed of thick-walled

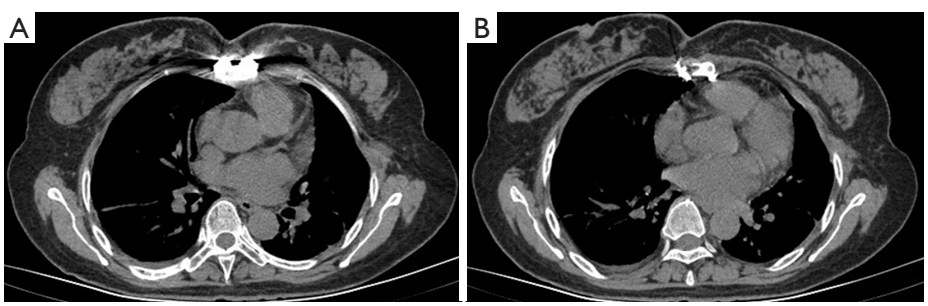

Figure 4 A 60-year-old woman was diagnosed with a cavernous hemangioma of the left atrial appendage. (A,B) Six months after surgery, the NECT examination depicted the absence of the tumor. NECT, non-contrast-enhanced computed tomography.

Table 1 Characteristics of cavernous hemangioma of the left atrial appendage in the published literature

Reference	Year	Author	Number of cases	Gender	Age (years)	Clinical presentation	Imaging method	Morphologic feature	Size (cm)	Pathological type	Therapeutic method	Prognosis
(7)	2009	Chiappini	1	Male	65	Syncope and paroxysmal atrial fibrillation	TTE	Encapsulated, a wild sessile implant basis	5.0×3.0	Cavernous	Median sternotomy	Without complications
(8)	2015	Darwazah	1	Male	62	Cardiac arrest induced by spinal anesthesia	–	A wide base with no stalk	1.0×1.0	Cavernous	Resection operation	–
(9)	2009	Acikel	1	Male	71	Cerebrovascular ischemia	TTE	Left atrial appendage and right atrium	4.0×1.0 2.5×1.0	Cavernous	Resection operation	No recurrence
(10)	2022	Fujita	1	Female	76	Cough, dyspnea, and leg oedema	Thoracoscopic pericardial fenestration	Left atrial appendage	2	Capillary	Antero-axillary thoracotomy	No pericardial or pleural effusions
(11)	2003	Oshima	1	Male	70	Asymptomatic	X-ray	Left appendage and protruding into the pericardial cavity	4.5×3.6	Capillary and cavernous	Resection operation	–
(12)	2021	Suzuki	1	Female	66	Hypertension	X-ray	Left appendage	5	Cavernous	Median sternotomy	Without complications
(13)	2013	Takahashi	1	Male	61	Asymptomatic, presenting exertional dyspnea with enlargement of tumor	X-ray	Left appendage	6.3×5.0	Mainly cavernous type hemangioma with mixed capillary type	Median sternotomy	Without complications
(14)	2004	Sata	1	Male	72	Recurrent pericardial tamponade	TEE	Left appendage and growing into the pericardial cavity	4.5×4.5	A combination of cavernous- and venous-type	Left anterolateral thoracotomy by way of the fourth intercostal space	No pericardial effusion
This case	2022	–	1	Male	60	None	CT	Left appendage, pedicled	6.5×9.0×7.0	Cavernous	Resection operation	Without complications

TTE, transthoracic echocardiography; TEE, transesophageal echocardiography; CT, computed tomography.

dysplastic arteries, venous-like vessels, and capillaries) (3). Capillary hemangiomas and cavernous hemangiomas are adjacent to the endocardium and have a broad base or thin stalk attached to the wall of the heart but do not infiltrate the myocardium. Moreover, most CHs are located in the myocardium and bear some similarities to the intermuscular hemangiomas of skeletal muscle.

Radiologic features

Echocardiography is the first option for imaging examination because it can be used to demonstrate the location and size of the lesions, and enables a qualitative diagnosis. However, computed tomography (CT) and magnetic resonance imaging (MRI) are superior methods for illustrating the invasion of the lesions. The echocardiographic findings of CHs are hyperechoic masses or nodules. CT often exhibits uneven and low-density lesions with lower density (cystic lesions) and high density (small and patchy calcification) in the tumor. On CECT, CHs display uneven and avid enhancement. Cavernous hemangiomas often show delayed enhancement with gradual filling from the edge to the center, which is similar to cavernous hemangiomas of the liver. On CMR, hemangiomas manifest as an equal signal on T1-weighted imaging (T1WI) and a high signal on T2-weighted imaging (T2WI) due to the slow blood flow. Over the course of echo, the signal increases on T2WI, which is known as the "bulb sign". The manifestation on contrast-enhanced MRI is similar to that on CECT (16). CMR also provides excellent contrast resolution and multiplanar capability, facilitating qualitative diagnosis and optimal anatomical evaluation of CHs. This anatomical information is especially useful for pre-surgical planning.

Differential diagnosis

Myxomas are the most frequent PCT, accounting for more than 50% of all cases. Papillary fibroelastomas represent the most common valvular tumor. Cardiac rhabdomyomas constitute up to 90% of cardiac tumors in infants and children aged less than 12 months. As a result, a careful differential diagnosis should be performed when providing a preliminary diagnosis of CH.

Myxomas are the most common benign cardiac tumor, accounting for almost half of all PCTs (3,6). Approximately three-quarters of all myxomas arise in the LA, with 23% occurring in the RA, and they often have a narrow pedicle attached to the oval fossa of the atrial septum (3,6). On echocardiography, the lesion appears as a hypoechoic or hyperechoic mobile mass. On CT, the lesion is heterogeneous with areas of high density (calcification) and/or low density (cystic degeneration and necrosis). On CMR, myxomas are typically demonstrated as an equal signal or heterogeneous intensity on T1WI and a high signal or heterogeneous intensity on T2WI. Sometimes, myxomas also show areas of decreased signal, which is attributable to calcification or hemosiderin (3,6). There is an uneven and slight enhancement on gadolinium enhancement.

Papillary fibroelastomas represent the most common valvular tumor (approximately 75% of all cardiac valvular tumors). They are also the second most common benign cardiac tumor (accounting for 16% of all patients) (3,6). A papillary fibroelastoma typically originates from the free edge of the valve, especially in the aortic valve, and is usually medially echoic on echocardiography but is not visible on CT. It presents as isointense on T1WI and T2WI images, with significant enhancement on gadolinium enhancement.

Rhabdomyomas are the most common PCT in children, accounting for 40–60% of cases (3,6). A rhabdomyoma often manifests as multiple masses in the myocardium of the LV or the interventricular septum, and hemorrhage and calcification are rarely seen. On echocardiography, a rhabdomyoma is a hyperechoic mass but is indistinguishable on CT, as the density of the mass is similar to that of the normal myocardium. Rhabdomyomas are isointense or slightly hyperintense on T1WI and hyperintense on T2WI images, and there is no significant enhancement on gadolinium enhancement.

CHs should also be differentiated from primary cardiac sarcomas, which account for most malignant PCTs (about 30%), and are the second most common PCT (about 10%) (3,6). A detailed description of some PCTs is summarized in *Table 2*.

Treatment and prognosis

Although a CH is a benign tumor, surgery should be performed as soon as possible due to the risk of developing angiosarcoma and the possibility that asymptomatic CHs may suddenly cause life-threatening complications. If a cardiac tumor is established, the patient should be treated at an interdisciplinary center. Surgeons, oncologists, radiotherapists, and radiologists need to cooperate comprehensively. On the one hand, surgery is beneficial for diagnosis; on the other hand, it can reduce the risk

Table 2 Clinical and imaging features of hemangioma, common benign, or malignant cardiac tumors

Cardiac masses	Benign or malignant	Percentage (%)	Susceptible age	Site	Morphological variation	US	CT	CMR			
								Cine-MR imaging	T1WI	T2WI	LGE
Hemangioma	Benign	5–10	All ages	Anywhere in cardiac or pericardium (especially the ventricle)	Polypoid or pedicled, endocardial or intramyocardial	Hyperechoic	Heterogeneous with calcification or cystic lesions	Hyperintense with focal ventricular wall motion abnormality	Iso- or slightly hyperintense	Hyperintense	Heterogeneous prolonged enhancement
Myxoma	Benign	50	30–60 years	Fossa ovale of atrial septum (left atrium 75%, right atrium 23%)	Calcification, cystic degeneration or necrosis	Hypoechoic or hyperechoic mobile mass	Heterogeneous with calcification	Mobile mass	Isointense, heterogeneous	Hyperintense, heterogeneous	Heterogeneous enhancement
Papillary fibroelastoma	Benign	16	60–80 years	The free edge of the valve, especially the aortic valve	Small with narrow stalk, calcification rare	Medial echo	Usually not visible	Hyperintense and movable valve mass with thin stalk	Isointense	Isointense	Significant enhancement
Rhabdomyoma	Benign	1	Infants and children	Ventricle or the interventricular septum	Lobulated nodules and multiple	Hyperechoic	Isodensity	Slightly hyperintense, intramural mass	Isointense or slightly hyperintense	Hyperintense	None
Angiosarcoma	Malignant	30	Middle-aged men	Almost in the right atrium	Circumscribed or diffusely infiltrative, hemorrhage and necrosis	Protruding into right atrium, hemorrhagic effusion	Heterogeneous	Heterogeneous, hemorrhage and necrosis	Heterogeneous hyperintense “cauliflower”	Heterogeneous hyperintense “cauliflower”	Avid enhancement with “Sunray” aspect
Synovial sarcoma	Malignant	10–15	Approximately 34.8 years, M:F =3.4:1	Pericardium, right atrium	Irregular and lobulated	Isoechoic	Low density	Heterogeneous mass with poly cysts or septa	Hypointense or isointense	Hyperintense mainly, heterogeneous	Heterogeneous enhancement
Rhabdomyosarcoma	Malignant	10	Infants, children, M:F =1:1.4	Anywhere in the hear	Smooth or irregular contour, central necrosis	Multiple, valves and pericardium invasion	Low density	Isointense	Isointense	Isointense	Heterogeneous enhancement

US, ultrasonography; CT, computed tomography; CMR, cardiac magnetic resonance; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging; LGE, late gadolinium enhancement.

of rupture or pulmonary embolism (17). Regarding CHs with a clear boundary, radical surgical resection should be performed as far as possible, regardless of tumor size. Patients generally recover well after surgery. A large, extensive CH is potentially at risk of recurrence and often has an unfavorable prognosis because it cannot be completely resected. All patients should be followed up regularly after surgery to determine whether there are new or recurrent lesions.

Conclusions

Hemangioma of the left atrial appendage is rarely encountered clinically. However, comprehensive knowledge of this condition may be helpful in the formulation of a diagnosis and for differential diagnoses.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-460/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was provided by the patient for publication of this case report and the accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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