



Giant Coronary Aneurysm with Coronary-Pulmonary Artery Fistula in a Jehovah's Witness

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Abstract

With an incidence of 3 in 100 million, giant coronary artery aneurysm (CAA) with coronary artery fistula (CAF) is a very rare condition. To prevent rupture, giant CAA with CAF should be swiftly treated. We present a Jehovah's Witness patient with giant CAA and coronary-pulmonary artery fistula. We resected the giant CAA in one piece, while ligating the CAF, without allogeneic blood transfusion. Due to rarity of these conditions, many thoracic surgeons lack direct experience in its surgical procedures. Herein, we share footage of this surgery as an example of how to safely resect CAA with minimal bleeding.

Keywords

- ▶ giant coronary artery aneurysm
- ▶ coronary-pulmonary artery fistula
- ▶ Jehovah's Witness

Introduction

Coronary artery aneurysms (CAA) and fistulas (CAF) have both become increasingly common diagnoses with the widespread use of coronary computed tomography and coronary angiography testing. CAA with maximum short axis diameters larger than 40 mm is categorized as giant, and giant CAA coupled with CAF is a very rare entity to come across. CAAs are associated with obstructive coronary artery disease, and rupture can result in sudden cardiac death.^{1,2} CAFs with significant left to right shunts can present with heart failure and angina symptoms, and elevate risk of infectious endocarditis.^{3–5} Although CAF is rarely indication for surgery on its own, when coupled with giant CAA, both should be surgically treated to prevent rupture.

Here we present a Jehovah's Witness patient who was incidentally diagnosed with giant CAA with coronary-pulmonary artery fistula. In regard to transfusion, the patient was only open to cardiopulmonary bypass and intraoperative cell salvage. We successfully resected the giant CAA

in one piece, while identifying and ligating its inflow orifice and surrounding CAF in an on-pump arrest procedure, with minimal bleeding and only intraoperative autologous blood transfusion.

Because this condition is so rare, many thoracic surgeons lack direct experience in its surgical procedures. Sharing footage of this surgery as an example of how to safely resect giant CAA and ligate CAF with minimal bleeding could aid as reference for future cases.

Case Report

A septuagenarian Jehovah's Witness female was referred to our hospital for surgical treatment of giant CAA with CAF. The patient was asymptomatic, and the giant CAA was noted as a mass shadow in her chest X-ray (▶**Fig. 1**). Transthoracic echocardiogram showed CAF with a continuous flow of 80 cm/s and mosaic pattern in the pulmonary artery, and a partially thrombosed giant CAA. Coronary computed tomography revealed the CAF to originate from branches of the right

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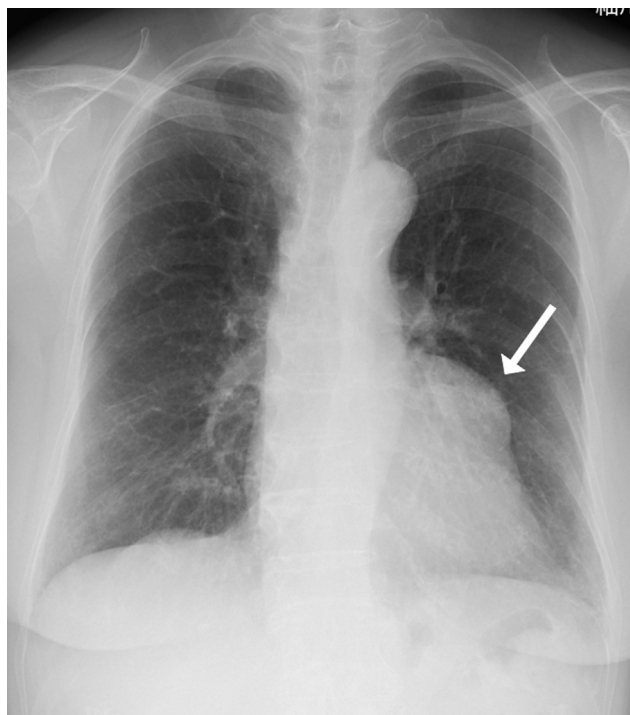


Fig. 1 Giant coronary artery aneurysm noted as mass shadow in chest X-ray (arrow).

coronary artery and left anterior descending artery, entering the pulmonary artery, and a giant CAA with a maximum short axis diameter of 50 mm originating from the CAF (→ Fig. 2). Although the patient did not present with cardiogenic symptoms, she eagerly consented to elective surgery. As a Jehovah's Witness, the patient only agreed to cardiopulmonary bypass and intraoperative cell salvage in terms of blood transfusion. Although we discussed the idea of an interventional approach with the cardiology department, the option was abandoned for two main reasons. First, approaching the CAA through the

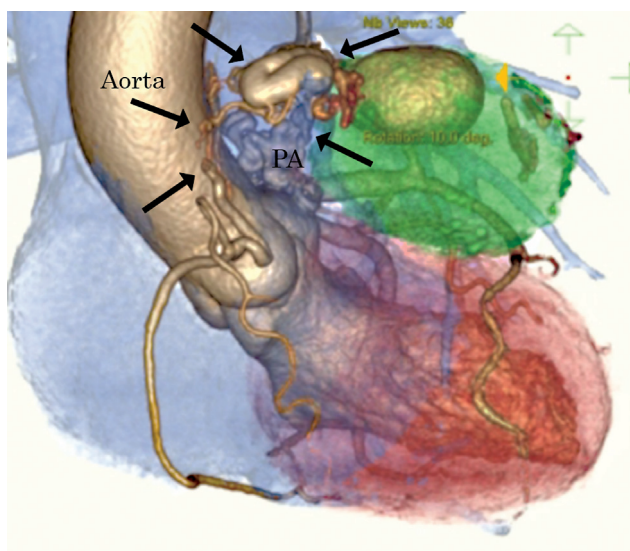


Fig. 2 Preoperative coronary computed tomography of the giant coronary artery aneurysm (CAA) and coronary artery fistula (CAF). Arrows: CAF vessels. Green mass: thrombosed portion of the giant CAA lumen. PA, pulmonary artery.

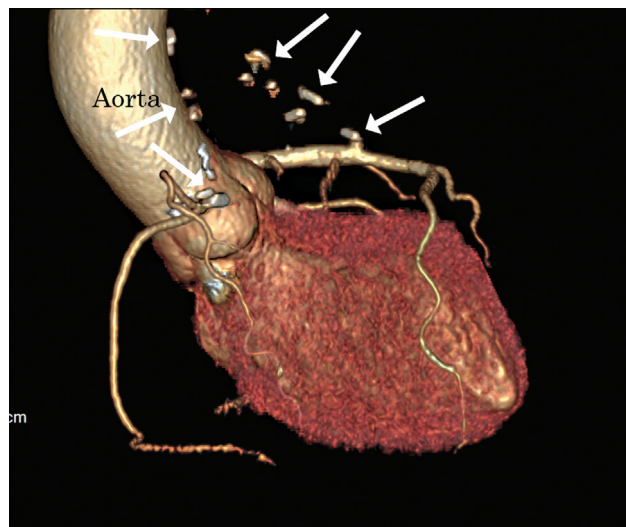


Fig. 3 Postoperative coronary computed tomography. Arrows: surgical clips used to ligate coronary artery fistula.

meandering CAF vessels would be very challenging with the added risk of perforating a CAF vessel. Second, the medical cost of coiling devices needed to completely embolize the giant CAA was unfathomable.

The patient underwent complete resections and ligations of both the giant CAA and CAF. On-pump arrest procedure was necessary to position the heart (lift and rotate from left to right) without jeopardizing intraoperative hemodynamics. First, the feeder coronary arteries (1 branch immediately branching off from the right coronary artery orifice, and 1 branch from the #6 region of the left anterior descending artery) of the CAF were ligated, and resected from the anterior pulmonary artery wall. Closure of CAF entry into the pulmonary artery was reinforced with felted sutures. Because the giant CAA originated from the CAF, we carefully removed the giant CAA in one piece with the entirely resected CAF still attached. Pathological examination of the giant CAA revealed a single feeder vessel from the CAF, and thrombus formation in 95% of the lumen (→ Video 1). Operation time was 265 minutes, with aortic cross-clamp time of 137 minutes, and total blood loss of 479 mL (not including 200 mL of cell salvage). Due to the sheer size of the giant CAA, carefully detaching the lesion from the cardiac surface without injuring the right ventricular wall nor CAF vessels caused the aortic cross-clamp time to exceed 2 hours; we prioritized minimal bleeding over operation time in this case. Postoperative course was uneventful, and she was discharged on postoperative day 16 (→ Fig. 3).

Video 1

Resection and ligation of the giant coronary artery aneurysm and coronary artery fistula. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0042-1757877>.

Discussion

CAFs are congenital anomalies of the coronary artery with a prevalence of 1 in 50 thousand in adults, without sex and race disposition. Small CAFs are usually asymptomatic, but larger CAFs with left to right shunt and/or coronary steal phenomenon can present with symptoms of angina.³⁻⁵ Giant CAAs are also rare with a prevalence of 10 in a million, and usually asymptomatic. The natural history of CAAs is still unclear, but they are associated with atherosclerosis, vasculitis such as Kawasaki disease and Takayasu disease, connective tissue disorders, trauma, percutaneous coronary intervention, and drugs.^{1,2}

The incidence of CAFs coupled with giant CAAs is very rare with an approximate incidence of 3 in 100 million. The limited number of case reports published in the past 30 years have in common that the giant CAAs originate from the CAF, meaning they have not been found to exist separately when coexistent. The precise pathology is unknown, but we speculate that the meandering and uneven flow of CAFs advance atherosclerosis and inflammation in the vessels to accelerate CAA development as well as its growth.

It is important to consider early surgical intervention once giant CAAs are diagnosed, because they can rupture, causing cardiac tamponade and sudden cardiac death. Reports of giant CAA rupture are very scarce, and correlation between CAA size and rupture risk is yet undetermined. Even with the limited data available on giant CAA and rupture cases, it is fair to say that preemptive surgery before rupture is the safer bet for the patient. This case is not remarkable, except the condition is extremely rare, and the patient was a Jehovah's Witness (which is a rare religion in some countries). However, many thoracic surgeons lack direct experience in resection of giant CAAs, thus sharing our experience with footage of the surgical procedure could aid in treatment plans for similar cases in the future.

We recommend surgical resection and ligation of giant CAAs as soon as it is diagnosed, as well as CAFs because it can harvest a (giant) CAA. There are two major surgical techniques in treating giant CAAs: one method is to carefully shave it off in one piece, and the other is to bisect the CAA and ligate the communicating vessels from CAA lumen. There is no consensus on which is the preferred surgical technique, but our recommendation is to remove the CAA in one piece, under cardioplegic arrest to safely rotate the heart to expose the entire CAA and find the exact layer to be dissected, especially in Jehovah's Witness patients to minimize bleeding; bisecting the CAA will lead to increased intraoperative bleeding. From our experience, we are confident that complete resection and ligation of both giant CAA and CAF can be safely performed with minimal cell salvage transfusion in a Jehovah's Witness patient, and that this report might aid in planning treatment for similar cases.

Conflict of Interest

None.

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