Aortic pseudoaneurysm with a fistula between the non-coronary sinus and right atrium: a case report

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Abstract

The authors present a case report of a 68-year-old man evaluated at the emergency department for repeated syncope, asthenia, and general malaise, suggesting heart failure in a patient with several comorbidities. At presentation, the patient was afebrile, but he had reported a low-grade fever in the previous 6 months. At first glance, transthoracic echocardiography was not clear, while transesophageal echocardiography revealed an echo-free image at the level of the non-coronary sinus of the aortic root, suggestive of a pseudoaneurysm communicating with the right atrium with continuous systo-diastolic flow, compatible with the aorto-cavitary fistula between the aortic root and the RA. Echocardiographic findings were confirmed by cardiac computed tomography. The case was discussed with the heart team and was considered suitable for surgery, but the patient suddenly died just before surgery due to impairment and friability.

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Key words: aortic pseudoaneurysm, heart failure, fistula, right atrium, endocarditis, echocardiography.

Contributions: all authors equally contributed to the draft of the manuscript and gave their final approval.

Conflict of interest: the authors declare no potential conflict of interest.

Ethics approval and consent to participate: no ethical committee approval was required for this case report by the Department, because this article does not contain any studies with human participants or animals.

Patient consent for publication: informed consent was obtained from the patient's relatives for publication of this case report and any accompanying pictures.

Availability of data and materials: all data underlying the findings are fully available.

Funding: none.

Received: 28 April 2024.
Accepted: 23 May 2024.
Early view: 29 August 2024.

Monaldi Archives for Chest Disease 2025; 95:3039
doi: 10.4081/monaldi.2024.3039

Key Clinical Message

Connections between the aorta and the right atrium (RA) are rare anomalies and usually involve the ascending aorta in the form of congenital coronary cusp fistulas or acquired connections associated with aortic dissection and infective endocarditis (IE). Both can remain clinically silent for a long time, and 50% of cases are associated with aortic regurgitation. Symptoms may be caused by mechanical obstruction, compression of the conduction system, or intracardiac rupture. The volume overload to the RA may lead to rapid development of pulmonary vascular disease. Early diagnosis and treatment, especially in the presence of undiagnosed IE and other congenital heart defects, are of paramount importance for the outcome.

Introduction

Aorto-cardiac fistulas, also called aorto cavitary fistulae (ACFs), are rare abnormal connections between the aorta and chambers of the heart. The incidence and prevalence of this entity are unknown, as ACFs are rare, and they are often found on post-mortem examinations [1]. ACFs can be congenital or acquired [2], associated with aortic dissection, frequently caused by an iatrogenic or infectious process [3]. In IE, this complication is rare and has been reported to be estimated at 1-2% of all cases [4]. Clinically, ACF patients can range from asymptomatic to signs and symptoms of heart failure and cardiogenic shock [5]. Without diagnosis, adequate closure, and treatment of the underlying cause of ACF, patients often die [4]. The diagnosis requires high clinical suspicion and the use of imaging modalities, including echocardiography. Even though a transthoracic echocardiogram (TTE) is the first diagnostic approach, a transesophageal echocardiogram (TEE) has a rate of diagnostic detection up to 97.8%. There is no consensus on

the management of ACF, and various management strategies exist, such as medical management of symptoms or infection, or attempts at surgical or percutaneous closure. There is little data in the literature on the etiology, symptoms, and management of acquired ACF; however, if left untreated, mortality has proven to be high [1,4,5]. We reported a case of IE-related fistula between the aorta and RA and its multimaging approach and outcome.

Case Report

The patient was a 68-year-old man, a former heavy smoker, with multiple comorbidities such as pulmonary emphysema-type chronic obstructive pulmonary disease, advanced arteriopathy obliterating lower limbs, exotoxic liver cirrhosis complicated by portal hypertension, ascitic decompensation, small esophageal varices, and diffuse gastropathy.

The patient was evaluated at the emergency department for repeated syncope, inability to maintain an upright position, and difficulty walking with easy fatigability. With a scrupulous anamnesis, it was discovered that the patient had had fever in the previous 6 months, associated with general malaise. The physical examination revealed poor general conditions due to malnutrition, a new onset systolic murmur of 3/6 L. During hospitalization, the patient was still afebrile, and laboratory results did not show markers of acute infection. All possible causes of syncope were investigated and found to be negative. He underwent a chest-abdomen computed tomography (CT) scan, which showed abundant bilateral pleural effusion and abundant free intra-abdominal effusion. Transthoracic cardiac ultrasound was requested because of a cardiac murmur and to define the severity of pulmonary systolic pressure, if any. TTE showed II-III degree diastolic dysfunction, slightly dilated right ventricle with preserved longitudinal contractile function (tricuspid annular plane systolic excursion 26 mm, S' at tissue doppler imaging 15.4 cm/sec, free wall strain 27%), slight bi-atrial dilatation. The presence of turbulent flow at the level of the tricuspid valve was interpreted as at least moderate tricuspid regurgitation (TR), and the derived pulmonary artery systolic pressure was mildly increased (pulmonary artery systolic pressure 48 mmHg). Although the TR was deeply investigated, it was not possible to identify the point of convergence on the tricuspid. The inferior vena cava was normal in diameter and hypocollapsible on inspiration, suggesting a slight increase in pressure in the RA. It was also found to have mild to moderate aortic valve stenosis with associated mild valvular regurgitation (Figure 1). Since the supposed TR was not clear, the patient underwent TEE evaluation that clearly showed the presence of a drop of echo at the level of the right coronary artery (RCA) and non-coronary artery communicating with (approximately 3 mm) the RA (5 mm communication) with continuous systo-diastolic flow suggestive of the fistulized abscess between aorta and RA and right chambers volume overload (Figures 2 and 3). Coronary CT confirmed the presence of a pseudoaneurysmal cavity of approximately 29×22×28 mm, extending under the proximal section of the RCA. The cavity had a wide communication with the non-coronary sinus (9 mm) and with the RA (7 mm), with a para-aortic pseudoaneurysm with fistula between the non-coronary sinus and RA (Figure 4). In addition to the specific findings, it was also found that they have three vessels disease. Considering the history of fever, microbiological cultures on blood and urine were done, but they were always negative. Doppler ultrasound of the supra-aortic trunks showed a stenosis of the right internal carotid artery of 70%. The patient was treated medically for heart failure with progressive improvement of signs and symptoms. The case was discussed in the heart team and approved for cardiac surgery, but died just the night before. Cirrhosis and diffuse atherosclerosis have been confirmed by the autopsy, but the cause of sudden death was not found. On the other hand, the ACF was confirmed (Figure 4).

Discussion

The etiology of ACFs can be due to primary causes such as congenital malformations [6,7], or secondary causes such as paravalvular abscesses, ruptured sinus of Valsalva aneurysms, aortic dissections, trauma, IE, or iatrogenic causes such as occurring after intravascular procedures or valve replacements [8,9]. In our case, ACF was found in a very compromised, fragile patient, with several comorbidities, a situation that made diagnosis and treatment particularly difficult and which ended in death. Moreover, the infection caused endocarditis likely happened several months before the present hospital admission, and the evidence of the fistula between the aorta and RA was obtained by change.

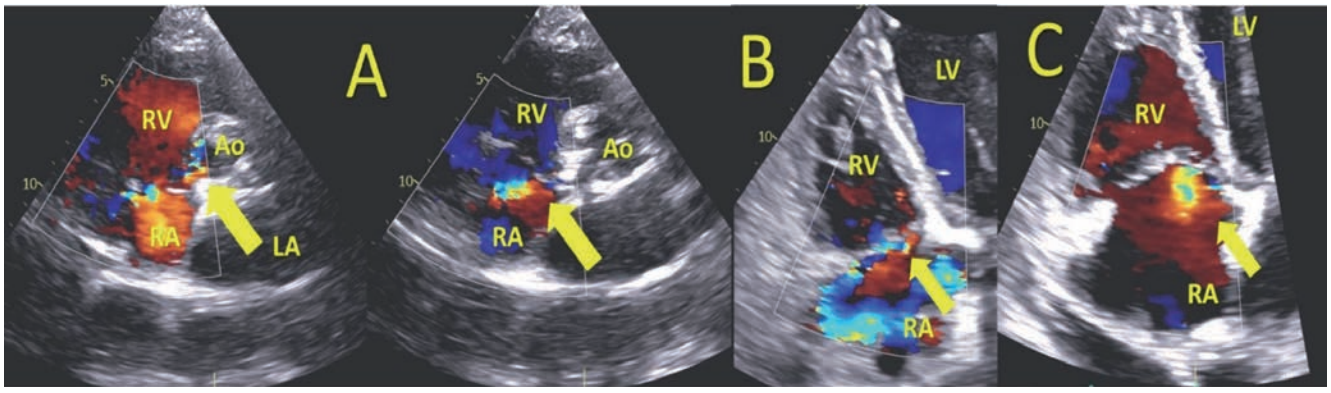

Figure 1. A) Parasternal short axis in transthoracic echocardiogram (TTE): strange turbulent flow from the aorta to the right atrium; B,C) A4C in TTE: turbulent sisto-diastolic flow directed into the right atrium, mixed with the jet of tricuspid insufficiency. Ao, aorta; LV, left ventricle; RV, right ventricle; RA, right atrium.

ACFs can be classified based on the location of the fistula from the aorta to the heart chamber, commonly from the aorta to the RA or pulmonary artery. Patients presenting with ACF may be asymptomatic and therefore be identified incidentally [10], or with mild to severe symptoms of heart failure, as pedal edema, dyspnea, fatigue, and chest pain, depending on the size of the fistula [5,11]. Patients with ACF have a continuous systolic murmur on auscultation, caused by blood flow through the shunt. Transesophageal echocardiography has been shown to provide better visualization than TTE, as it has a higher sensitivity and specificity for diagnosing ACFs [2,4,12]. Angio-CT may be useful to characterize the underlying cause, as in the case of aortic dissection, as an additional imaging modality [1]; moreover, by combining the two modalities, the sensitivity of diagnosing abscess/pseudoaneurysm/fistula can increase up to 100%. The shunt flow severity can be determined by the Qp/Qs ratio, which is usually determined by oximetry during right heart catheterization [13]. There is no consensus in the literature regarding the management of ACFs due to their rarity, unlike shunts such as atrial septal defects (ASD), in which the decision for shunt closure is established by the Qp/Qs ratio [14,15]. Symptomatic patients with ACF often experience volume overload and signs of heart failure

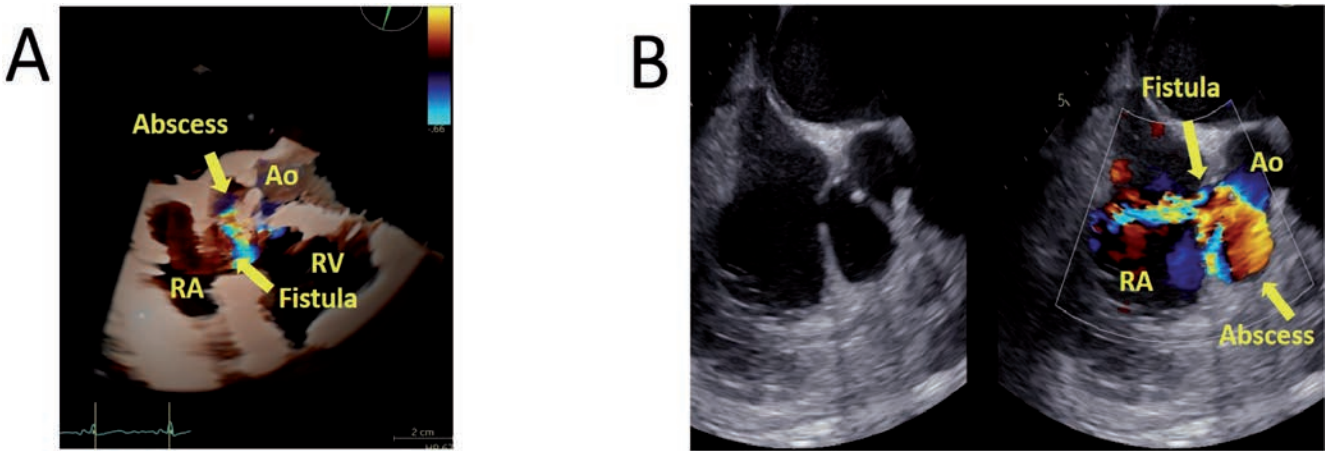

Figure 2. A) Long axis in transesophageal echocardiogram: turbulent flow through a fistula from the aortic abscess to the right atrium; B) 4 chambers: focus on the atria, which shows, at color Doppler, a turbulent flow from the aortic abscess to the right atrium. Ao, aorta; LV, left ventricle; RV, right ventricle; RA, right atrium.

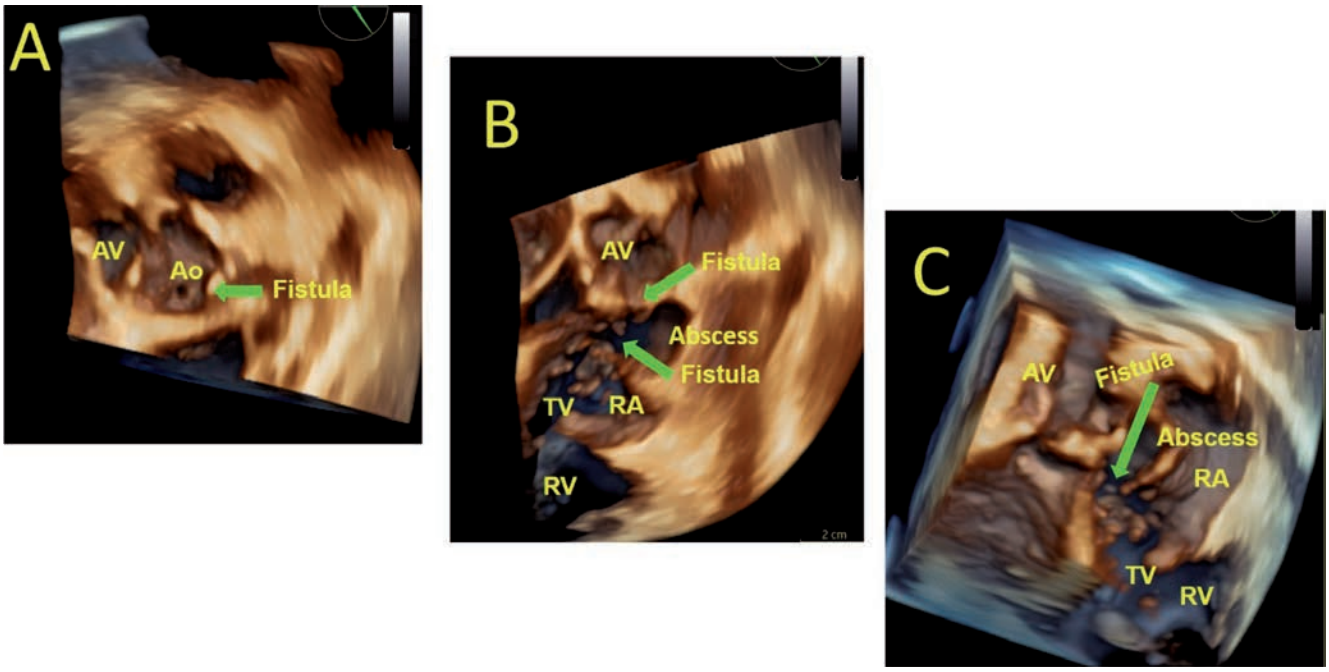

Figure 3. Transesophageal echocardiogram 3D reconstruction. A) Non-coronary sinus with the fistula; B) aortic root and right chamber; C) aortic root and right chambers with abscess and fistula. AV, aortic valve; Ao, aorta; RA, right atrium; RV, right ventricle; TV, tricuspid valve.

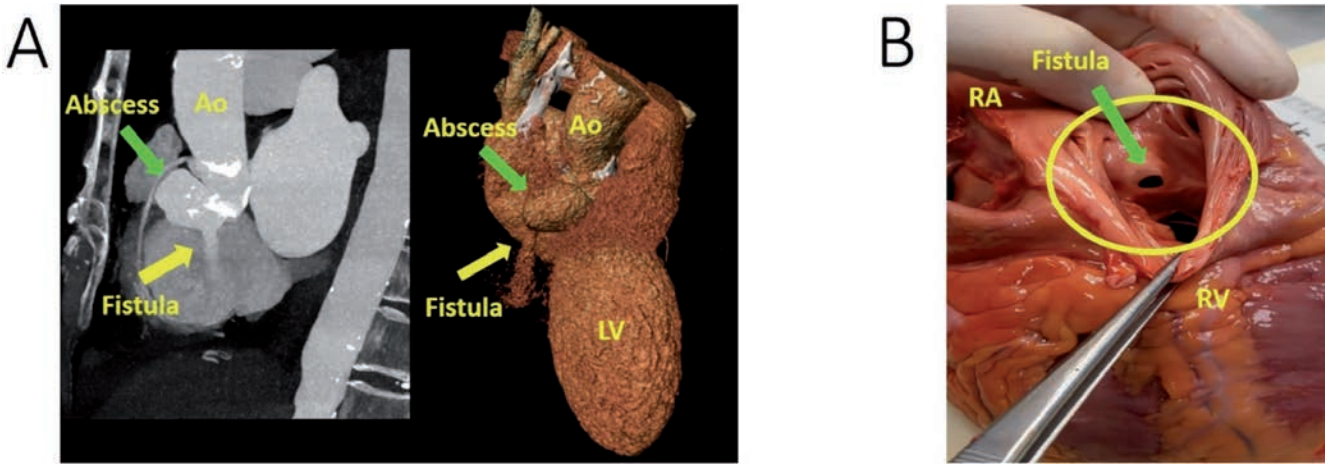

Figure 4. A) Coronary computed tomography: aorto-right atrium fistula. 3D reconstruction of the fistula between the pseudoaneurysm (green arrow) of the aorta and the right atrium. The yellow arrow points out the jet between the abscess and the right atrium. The right chambers were removed from the heart; B) autopsy findings: fistula from the aorta into the right atrium. Ao, aorta; LV, left ventricle; RA, right atrium; RV, right ventricle.

are due to shunting of blood from the aorta to the right or left atrium and then to the right or left ventricle [7]. Large shunts have been observed to result in increased severity of heart failure and increased volume overload, and can be associated with ACF up to 52% and it is an independent risk factor of mortality [4,16]. Qp/Qs ratio could become important in the decision-making process for the treatment of ACF, similar to the decision-making process for the treatment of ASD [14,15]. Our patient had a high-flow ACF with already dilated right chambers responsible for the heart failure, which was superimposed on the clinical status determined by the decompensated cirrhosis, with the indication of surgical closure. The first surgical repair and closure of an ACF was reported by Temple *et al.* in 1966. The first transcatheter closure of ACFs was reported 20 years later by Hayward *et al*. in 1988 [17], by a detachable balloon device. Recent successful attempts at percutaneous fistula closure have also been made utilizing occluder devices or coil embolization [18]. Recent systematic reviews shed light on this highly morbid condition. Once recognized, surgical fistula closure appears to be superior to conservative management in patients with post-IE [19].

Conclusions

Aorto-cardiac fistula is a rare disease, often fatal because it occurs in compromised patients, as in our case report, but its entity is progressively reported; therefore, it must be looked for in the differential diagnoses. Heart failure and/or hemodynamic instability are the most frequent symptoms in patients with ACFs, carrying a high risk of mortality. Therefore, ACF could represent a possible differential diagnosis in patients with IE previous invasive cardiac surgery. TEE is the best imaging method to diagnose ACF due to its high sensitivity and specificity. The common locations where they can be found are from the aorta to the RA and from the aorta to the pulmonary artery. Once recognized, closure of the fistula by transcatheter or surgical means appears to be superior to conservative management.

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