Clinical Case Report

Lymphocyte-rich capillary–cavernous hemangioma of the mitral valve: a case report and review of the literature

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A B S T R A C T

Valvular hemangioma incidence is extremely low. In this report, we describe a 62-year-old man who presented with mild edema of the lower limbs. An echocardiogram revealed an incidental 1.3-cm diameter mass on the anterior mitral valve leaflet for which he underwent surgical resection and mitral valve replacement. Histopathological examination showed a lymphocyte-rich capillary–cavernous hemangioma. The exuberant lymphoid stroma is unusual for hemangioma and represents an undescribed pattern of cardiac hemangioma. Including the present report, only 13 cases of mitral valve hemangioma have been reported to date. Most patients are adult. Mitral hemangioma originates in the atrial aspect of the valve and involves more commonly the anterior leaflet. The average maximum diameter of the lesion is 1.7 (S.D.=0.75) cm. Pure cavernous hemangioma is the predominant type of mitral hemangioma. Most of them are described as pedunculated or polypoid. Surgical excision appears to be curative. Recurrences have not been reported. Lymphocyte-rich cardiac hemangioma represents a peculiar type of hemangioma which should be included in the differential diagnosis of other vascular lesions.

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1. Introduction

Cardiac hemangiomas are very uncommon benign vascular tumors representing only 1.5% (range=0–2%) of primary tumors of the heart [1]. Steger et al. [2] observed only one case of hemangioma out of 78 patients that underwent tumorectomy of primary cardiac tumors in a period of 41 years. Probably because cardiac valves are structures poorly vascularized, this location is the least frequent of the hemangioma [3,4]. To the best of our knowledge, only 12 mitral valve hemangiomas have been reported to date in the literature [3,5–15].

In this report, we present an incidental capillary–cavernous hemangioma located in the mitral valve. The tumor showed prominent infiltration of mature lymphocytes in the interstitium. The unique nature of this lesion which is different from other cardiac hemangiomas prompted us to publish this case report.

2. Case report

A 62-year-old Caucasian man undergoing periodic control for arterial hypertension presented with mild edema of the lower limbs for a month. He was in treatment with olmesartan–amlodipine, atenolol, and torasemide. The electrocardiogram showed sinus bradycardia at 54 beats per minute and hemiblock of the anterior subdivision of the His bundle with negative T waves in III and aVF leads. An echocardiogram revealed an incidental, 1.3-cm diameter, noncalcified mass on the anterior mitral valve leaflet with echolucent areas (Fig. 1). This mass was rounded, pedunculated with a short pedicle; it had a smooth surface and was mobile.

Past medical history was positive for diabetes mellitus Type 2 for 3 years, dyslipidemia, and chronic renal failure. He was a smoker of 10 cigarettes a day.

Laboratory tests including routine hematology and coagulation were all within the normal range. Biochemistry study showed glucose 114 mg/dl, urea 55 mg/dl, creatinine 1.49 mg/dl, and estimated glomerular filtration 50 ml/min/1.

A clinical diagnosis of papillary fibroelastoma was established, and cardiac surgery was indicated.

Intraoperatively, the pedunculated mass had a wide implantation base located between A2 and A3 scallops with slight invasion of the mitral annulus in zone A3. A mild mitral insufficiency was noticed. After resection of the mass, repair of the mitral was performed with ring and...
tendinous cord. However, after leaving the pump, a severe leak was ob-
served in the periannular zone. Extracorporeal circulation was resumed,
and the mitral was replaced by a standard CarboMedix prosthesis num-
ber 29. The operation lasted 163 min with 145 min of cardiac ischemia.

The postoperative course was uneventful with stable function. After the
intervention, there was an improvement in renal function (creatinine
1.27 mg/dl, glomerular filtration 60.2 ml/min). The electrocardiogram
showed sinus rhythm at 80 beats per minute, persistence of His bundle
hemiblock, and T negative in III and aVF leads. No significant mitral re-
gurgitation was detected. The mean prosthetic gradient was 8.0
mmHg. Two months after surgery, the patient remained well and
asymptomatic with no signs of tumor recurrence.

A segment of mitral valve measuring 1.7×1.6×0.3 cm including a
purplish mass was received in the pathology service. Grossly, the pur-
plish and solid excised mass measured 1.6×1.1×1 cm. The surface was
smooth. The well-delimited mass was attached to the surface of the
valve by a short pedicle (Fig. 2). On cross-section, the lesion was spongy
with focal hemorrhagic areas. Histopathological examination revealed
that the mass was a vascular lesion with a dense lymphoid infiltrate
(Fig. 3A). It was composed of small thin-walled vessels with rounded
or oval contours lined with flattened typical endothelial cells (Fig. 3B).
Areas of irregularly dilated and congested, blood-filled, thin-walled,
gaping blood vessels were more commonly seen in the center of the
lesion. Hypertrophic capillary vessels with virtual lumina forming
an anastomosing network were prominent (Fig. 3C). Occasional mitoses
were seen in some poorly canalized vessels in the deep of the lesion.
Atypia or necrosis was absent. The loose stroma showed a dense diffuse
infiltration of inflammatory cells most of which were lymphocytes.
Plasma cells, groups of hemosiderophages, and interspersed red blood
cells were the other cells present (Fig. 3D). Capillary and cavernous
components were balanced (Fig. 4). The lymphocytes were mature
(Fig. 5A). Some vessels showed abundant lymphocytes in their lumina
(Fig. 5B).

On immunohistochemical staining, there was a mixture of CD3+ T-
cells (Fig. 5C) and CD20+ B-lymphocytes (Fig. 5D) in the interstitium
and in the vessels. However, T-cells were predominant in the intersti-
tium and in vascular lumens (Fig. 5C and D). The endothelial cells
were positive for CD31 (Fig. 6A) and CD34 (Fig. 6B). Alpha-smooth muscle
actin-positive pericytes were seen around many individual vascular
channels (Fig. 6C). Pathologic diagnosis was lymphocyte-rich capillary-
cavernous hemangioma.

In the rest of the valve, some Lambí’s excrescences were observed
(Fig. 6D).

3. Discussion

Cardiac hemangiomas are classified as capillary, cavernous, mixed
capillary and cavernous, and arteriovenous hemangiomas [16]. Epitheli-
oid hemangioma and lobular capillary hemangioma (pyogenic granulo-
ma) are separate and distinct entities.

Detection of valvular hemangiomas is infrequent. Including the
present report, only 13 cases of mitral valve hemangioma have been re-
ported to date [3,5–15] (Table 1). Most patients are adult (mean age=42.7; S.D.=19.8 years; range=17–76 years), and only three cases of
children have been reported (mean age=5.7; S.D.=4.0 years; range=
1–8 years). No clear sex difference has been observed. The presenting
symptoms may include palpitations, exertional dyspnea, syncope, angina
or atypical chest pain, heart failure due to hemodynamic disturb-
ances, suspicion of infective endocarditis, and sudden death. Two-

Fig. 1. Echocardiogram showing a pedunculated, noncalcified mass (arrow) on the anterior mitral valve leaflet with echolucent areas. The pedicle was short.

Fig. 2. Macroscopic aspect of the purplish, rounded, pedunculate mass and the resected
segment of the mitral valve. The arrow indicates the site of implantation of the mass.
Dimensional echocardiography is the most appropriate imaging modality for the detection of this tumor. Coronary angiography may delineate feeding vessels to the tumor and reveal a characteristic tumor blush [13]. Computed tomography scan or magnetic resonance imaging helps to evaluate the dimensions and invasiveness of the lesion. Nevertheless, surgical resection and histopathological examination remain the “gold standard” diagnosis of choice.

Mitral hemangioma originates in the atrial aspect of the valve and involves more commonly (66.7%) the anterior leaflet. The average of maximum diameter of the lesion is 1.7 (S.D.=0.75) cm (range=0.8–3 cm). Pure cavernous hemangioma is the most common type (58.3%) of mitral hemangioma. Most of them (72.7%) are described as pedunculated or polypoid. A cystic or partially cystic appearance is observed in 18.2% of cases. Surgical excision of mitral hemangiomas appears to be curative and is the treatment of choice. Mean follow-up of the cases was 9.3; S.D.=5.6 months (range=3–19 months). No recurrences have been described.

Diffuse neutrophilic infiltration in a valvular hemangioma has been reported in a case with superimposed infective endocarditis [6].

The case herein reported was a benign lesion based on absence of cellular atypia, high mitotic count, or necrosis and the presence of alpha-smooth actin-positive pericytes around individual vascular channels. Mononuclear inflammation can be seen in cardiac hemangiomas [17,18] and may account for the response to antiinflammatory treatment or the spontaneous resolution in some lesions [4,19]. However, an exuberant diffuse infiltration of mature lymphocytes as seen in our case has not been reported in this location. A review of the literature showed only one case of extracardiac hemangioma showing prominent lymphoid infiltration of the interstitium [20]. On the other hand, Kerstetter et al. [21] reported a unique case of soft tissue hemangioendothelioma with prominent lymphoid infiltrate which represented a diagnostic challenge.
These cysts rarely persist in adults. In these cases, the cysts are larger dental postmortem.

Blood cysts of the atrioventricular valves are most frequently an incidental finding. Endothelial cells show intense and diffuse positivity for CD31 (>200) (A) and CD34 (>200) (B). Endothelial cells are frequently surrounded by a layer of pericytes showing positivity for alpha-smooth muscle actin (C). Lamb's excrescences showing no branching, finger-like projections extending from the valve surface (H&E, ×100) (D).

Closely similar to the thinking of Kerstetter et al. [21], we believe that the dense lymphoid stroma is exceptional for hemangioma and suggests that it may represent some type of homing response rather than simply inflammation. The homing or tissue-specific selective and microenvironment-specific lymphocyte vascular adhesion is possibly mediated by chemokines or chemokine receptors. These factors allow the coordination of lymphocyte traffic.

Differential diagnosis of mitral hemangioma includes blood cyst, varix, intravascular histiocytosis of the valve, and organized thrombus. Blood cysts of the atrioventricular valves are most frequently an incidental postmortem finding of fetus and infants<6 months of age. These cysts rarely persist in adults. In these cases, the cysts are larger and may cause cardiac dysfunction [22,23]. Histopathologically, they are unilocular and consist of a thin wall of connective tissue lined with flat endothelial cells by both sides. The lumen contains a bloody fluid [23]. Varix of the heart is an endocardial, unilocular, blood-filled cyst lined by endothelial cells. The wall usually shows collagenization. The lumen contains organizing thrombi and may show phleboliths. The lesion is usually found in the inferior rim of the fossa ovalis in right atrium and corresponds to a dilated vein [24,25]. Intravascular histiocytosis has been described in the aortic valve [26]. The condition is characterized by dilated thin-walled vessels, some of them containing collections of mononuclear histiocytes within their lumina. The lesion can be associated with a systemic process. Thrombosis may occur in the native mitral valve and simulate a neoplasm [27,28]. Thus, an organizing valvular thrombus may show abundant small vessels mimicking a vascular tumor (hemangioma-like appearance). However, persistent nonorganized thrombus zones, residual granulation tissue, and focal areas of intravascular papillary endothelial hyperplasia are usually diagnostic keys.

4. Conclusion

In sum, the exuberant lymphoid stroma is unusual for hemangioma and represents an undescribed pattern of cardiac hemangioma. We believe that the designation of lymphocyte-rich cardiac hemangioma in this case is justified. The lesion represents a peculiar type of hemangioma which could be included in the differential diagnosis of other vascular lesions.

References


Table 1
Hemangioma of the mitral valve: review of the literature

<table>
<thead>
<tr>
<th>Case/Reference</th>
<th>Age (year)/Sex</th>
<th>Presenting ailment</th>
<th>Leaflet</th>
<th>Maximum diameter (cm)</th>
<th>Pathology</th>
<th>Follow-up (months)/Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/[3]</td>
<td>–</td>
<td>Sudden death</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>2/[5]</td>
<td>33/F</td>
<td>Palpitations, chest tightness</td>
<td>Anterior</td>
<td>1.0</td>
<td>Cavernous</td>
<td>12/No recurrence</td>
</tr>
<tr>
<td>3/[6]</td>
<td>24/F</td>
<td>Facial asymmetry, left arm weakness, fever</td>
<td>Anterior</td>
<td>–</td>
<td>Pedunculated capillary with dense infiltration of neutrophils (infective endocarditis)</td>
<td>–</td>
</tr>
<tr>
<td>4/[7]</td>
<td>8/M</td>
<td>Palpitations, dyspnea</td>
<td>Anterior</td>
<td>3.0</td>
<td>Polypoid cavernous</td>
<td>19/No recurrence</td>
</tr>
<tr>
<td>5/[8]</td>
<td>8/M</td>
<td>Palpitations, dyspnea</td>
<td>Anterior</td>
<td>3.0</td>
<td>Pedunculated cavernous</td>
<td>6/No recurrence</td>
</tr>
<tr>
<td>6/[9]</td>
<td>76/M</td>
<td>–</td>
<td>Posterior</td>
<td>1.5</td>
<td>Polypoid–cavernous</td>
<td>–</td>
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<td>7/[10]</td>
<td>33/F</td>
<td>Shortness of breath, chest pressure, headness</td>
<td>Anterior</td>
<td>2.0</td>
<td>Cystic cavernous</td>
<td>6/No recurrence</td>
</tr>
<tr>
<td>8/[11]</td>
<td>–</td>
<td>–</td>
<td>Posterior</td>
<td>2.0</td>
<td>–</td>
<td>Not specified/No recurrence</td>
</tr>
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<td>9/[12]</td>
<td>17/M</td>
<td>Syncope event</td>
<td>Anterior</td>
<td>1.4</td>
<td>Pedunculated capillary</td>
<td>12/No recurrence</td>
</tr>
<tr>
<td>10/[13]</td>
<td>49/F</td>
<td>Dyspnea</td>
<td>Posterior</td>
<td>0.9</td>
<td>Pedunculated cavernous</td>
<td>8/No recurrence</td>
</tr>
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<td>11/[14]</td>
<td>48/M</td>
<td>Unstable angina</td>
<td>Posterior</td>
<td>2.0</td>
<td>Pedunculated cavernous</td>
<td>–</td>
</tr>
<tr>
<td>12/[15]</td>
<td>1/F</td>
<td>Dyspnea</td>
<td>Anterior</td>
<td>0.8</td>
<td>Partially cystic cavernous</td>
<td>–</td>
</tr>
<tr>
<td>13/Report</td>
<td>62/M</td>
<td>Mild edema in lower extremities</td>
<td>Anterior</td>
<td>1.6</td>
<td>Pedunculated capillary–cavernous. Dense, diffuse infiltration of lymphocytes</td>
<td>2/No recurrence</td>
</tr>
</tbody>
</table>


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