A Case of Giant Coronary Artery Aneurysm with Fistulous Connection to the Pulmonary Artery: a Case Report and Review of the Literature

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Abstract

Giant coronary artery aneurysm is a rare condition with a reported prevalence of 0.02%. Herein, we report the case of a 79-year-old woman with a giant coronary aneurysm arising from a branch of the left anterior descending coronary artery that had a fistulous connection to the pulmonary artery. The aneurysm was removed and inflow and outflow arteries were closed surgically. Histology showed prominent mucinous degeneration and infiltration of inflammatory cells in the medial layer. After successful surgery, the patient was discharged uneventfully.

Key words: giant coronary artery aneurysm, fistula, shunting, heart failure

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Introduction

Localized dilatation of the coronary artery exceeding the diameter of the adjacent normal segment by 50%, termed a coronary artery aneurysm, is not uncommon, but it may worsen outcomes (1). In adults, most coronary artery aneurysms are atherosclerotic in origin, and other underlying causes include Kawasaki disease, Takayasu arteritis (2), collagen vascular disease (3), and IgG4-related coronary periarteritis (4); in addition, coronary artery aneurysm may develop after angioplasty, especially after drug-eluting stent implantation (5). “Giant” coronary artery aneurysms, which are defined as having a diameter of 2 cm or greater, are rarer with a reported prevalence of 0.02% (6). Although evidence-based treatment guidelines are limited, surgical resection may be selected due to potential life-threatening complications, such as rupture, thrombosis, and distal coronary embolization. A coronary artery fistula is also a rare anomaly with an incidence ranging from 0.1% to 0.8% in the adult population (7). In this case report, we provide data from a patient who had heart failure symptoms and was diagnosed to have a giant coronary artery aneurysm with a fistulous connection to the pulmonary artery.

Case Report

A 79-year-old woman was referred to our hospital due to dyspnea on exertion and edema in the lower extremities. Her medical history was unremarkable from the cardiologic point of view. Initial vital signs showed a body temperature of 36.8 degrees, blood pressure of 140/60 mmHg, heart rate of 112 beats/min, and tachypnea of 28/min. A continuous Levine grade 3/6 heart murmur was heard at the second right sternal border. The patient did not have apparent previous illness suggestive of Kawasaki disease. Chest X-ray revealed mild dilatation of the left second bow and electrocardiography revealed atrial fibrillation. Laboratory studies showed a white blood cell (WBC) count of 7,680/μL; hemoglobin level of 12.2 g/dL, platelet count of 23.8×10¹⁴/μL, C-reactive protein concentration of 5.76 mg/dL, and serum procalcitonin level of 0.05 ng/mL. Antinuclear antibody was negative, MPO-ANCA and PR3-ANCA were both <10 EU, and the serum IgG4 level was 11.9 mg/dL (within normal
The serum N-terminal pro-B-type natriuretic peptide level was increased to 3,243 pg/mL. Arterial blood gas analysis evaluated under room air showed partial pressures of oxygen and carbon dioxide of 91.8 mmHg and 37.4 mmHg, respectively. The 2-D transthoracic echocardiography showed a mild pericardial effusion and a large cystic mass containing smoke-like echoes adjacent to the left anterior descending artery (LAD). Blood flow into the aneurysm could be observed as color Doppler signals (B). Axial-section of CT image. Coronary artery aneurysm (dimensions 45×46 mm) with heterogenic contrast enhancement is seen (arrows). D, E. 3-Dimensional heart reconstruction of CT images. A giant coronary artery aneurysm (47×49×59 mm) was present at the branch artery of the left anterior descending artery (LAD) (D, E, arrows), and the proximal portion of LAD was dilated and tortuous (D, arrowheads). Several smaller-sized aneurysms were observed at the proximal portion of the right coronary artery, some of which showed a fistulous connection to the pulmonary artery (E, arrowheads). F. Coronary angiography. Left coronary artery angiography demonstrated a giant coronary aneurysm originating from branch of LAD.

Under cardiopulmonary bypass via median sternotomy, the giant aneurysm located at coronary artery branching from LAD (Fig. 2A) was resected and the proximal and distal openings were closed with pledges (Fig. 2B). Pericardial effusion was turned out not to be bloody. No intraneurysmal thrombus was grossly visible. Coronary artery bypass graft surgery was not performed. Main pulmonary artery was opened longitudinally and the fistula was also closed directly. Histopathology of the excised aneurysm showed lymphocytic infiltration, widespread myxoid degeneration, and fragmented elastic fibers in the medial layer (Fig. 3). IgG4 staining depicted no IgG4-positive plasma cell infiltration (data not shown). The symptoms of heart failure of the patient were ameliorated from NYHA functional class III preoperatively to class I post-operatively. The patient was discharged 2 weeks after surgery on anticoagulants and digoxin, and was asymptomatic 2 months after surgery.
We report a patient who was referred to our hospital due to heart failure symptoms, and who was eventually found to have a giant coronary aneurysm originating from LAD with a fistulous connection to the pulmonary artery. Although the shunt flow was modest, with the Qp/Qs ratio of 1.5, surgical
treatment was selected because of the presence of giant coronary aneurysm and heart failure symptom. Surgical removal of the aneurysm and closure of the proximal opening by epicapedal patch was successfully performed, which led to amelioration of the patient’s symptoms.

For giant coronary aneurysm, surgical treatment may be indicated to prevent spontaneous rupture, dissection, myocardial ischemia, or thromboembolic events, although the optimal management strategy has not been established due to rarity of this disorder (8). Coronary artery fistula is also a rare condition that may drain into the right heart chamber or into the pulmonary artery, the prevalence of the latter of which has been reported to be 17% (9); in addition, only a few patients with both giant coronary artery aneurysm and coronary artery fistula have been reported to date (10-12).

Dyspnea on exertion is one of the commonest symptoms of patients with coronary artery fistulas (13), although coronary artery fistulas are usually asymptomatic (14). Rittenhouse et al. reported that surgical correction may be considered for coronary artery fistula when the Qp/Qs ratio is 2.0 or greater (15). When the shunt flow is lower, as in the present case, indications for operation are not precise; however, operation may be considered when myocardial ischemia and/or heart failure are evoked by coronary flow steal and/or shunting (16, 17). Although optimal management strategy has not been established for giant coronary aneu-

Table 1. Cases of Giant Coronary Artery Aneurysms Reported in 2011

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Sex</th>
<th>Coronary artery</th>
<th>Fistula</th>
<th>Drain</th>
<th>Presentation</th>
<th>3DCT</th>
<th>Management</th>
<th>Author</th>
<th>Reference</th>
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<td>1</td>
<td>57</td>
<td>M</td>
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<td>Jung et al.</td>
<td>(20)</td>
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<td>2</td>
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<td>-</td>
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<td>Nazareth et al.</td>
<td>(21)</td>
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<td>-</td>
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<td>Rudan et al.</td>
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<td>4</td>
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<td>5</td>
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<td>F</td>
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<td>surgical</td>
<td>Mita et al.</td>
<td>(23)</td>
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<td>surgical</td>
<td>Berdajs et al.</td>
<td>(24)</td>
</tr>
<tr>
<td>8</td>
<td>N/D</td>
<td>M</td>
<td>RCA</td>
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<td>yes</td>
<td>coil embolization</td>
<td>Urban et al.</td>
<td>(25)</td>
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<tr>
<td>9</td>
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<td>Greenhouse et al.</td>
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<td>Rios Gomez et al.</td>
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<td>Kobayashi et al.</td>
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<tr>
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<td>Pala et al.</td>
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<tr>
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<td>Dhakam et al.</td>
<td>(30)</td>
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<td></td>
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<td>surgical</td>
<td>Mora et al.</td>
<td>(31)</td>
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<tr>
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<td>Liu et al.</td>
<td>(32)</td>
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<td>(33)</td>
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<td>Emaminia et al.</td>
<td>(34)</td>
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<tr>
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<td>-</td>
<td></td>
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<td>Emaminia et al.</td>
<td>(34)</td>
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<tr>
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<td>surgical</td>
<td>Radwan et al.</td>
<td>(8)</td>
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<td>surgical, stent</td>
<td>Kopjar et al.</td>
<td>(36)</td>
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<td>(37)</td>
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<td>Voth et al.</td>
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<tr>
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<td>surgical</td>
<td>Cebeci et al.</td>
<td>(39)</td>
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</table>

N/D indicates not described. 3DCT is said to be yes when the 3D reconstructed CT image was shown in the corresponding paper. CS: coronary sinus, LV: left ventricle, PA: pulmonary artery. Stent indicates covered stent implantation.

Figure 4. Number of publications discussing giant coronary aneurysm according to the year of publication. The papers describing giant coronary aneurysm, irrespective of concomitant fistula formation, were searched using PubMed (http://www.ncbi.nlm.nih.gov/pubmed/) by entering the following key words of the title as search terms: “giant”, “coronary”, and “aneurysm” or “aneurysms”.

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A giant coronary artery aneurysm is considered to be a very rare pathological condition with a reported prevalence of 0.02% (6); however, detection of such lesions seems to have become more frequent owing to the advancement of imaging modalities, especially CT. In fact, the number of reports describing giant coronary aneurysm has recently been increasing, and it was found that more than 60% of such papers have been published from 2006 onward (Fig. 4). In Table 1, we list a summary of the case reports that were published in 2011 in which giant coronary aneurysm was described, irrespective of the presence or absence of fistula formation (Table 1). In these studies, male prevalence was 75% and the association with fistula was observed in 6 (25%) of 24 coronary aneurysms. Notably, 3-D reconstruction of CT images was demonstrated in 16 (67%) of 24 lesions, supporting that dissemination of multidetector CT may facilitate the discovery of giant aneurysm, which might be asymptomatic in initial presentation.

In summary, we have documented a patient with a giant coronary aneurysm with a fistulous connection to the pulmonary artery, who was successfully treated by aneurysmal resection and patch closure of the inflow and outflow arteries. Due to the rarity of such a condition, selection of appropriate therapeutic strategies and investigation of underlying conditions should carefully be performed.

The authors state that they have no Conflict of Interest (COI).

Hideaki Morita and Hideki Ozawa equally contributed to this work.

References


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