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REVIEW ARTICLE

Giant aneurysm of the right coronary artery, report of a case and review of the literature

Aneurisma gigante de la arteria coronaria derecha, reporte de un caso y revisión de la literatura

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Abstract

Coronary artery aneurysms are described as a localized dilatation that exceeds the normal diameter by 1.5 times. This is a rare condition; its incidence varies from 0.3% up to 5.3% of all coronary angiographies. Those aneurysms that exceed 4 times the diameter of a normal artery are considered giant aneurysms, which are even more uncommon, presenting between 0.02% and 0.2% of all cases. There is controversy regarding its pathophysiology, however, up to 50% of the cases are related to atherosclerosis. They are diagnosed more frequently between the sixth and seventh decade of life. The main clinical manifestations are related to ischemic heart disease. Regarding their treatment, there is no general consensus toward its management in adult patients. The options are medical, surgical, or percutaneous treatment. We report the presence of a giant aneurysm of the right coronary artery and giant ectasia of the left coronary system with active thrombosis in a man with a history of an abdominal aortic aneurysm, with endovascular treatment and a non-ST segment elevation myocardial infarction with no reperfusion strategy, who required a coronary computed tomography, identifying the anatomical characteristics of this disease.

Key words: *Coronary artery aneurysms. Giant aneurysm. Ectasia. Ischemic heart disease. Coronary tomography. Mexico.*

Resumen

Los aneurismas de las arterias coronarias se definen como una dilatación localizada que excede el diámetro normal en 1.5 veces. Esta es una condición poco frecuente, su incidencia varía del 0.3 hasta el 5.3% de las angiografías coronarias. Los aneurismas que exceden cuatro veces el diámetro del vaso normal se consideran gigantes. Estos son aún más raros y se presentan en el 0.02 a 0.2% de todos los casos. Existe controversia en cuanto a su fisiopatología, sin embargo, hasta el 50% de los casos se relacionan con la aterosclerosis. Se diagnostican más frecuentemente entre la sexta y séptima décadas

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de vida. Las principales manifestaciones clínicas están relacionadas con la cardiopatía isquémica. Respecto a su tratamiento, no existe un consenso del manejo en los pacientes adultos, las opciones son: médico, quirúrgico o intervencionismo. Reportamos la presencia de un aneurisma gigante de la coronaria derecha y ectasia gigante del sistema izquierdo con trombosis activa en un hombre con antecedentes de un aneurisma en la aorta abdominal, tratado por vía endovascular, e infarto agudo al miocardio sin elevación del segmento ST no reperfundido. Requirió de estudio de angiotomografía coronaria, el cual permitió la identificación de las características anatómicas de esta enfermedad.

Palabras clave: *Aneurismas de las arterias coronarias. Aneurisma gigante. Ectasia. Cardiopatía isquémica. Angiotomografía coronaria. México.*

Introduction

Coronary artery aneurysms (CAAs) are incidental, uncommon findings that often are not identified in patients with coronary artery disease. The causes can be atherosclerosis, trauma, congenital, Kawasaki's disease, and vasculitis, which should be analyzed within the context of age and gender[1,2].

The finding of a giant aneurysm of the right coronary artery and giant ectasia of the left system with active thrombosis merits a review of literature on the subject.

Case report

We report the case of a 66-year-old man with a history of heavy smoking, hypertension, abdominal aortic aneurysm treated by the endovascular route, and acute myocardial infarction without ST-segment elevation, not re-perfused, 5 months before his assessment. The patient was admitted to our institution with aortic stent active infection. He had no cardiovascular symptoms. He was found with normocytic normochromic anemia; electrocardiogram in sinus rhythm with left atrial abnormality, transthoracic echocardiogram showed concentric hypertrophy of the left ventricle; akinesia of the middle and basal thirds of the inferolateral wall; left ventricle ejection fraction was 52%, tricuspid annular plane systolic excursion was 21 mm, there was slight biauricular dilation and pulmonary artery systolic pressure of 44 mmHg. SPECT with technetium 99 demonstrated infarction of the lower wall and the inferoseptal region that spread from the middle to the basal third, not transmural in the apical and middle thirds, and transmural infarction in the basal third, with slight ischemia in the residual tissue.

Computed tomography (CT) angiography demonstrated a right coronary artery giant aneurysm, diffuse ectasia, and circumflex artery aneurysm (Figs. 1-3).

The patient underwent resection of the infected juxtarenal abdominal aortic aneurysm and stent, aorto-bi-iliac bypass with silver-impregnated prosthesis, aortoenteric fistula resection, and placement of a retrocolic omental patch. He had a satisfactory evolution. Conservative management of the giant coronary aneurysm with antiplatelet and anticoagulant drugs was decided.

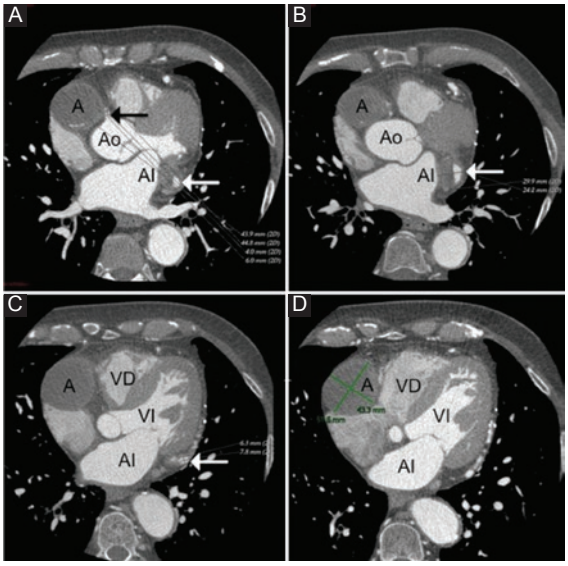

Figure 1. A and D: Right coronary artery, 6-mm permeable proximal arteries with a diameter of 4.1 × 4.3 mm (black arrow: right coronary ostium). Subsequently and up to the crux cordis there is no coronary flow. Diffuse right coronary aneurysmal dilation is observed. The diameter of the proximal third is 44 × 45 mm, that of the middle third, 51 × 43 mm, and that in the distal third, 8.5 × 10.2 mm. **B and C:** circumflex artery. Permeable in all its extension with diffuse ectasia, proximal diameter of 6.2 × 7.6 mm, middle of 9.9 × 11.6 mm, and distal of 9.8 × 7.1 mm, aneurysmal dilation and intramural thrombus (white arrow), in the proximal and middle thirds of these segments. Vascular diameters are 23.5 × 22.6 mm and 29.9 × 24.2 mm. A: giant aneurysm of the right coronary artery; AI: left atrium; Ao: aorta; VD: right ventricle; VI: left ventricle.

Review of the topic

CAAs are defined as localized and irreversible dilation that exceeds 1.5 times the diameter of normal

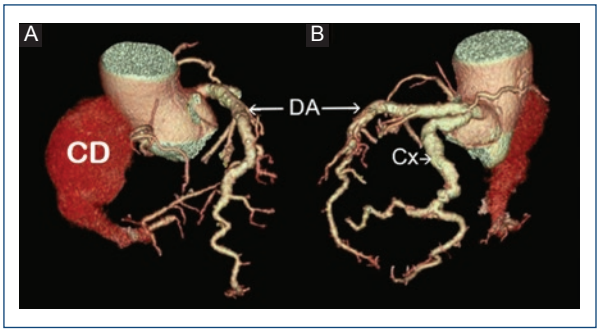

Figura 2. 3D reconstruction. **A:** right coronary artery giant aneurysm image. **B:** circumflex artery diffuse ectasia.
CD: right coronary; Cx: circumflex artery; DA: anterior descending artery.

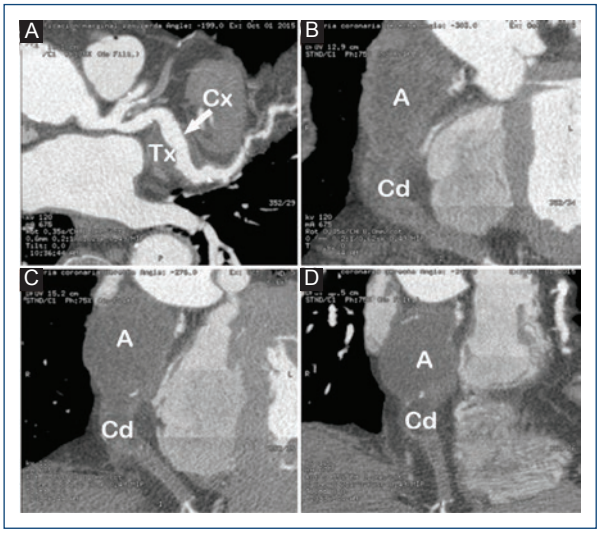

Figure 3. Multiplanar reconstruction showing: diffuse circumflex artery (Cx) ectasia with aneurysmal dilation in the proximal and middle third with intramural thrombus (Tx) (**A**), and giant aneurysm of the right coronary artery (**B-D**). A: aneurysm; Cd: right coronary artery (distal to the aneurysm with ectasia).

adjacent segments, or the diameter of the largest coronary vessel. In contrast, the term ectasia is used to describe a diffuse dilatation of the coronary arteries involving 50% or more of the length of the artery[1,2].

CAAs were first described by Morgagni in 1761 and the first CAA report was made by Bourgon in 1812. In 1929, Packard and Wechsler reported 21 cases[3].

CAA incidence ranges from 0.3 to 5.3% in the Coronary Artery Surgery Study, one of the largest, which included 20,087 patients undergoing coronarography, and reported coronary aneurysms in 4.9% of patients[4].

The right coronary artery is the most commonly involved, in 40 to 70%, the circumflex artery in 23% and the anterior descending artery in 32%, although it depends on the series. The involvement of three vessels or the left coronary trunk is much rarer (3.5%). Coronary aneurysms due to atherosclerosis or of inflammatory cause are usually multiple and involve more than one coronary artery. In contrast, congenital, traumatic or dissection-related aneurysms, typically involve a single artery[5].

They occur more in men and mean age at diagnosis ranges between 54 and 65 years. An association between coronary and aortic aneurysms has been described in up to 30% of cases[6].

There is no universally accepted definition of giant coronary aneurysm. In the medical literature, definitions have been proposed such as a diameter larger than 20 mm, 40 mm, and 50 mm, as well as 4 times the diameter of the reference vessel. The data published on giant coronary aneurysms report a prevalence of 0.02-0.2% (prevalence of giant aneurysms ≥ 50 mm in diameter is 0.02%)[7].

Classification

Morphologically, CAAs are defined as saccular when their longest transverse diameter exceeds the longitudinal and fusiform when their longitudinal diameter is longer than their longest transverse diameter.

Histopathologically, CAAs are classified as true when the vascular wall contains all normal vascular layers, or as pseudoaneurysms (typically saccular) when there is a loss of integrity of the normal vascular wall, which results in the formation of thin-walled structures lacking a complete arterial wall[8]. Markis et al., in 1976, proposed a classification based on coronary dilation degree of involvement and distribution: type I, diffuse ectasia with aneurysmal lesion in two or three vessels; type 2, diffuse ectasia in one vessel and localized ectasia in another vessel; type 3, diffuse ectasia in one vessel; and type 4, localized ectasia in one vessel[1,9].

Pathophysiology

As regard giant aneurysms related risk factors and pathophysiological mechanisms, there is controversy in the literature; however, the presence of diabetes mellitus, smoking, and dyslipidemia has been reported to be more common in patients with coronary aneurysms in comparison with healthy controls[10].

Table 1. Etiology of coronary aneurysmal disease

(A) Congenital
Congenital heart diseases
Coronary arteriovenous malformations
Hereditary disorders
– Marfan syndrome
– Type 1 neurofibromatosis
– Polycystic kidney disease
– Hereditary hemorrhagic telangiectasia
– Ehlers–Danlos type IV
– Fibromuscular dysplasia
(B) Acquired
Atherosclerosis
Kawasaki's syndrome
Drugs (cocaine)
Connective tissue disorders
– Systemic lupus erythematosus
– Behcet's disease
– Relapsing polychondritis
– Takayasu arteritis
– Direct coronary atherectomy
– Balloon or stent angioplasty
– Intracoronary post–brachytherapy
Graft vasculopathy

Modified from Chrissoheris et al.[11]

Although the underlying pathological processes vary for each etiology, there is a common process inherent to weakening and subsequent dilatation of the vessel wall.

There are multiple related etiologies. It is attributed atherosclerosis in up to 50% of cases, while 20-30% are considered to be of congenital origin. In most of these patients, ectasia coexists with coronary artery disease[10]. About 10-20% are associated with connective tissue or inflammatory diseases[2].

A large number of connective tissue and inflammatory disorders have been linked to coronary aneurysms (Table 1)[11]. The best known is the association with Kawasaki's disease, although it has also been reported in patients with Takayasu arteritis, systemic lupus erythematosus, rheumatoid arthritis, Behcet's disease, Marfan syndrome, and Ehlers-Danlos syndrome[12].

Coronary aneurysms have also been observed in association with infections, drug use, and trauma. Iatrogenic etiology, after coronary angioplasty attempt, is a rare cause (0.3-0.6%); these cases are triggered after the implantation of a drug-releasing stent due to the effects of antiproliferative agents in the intima, secondary to an arterial wall deep injury due to dilatation with large balloons or stents, insufflations at high atmospheres or atherectomy[13,14].

Histopathological findings on autopsies have revealed extensive changes related to atherosclerosis, with destruction or thinning of the vascular wall middle layer, similar to those found in coronary artery disease. Metalloproteinases have been implied in coronary aneurysm formation, secondary to extracellular matrix protein degradation[15].

Clinical presentation

In most cases, CAAs are asymptomatic. Regularly, clinical manifestations are similar to those observed in coronary artery disease but can vary according to the underlying cause. CAAs promote thrombosis through an abnormal flow condition; within the aneurysm, coronary flow is relatively slow or static, which promotes platelet activation and thrombi formation. The presence of chronic thrombi can also promote thrombogenesis, by providing procoagulant factors and fibrin, which acts as a nest for the formation of a new clot. In consequence, a common finding is the presence of thrombosis with subsequent embolization, which causes angina, dyspnea, ischemia, infarction, or sudden death[5,7,16].

Diagnosis

The gold standard for the diagnosis of CAAs is invasive coronary angiography, which provides information about the size, shape, location, and number of aneurysms. However, flow can only be evaluated within the lumen and does not provide information on the structure of the vascular wall. The employment of intravascular ultrasound allows to differentiate between true and false aneurysms[17].

Currently, coronary CT angiography has a high spatial and temporal resolution. It provides a safe non-invasive approach to accurately delineate the structure of the arteries, and precise information is obtained about the dilation characteristics, longest diameter, length, shape, number, morphological characteristics, presence of associated stenosis, precise anatomical localization, and relationship with surrounding anatomical structures. However, this method requires radiation exposure and the use of iodinated contrast medium.

Coronary magnetic resonance angiography is a diagnostic alternative, whose usefulness has been reported by several researchers. The echocardiogram allows visualizing dilatations close to the origin of the coronary arteries[18].

Treatment

The appropriate treatment for CAAs is controversial and depends on the particular clinical situation. There

is abundant literature on the management of pediatric patients with coronary aneurysms associated with Kawasaki's disease; however, in adult patients, where etiologies are different, no treatment has been validated in clinical trials[12].

Therapeutic options in CAA consist of pharmacological, surgical, and endovascular treatment. Medical therapy consists in preventing thromboembolic complications through the administration of antiplatelet agents and anticoagulants.

Surgical management is appropriate in symptomatic patients with significant coronary artery disease, evidence of embolization, myocardial ischemia, or at risk of aneurysm rupture.

In the context of patients with the acute coronary syndrome, coronary angiography usually demonstrates the presence of thrombus; however, chronicity of the thrombus is unknown, since it is a common finding even in asymptomatic patients[15].

In patients with Kawasaki's disease and acute coronary syndrome, treatment with thrombolysis using intracoronary urokinase, in addition to anticoagulation with heparin or oral anticoagulants, has been described.[19]

Lima et al. reported two cases of left coronary trunk CAA treated with warfarin and acetylsalicylic acid[20].

In patients with an acute coronary syndrome without ST-segment elevation and CAA, Boyer et al. suggested considering revascularization of the aneurysm vessels with thrombolysis in myocardial infarction with 0 or 1 flow quality, as well as in patients with angina, recurrent ischemia, sustained ventricular tachycardia, or hemodynamic instability. If these findings are not present during initial clinical evaluation and diagnostic angiography, they recommend a conservative strategy that includes dual antiplatelet therapy, anticoagulation with unfractionated or low molecular weight heparin and considering the use of glycoprotein IIb/IIIa inhibitors for 24-48 h, particularly when there is an angiographically significant or evident thrombus[15].

For long-term management in patients with CAA and some other additional indication for chronic anticoagulation, the use of acetylsalicylic acid at 81 mg/day and anticoagulation with warfarin is recommended, with International Normalized Ratio targets between two and three.

In patients without prior indication for anticoagulation, Boyer et al. recommend dual antiplatelet therapy with 81 mg of acetylsalicylic acid and clopidogrel, prasugrel, or ticagrelor, regardless of choosing a conservative or invasive strategy. The duration of dual antiplatelet therapy is not defined; however, maintaining it for a prolonged period is recommended[15].

The role of the new oral anticoagulants for the treatment of CAA is not known. Off-label use can be considered after discussion with the patient, including careful consideration of possible risks and benefits[15].

It should be emphasized that long-term medical treatment based on antiplatelet agents and anticoagulants is inferred from the beneficial effects observed in patients with Kawasaki's disease and associated giant aneurysms. However, this treatment has not been tested in randomized prospective trials, and thus treatment choice should be based on a careful risk-benefit evaluation[7].

In cases of CAA with ischemic symptoms despite medical treatment, percutaneous or surgical revascularization is necessary. Percutaneous implantation of polytetrafluoroethylene (PTFE)-coated stents has gained popularity due to their ability to effectively limit the expansion of CAAs by reducing parietal stress within the aneurysm, and thus avoid its rupture. Some authors have suggested that PTFE-coated stents should be restricted to patients whose aneurysms are <10 mm in diameter. Implantation of bare and drug-eluting stents has been used with good results. Other authors have documented the successful treatment of CAAs by embolization with coils. Coronary intervention entails challenges due to the risk of distal embolization, secondary branch occlusion when using coated stents, and the possibility of stent malapposition, due to the increased risk of thrombosis and intra-stent restenosis. The use of intravascular ultrasound or optical coherence tomography may improve the definition of the lesions, including vessel diameter, the presence of thrombus and the relationship of CAAs with secondary vessels[15].

Surgery may be considered in the presence of giant aneurysms, left coronary trunk involvement, bifurcation, or multiple-vessel lesions, evidence of embolism from the aneurysm to the distal coronary bed and progressive growth of an AAC documented by serial angiographic measurements. Surgical treatment involves coronary artery bypass, with or without aneurysm ligation or resection[15,19].

To clarify some of the questions that remain unanswered, the results are awaited of the international registry of coronary aneurysms (Rationale and design of a multicenter, international, and collaborative CAA Registry CAAR; NCT02563626), which has recently completed patient recruitment and is expected to

become the largest series of patients with coronary aneurysms so far[21].

Conclusion

Giant coronary aneurysms are an uncommon finding where the right coronary artery is mostly affected. There are multiple etiologies related to their development; however, coronary atherosclerosis is the most common cause of patients undergoing angiography. Various imaging methods can be used for diagnosis, among which coronary angiography is the gold standard; however, coronary artery CT angiography is a non-invasive method that adequately identifies ectasia and coronary aneurysms anatomical features.

Its clinical presentation varies from the asymptomatic patient to the most challenging scenario, an acute coronary syndrome. Treatment is a controversial issue because there is a lack of perspective and randomized clinical trials evaluating the variety of available therapeutic options, which range from pharmacological treatment with antiplatelet and anticoagulant agents to invasive treatment through percutaneous coronary intervention or aortocoronary bypass surgery. The finding of a giant aneurysm requires, regardless of the symptoms, for management to be individualized by assessing the possible risks and benefits.

Conflicts of interest

None.

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Ethical disclosures

Protection of people and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained written informed consent of the patients and/or subjects mentioned in the article.

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