Review paper

Coronary artery fistula: key clinical and therapeutic management aspects. A comprehensive literature review

Przetoka tętnicy wieńcowej – kluczowe zagadnienia dotyczące postępowania klinicznego i terapeutycznego. Kompleksowy przegląd piśmiennictwa

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Słowa kluczowe: przetoka tętnicy wieńcowej, niewydolność serca, zawał mięśnia sercowego, przetoki.

Abstract

Coronary artery fistula (CAF) is a rare structural anomaly characterized by an abnormal connection between a coronary artery and a vessel or cardiac chamber. The natural history of CAF is highly variable, and it may cause significant cardiac morbidity; however, it has yet to be fully elucidated. Although uncommon, CAF is an important consideration in the differential diagnosis of common pulmonary and cardiac issues, such as dyspnoea or chest pain. While small fistulae are often asymptomatic and require no treatment, symptoms and complications, such as myocardial infarction, heart failure, or arrhythmias, especially in cases of significant fistula size, may necessitate percutaneous or surgical closure. The management of each fistula is tailored to its specific characteristics. Notably, most studies on CAF are case reports with small patient cohorts, and several aspects of CAF management still lack clear guidelines.

Streszczenie

Przetoka tętnicy wieńcowej (CAF) to rzadka anomalia strukturalna charakteryzująca się nieprawidłowym połączeniem tętnicy wieńcowej z naczyniem lub jamą serca. Historia naturalna CAF jest bardzo zmienna i może powodować znaczną zapadalność na choroby serca; jednak nie zostało to jeszcze w pełni wyjaśnione. Chociaż CAF jest rzadka, to należy brać ją pod uwagę w diagnostyce różnicowej w przypadku występowania powszechnych objawów chorób płuc lub serca, takich jak duszność lub dolegliwości bólowe klatki piersiowej. Mimo że małe przetoki często przebiegają bezobjawowo i nie wymagają leczenia, to objawy i powikłania, takie jak zawał mięśnia sercowego, niewydolność serca lub zaburzenia rytmu, szczególnie w przypadku przetok o znacznych rozmiarach, mogą wymagać ich przezskórnego lub chirurgicznego zamknięcia. Postępowanie w przypadku każdej przetoki jest dostosowane do jej specyficznych cech. Warto zauważyć, że większość badań dotyczących CAF to opisy przypadków z udziałem małych grup pacjentów. W kilku aspektach postępowania z CAF nadal brakuje jasnych wytycznych.

Introduction

Coronary artery fistula (CAF) is an uncommon anatomical abnormality characterized by an abnormal connection between a coronary artery and a vessel or cardiac chamber. While many CAFs are small and clinically insignificant, some can be substantial, resulting in significant blood flow diversion away from the coronary circulation, leading to various haemodynamic consequences. The first documented description of anomalous coronary arteries with fistulae dates back to 1841, by the Austrian anatomist Josef Hyrtl [1]. Another description of the CAF was from 1865 by German anatomist Wilhelm Krause [2]. Over the following 8 decades, CAFs were primarily identified post-mortem. In 1947 Biorck and Crafoord performed the first pre-mortem diagnosis and repair of a CAF during an operation on a young boy initially suspected of having a patent ductus arteriosus. A CAF was discovered and ligated, resulting in subsequent symptomatic improvement and resolution of abnormal physical findings [3].

Epidemiology of coronary artery fistulae

The precise prevalence of CAF remains unknown; however, its estimated incidence in the general population is approximately 0.002% [4]. CAF accounts for approximately 0.2–0.4% of all congenital heart diseases [5] and approximately 14% of all coronary anoma-

lies [6]. Moreover, CAF has been reported in 0.06% of children undergoing echocardiography [7] and in approximately 0.13–0.22% of adults undergoing coronary angiography [8–10]. CAF exhibits no racial or sex predilection [11, 12]. In 20–45% of cases, congenital CAF is associated with other cardiac malformations, including ventricular or atrial septal defects, patent ductus arteriosus, bicuspid aortic valve, pulmonary atresia, and tetralogy of Fallot [13, 14].

Aetiology

Most CAFs are congenital in origin. However, they have also been reported as acquired conditions following chest trauma, endomyocardial biopsy, neoplasms, acute myocardial infarction, infective endocarditis, coronary angioplasty, cardiac surgery, or rupture of a coronary artery aneurysm [15, 16].

Morphology

Multiple fistulae are present in 10.7–16% of cases, while a single fistula is significantly more common, occurring in up to 90% of all cases [11, 12]. Fistulae originating from the proximal parts of the coronary artery or its branches are often large. When located distally, they are typically smaller and more tortuous [17]. Approximately 52% of cases have fistulae originating from the right coronary artery, approximately 30% from the left anterior descending coronary artery, and about 18% from the circumflex coronary artery [18]. Low-pressure structures are the most common sites for CAF drainage. The most common drainage sites are the right ventricle (41%), right atrium (26%), pulmonary artery (17%), coronary sinus (7%), left atrium (5%), left ventricle (3%), and superior vena cava (1%) [12].

Pathophysiology and clinical presentation

The pathophysiological manifestation of CAF is dependent on the blood flow resistance along the fistula and the difference in blood pressure between the coronary artery and the draining area of the fistula. Blood flow resistance is related to the size, tortuosity, length of the fistula, and the morphological and biochemical properties of the blood. Although the size of the CAF is a major determinant of its clinical significance and further management, there is no consensus regarding how to categorize the absolute fistula size. For example, fistula sizes were determined by Shah *et al.* in reference to the native coronary arteries proximal to the fistula origin, with CAF up to 2 times the diameter of the distal native coronary artery determined as moderate. Fistulae > 2 to ≤ 3 times the size were determined as large, and those > 3 times the size of the native coronary artery were considered giant CAF [19], Al-Hijji *et al.* categorized CAF size as small, medium, or large based on the fistula diameter relative to the largest diameter of the coronary vessel not feeding the coronary fistula. Fistulae < 1, 1–2, or > 2-times the largest diameter were considered as small, medium, or large, respectively [20]. Latson defined medium-sized fistulae when the CAF diameter is larger than twice but less than 3 times the expected proximate normal coronary artery diameter, or it is associated with similar ranges of dilation of the proximal associated coronary artery; fistulae with smaller dimensions were categorized as small, and those with larger dimensions as large fistulae [21]. Kiefer *et al.* defined small connections of coronary arteries as having vessel diameters of < 1 mm [22]. Most CAFs, especially small fistulae, are asymptomatic and diagnosed incidentally. Some CAFs can spontaneously close, while larger ones tend to enlarge over time [11]. Physical examination results are usually normal. Occasionally, diastolic, systolic, or continuous murmurs are observed. Typically, a cardiac murmur is continuous with diastolic accentuation, with its location on the chest wall being the most audible in the area of the fistula drainage site. If the fistulae drain into the right atrium, a cardiac murmur is audible along the edge of the sternum. When fistulae drain into the left ventricle, the murmur is most audible in the apex region, and if they drain into the pulmonary artery, it is most audible in the second intercostal space to the left of the sternum [11, 17, 21, 23]. The main pathophysiological changes are believed to be the coronary steal phenomenon, which is myocardial ischaemia resulting from the diversion of blood from normal myocardial circulation, and volume overload, which could lead to dilatation of the heart atria and chambers. Dilatation of the cardiac regions depends on the drainage site of the fistula. The left-to-right shunt causes volume overload in the right heart, whereas the left-to-left shunt causes volume overload in the left heart [11, 17]. Symptomatic fistulae are CAFs that are potentially responsible for complications, such as myocardial ischaemia, myocardial infarction, unexplained cardiac chamber dilation and/or dysfunction, congestive heart failure, pulmonary hypertension, arrhythmias, endocarditis, vessel aneurysm and rupture, or vessel thrombosis. The most common clinical symptoms of CAF include fatigue, dyspnoea, chest pain, or syncope [17, 18, 24]. The risk of fistula complications and symptoms increases with age. Moreover, a suggestion has been made regarding the association between CAFs and coronary atherosclerosis, but the relationship remains uncertain [25]. Approximately 30% of the CAFs in adults are associated with coronary atherosclerosis [26]. CAFs may lead to premature atherosclerosis in the shear-induced intimal damage mechanism owing to turbulent vascular blood flow [27]. Moreover, the risk of atherosclerosis may be higher if ectasia and dilatation of the coronary artery persist or progress [28].

The natural history of CAF is highly variable and has not yet been fully elucidated.

Diagnostic and clinical management

Physical examination is the first step in diagnostic management; however, similarly to a 12-lead electrocardiogram or chest radiography, it does not provide a definitive diagnosis. According to American College of Cardiology and American Heart Association recommendations for congenital heart disease, if a continuous murmur is present, its origin should be determined by echocardiography, magnetic resonance imaging (MRI), computed tomography (CT), angiography, or cardiac catheterization [29]. Transoesophageal echocardiography (TEE) and transthoracic echocardiography (TTE) may be useful diagnostic tools for the detection of CAF because they may reveal their origin, course, and termination – particularly TEE, as well as cardiac morphology and function for the assessment of the haemodynamic significance of fistulae, exclusion of other cardiac abnormalities, and surveillance after therapy. However, most CAF cases require other imaging approaches, such as CT, MRI, invasive coronary angiography (ICA), or intravascular ultrasound (IVUS), to identify or confirm the diagnosis of CAF and provide more information regarding the anatomy and pathophysiology of the fistulae, which is helpful for planning their clinical management. Patients with small fistulae have a good prognosis without treatment. However, clinical monitoring with echocardiography every 3–5 years can be useful for patients with small, asymptomatic CAF to exclude the development of symptoms and enlargement and/or dysfunction of the cardiac chambers [20, 29]. The presence of symptoms, complications, and a significant shunt are the main indications for CAF closure techniques [20, 30]. There are 2 CAF closure modalities: surgical repair and transcatheter closure. Deciding between transcatheter closure and surgical intervention for CAFs can be challenging. Surgical intervention should be considered when there are other indications for cardiac surgery, including heart valve dysfunction as significant valve stenosis or regurgitation or multivessel coronary artery disease and low surgical risk [20, 31]. Transcatheter closure of CAF is feasible and should be considered in select patients. Some anatomical features of CAF may impact the success of transcatheter fistula closure and increase the risk of procedural complications, such as localization of fistula origin and termination region, number, magnitude, tortuosity of fistulae, and presence of a landing zone for device deployment without compromising the distal coronary artery. Transcatheter approaches may be challenging or unfeasible because of high fistulae tortuosity and the inability to deliver a catheter to the distal portion of the fistula to reduce the risk of device prolapse/migration and thrombus propagation [20, 21, 31, 32]. Several percutaneous closure techniques, primarily using occluders or coils, are available. The first report of a liquid embolic agent for successful percutaneous embolization of CAF was published by Tchantchaleishvili *et al.* in 2015 [33, 34]. Recanalization of the treated CAF may occur; therefore, imaging monitoring is necessary in these patients.

Procedural complications

The primary complications of percutaneous CAF closure include coronary dissection and myocardial infarction (MI). MI can manifest either at the time of the procedure or subsequently due to localized thrombosis and/or embolization of the coronary artery [19]. Nonetheless, consensus regarding anticoagulation or antiplatelet therapy after CAF closure remains elusive. In a study by Ilkay *et al*., patients were not routinely treated with oral anticoagulants or antiplatelet therapy post-procedure, and only 1 patient exhibited complete atrioventricular block; all patients remained asymptomatic, and no late complications or fatalities were recorded [35]. El-Sabawi *et al*. reported that patients were not routinely administered oral anticoagulants or antiplatelet therapy following the procedure; however, postprocedural MI occurred in 4 patients, with one occurring at 1 year, despite anticoagulation and aspirin use [32]. Shah *et al*. employed anticoagulation for patients with closed large/giant fistulae and dual antiplatelet therapy for moderate-sized fistulae [19]. After transcatheter CAF closure using vascular occlusion devices, Jiang *et al*. recommended the administration of oral aspirin (3 mg/kg daily) for 6 months to mitigate thromboembolic events following coil embolization [36]. Hou *et al*. routinely prescribed antiplatelet therapy with aspirin or dipyridamole for at least 6 months after the surgical management of CAF, except in cases of concomitant mechanical valve replacement, where only warfarin was prescribed [37]. Regarding the prevention of endocarditis, in accordance with the European Society of Cardiology guidelines, antibiotic prophylaxis is recommended for dental extractions, oral surgical procedures, and procedures requiring manipulation of the gingival or periapical region of the teeth in patients with a previous episode of infective endocarditis or those treated with surgery or transcatheter procedures involving postoperative palliative shunts, conduits, or other prostheses. Following surgical repair, and in the absence of residual defects, antibiotic prophylaxis is advocated solely for the initial 6 months following the procedure [38].

Conclusions

The management of CAF lacks uniform guidelines, leaving some fundamental questions unanswered.

However, CAF, while rare, is a potentially serious condition necessitating vigilant motoring and follow-up.

Conflict of interest

The authors declare no conflict of interest.

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