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Severe coronary artery ectasia and abdominal aortic aneurysm

Summary

Coronary artery ectasia (CAE), a discrete or fusiform arterial dilatation, is an uncommon angiographic finding. We report the case of a patient presenting with an acute coronary syndrome in whom further evaluation revealed coronary artery disease with severe CAE in the presence of an abdominal aortic aneurysm (AAA). Since both entities are strongly associated with local and systemic atherosclerosis, they have traditionally been viewed as a variant of atherosclerosis. The combined occurrence of CAE and AAA, as in the present case, raises questions about common pathogenetic mechanisms apart from atherosclerosis. The respective evidence will be reviewed.

Key words: coronary artery disease; coronary artery ectasia; aortic aneurysm

Zusammenfassung

Die Koronarektasie, eine umschriebene oder fusiforme Dilatation der Koronararterien, ist ein seltener angiographischer Befund. Wir berichten über einen Patienten, welcher sich mit einem akuten Koronarsyndrom präsentierte und bei dem die weitere Abklärung eine koronare Herzkrankheit mit schwerer Koronarektasie sowie ein abdominales Aortenaneurysma ergab. Da beide Entitäten mit lokaler und systemischer Arteriosklerose assoziiert

sind, werden sie meist als eine Variante der Arteriosklerose angesehen. Das gemeinsame Vorkommen einer Koronarektasie mit einem Aortenaneurysma, wie im vorliegenden Fall, wirft Fragen zu möglichen gemeinsamen pathogenetischen Mechanismen unabhängig von der Arteriosklerose auf. Die entsprechende Evidenz wird diskutiert.

Schlüsselwörter: koronare Herzkrankheit; akutes Koronarsyndrom; Koronarektasie; abdominales Aortenaneurysma

Introduction

Coronary artery ectasia (CAE) is defined as a discrete or fusiform arterial dilatation with a diameter of at least 1.5 times the diameter of an adjacent normal coronary segment [1]. CAE is an uncommon finding with estimates of its incidence varying from 1.2 to 4.9% in large angiographic series [1–4]. The risk of developing an abdominal aortic aneurysm (AAA) is 2–5% in the general population [5, 6]. We describe the case of a patient presenting with an acute coronary syndrome in whom further evaluation revealed coronary artery disease with severe CAE in the presence of an AAA. The combined occurrence of CAE and AAA, as in the present case, raises interesting questions about a common pathophysiological link. The respective evidence will be reviewed.

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Case report

A 72-year-old male patient was admitted to the emergency room because of acute chest pain present for five hours. Prior to this episode he had been free of cardiovascular symptoms. He has been treated for arterial hypertension and longstanding type 2 diabetes. His family history was positive for coronary artery disease and he had quit smoking 27 years ago.

On admission his heart rate was 114 per minute and his blood pressure 155/105 mm Hg. The heart sounds were normal, a rough 3/6 crescendo systolic murmur was heard best at the second right intercostal space. He had no peripheral edema, normal jugular veins and palpable peripheral pulses. The lungs were free on auscultation and oxygen saturation was 95% while breathing ambient air. The ECG showed a sinus rhythm with left anterior fascicular block, q-waves in the inferior leads, prominent R-waves in leads V₂–V₃ and mild ST-segment depression in leads V₃–V₅. The initial blood tests were remarkable for elevated levels of troponin I (0.85 ng/ml, normal <0.5), creatine kinase (267 U/l, normal <195), glucose (17.2 mmol/l), as well as total (5.5 mmol/l) and LDL-cholesterol (3.98 mmol/l). By echocardiography, left ventricular systolic function was mildly reduced with an ejection fraction of 45% due to inferoposterior akinesia. Moderate aortic stenosis with a mean gradient of 30 mm Hg and an aortic valve area of 0.9 cm² was also present.

The patient was taken to the coronary care unit with a diagnosis of an acute coronary syndrome with inferoposterior non ST-elevation myocardial infarction and treated with nitrates, platelet inhibitors, low molecular weight heparin, a betablocker and a statin. Creatine kinase rose to a maximum of 1865 U/l 22 hours after admission.

Coronary angiography showed three-vessel disease combined with severe segmental ectasia of all coronary arteries (fig. 1). No clear culprit lesion could be identified and it was assumed that embolisation from CAE in the right or left circumflex coronary artery could have caused the acute coronary syndrome. Abdominal aortography, performed because of difficult arterial access, demonstrated marked tortuosity of the iliac arteries and an AAA. Further examination by computed tomography confirmed an infrarenal, calcified aneurysm of nearly 6 cm in diameter (fig. 2).

With medical treatment the patient could be mobilised without complications and remained free of chest pain during the further hospital course. After thorough discussion with the patient and his family about the potential risks and benefits, it was decided not to pursue high risk interventional or surgical procedures at this point. During a follow-up of four months the patient did not experience recurrent cardiovascular symptoms with medical treatment.

Discussion

CAE and AAA are both disorders with pathological dilatation of the arterial system. An increased prevalence of CAE in patients with

AAA has been reported previously [7, 8], the inverse relation, as in the present case, has not been directly analysed.

Since both entities are strongly associated with local and systemic atherosclerosis, they

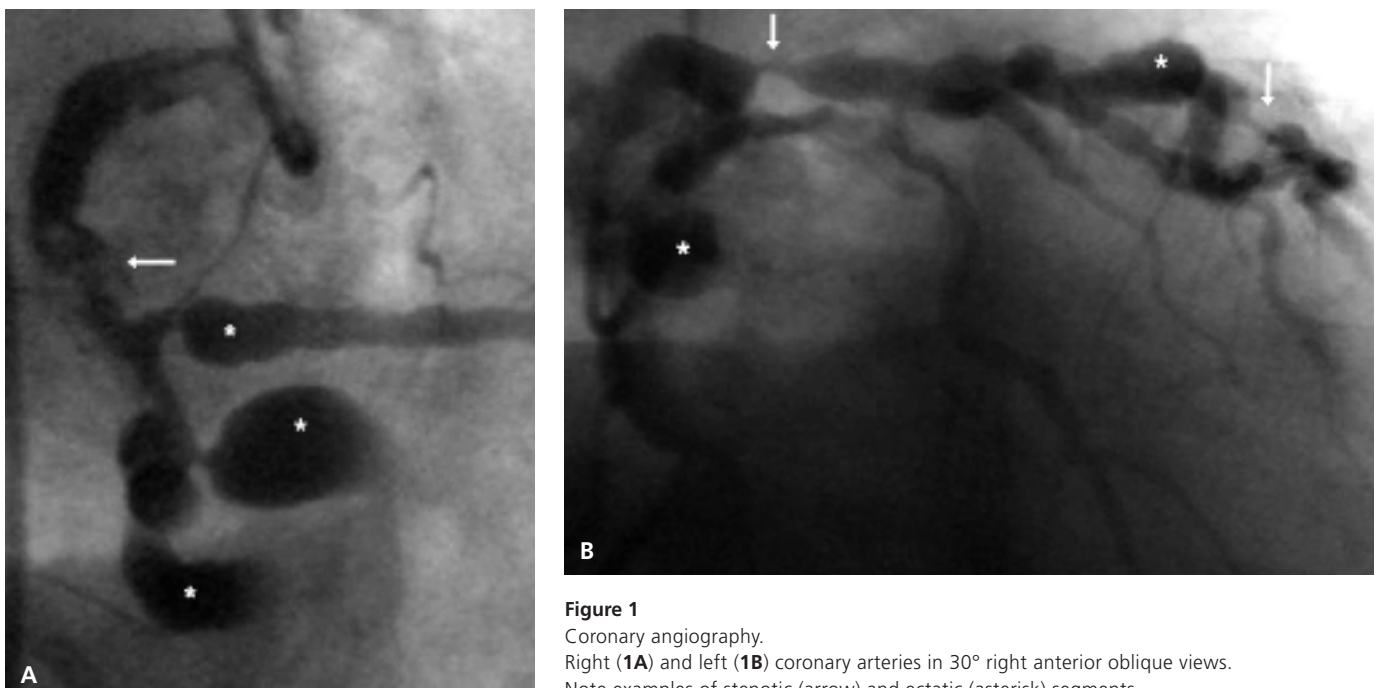


Figure 1

Coronary angiography.

Right (**1A**) and left (**1B**) coronary arteries in 30° right anterior oblique views.

Note examples of stenotic (arrow) and ectatic (asterisk) segments.

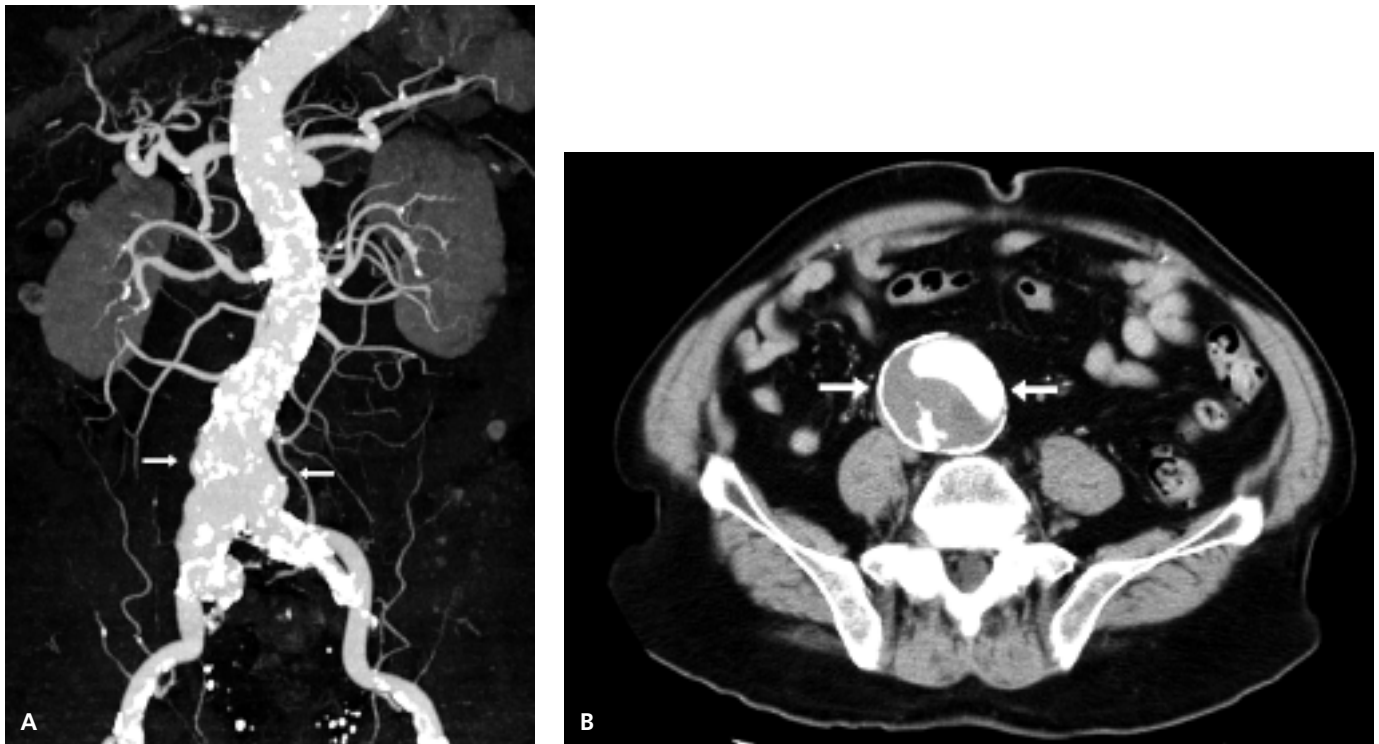


Figure 2
Abdominal computed tomography.

A Overview of the abdominal aorta and iliac arteries. Note extensive calcification of the vessel wall, an aneurysm just proximal to the aortic bifurcation (arrow) and tortuosity of the iliac arteries.

B Cross section at the level of the arrows in figure 2A showing a partly thrombosed aortic aneurysm with a diameter of 6 cm (arrow).

have traditionally been viewed as a variant of atherosclerosis [9, 10], but a clear causal relationship is not established. CAE and AAA share similar histologic characteristics with destruction of the musculoelastic elements of the tunica media [11, 12]. In contrast, atherosclerosis is primarily an endothelial disease, although thinning of the vascular media can occur in later stages [13]. CAE and AAA may thus be a variant of atherosclerosis with an exaggerated remodeling process or the result of additional pathogenetic factors involving primarily the media of the vessel wall. There is evidence to support both of these hypotheses. Atherosclerosis is considered an inflammatory disease in which immune mechanisms interact with metabolic risk factors to initiate and propagate lesions in the arterial circulation [14]. In the same line of evidence, chronic transmural inflammation is viewed as the primary pathophysiological process in AAA formation [15] and in some patients, CAE has been attributed to inflammatory or connective tissue disease. Inflammation of the aortic wall occurs through an unknown immunologic stimulus attracting inflammatory cells which release chemokines, cytokines and reactive oxygen species resulting in activation of proteases leading to medial degradation [16]. Among these proteases, matrix metalloproteinases seem to play a crucial role in vascular remodeling and atherogenesis [17]. Whether the same mechanisms are operative in CAE is

not known. However, elevation of inflammatory markers such as interleukin-6 has been shown in both AAA and CAE [18, 19]. Intrinsic defects of the media or systemic factors may predispose the media to form aneurysms. In various reports CAE has been described as isolated congenital lesion [20] or for instance in association with Ehlers-Danlos syndrome [21] pointing to weakness of elastin in the media. Prolonged exposure to herbicides containing acetylcholinesterase inhibitors has been postulated to be another cause of CAE [22]. Stimulation of nitric oxide by acetylcholine causes relaxation of vascular smooth muscle cells. Genetic susceptibility is an etiologic factor in both CEA and AAA [23, 24]. Interestingly, a low frequency of diabetes has been reported in AAA and CEA [4, 25]. It has been hypothesized that compensatory arterial enlargement in the course of the atherosclerotic remodeling process is impaired, among other factors, due to downregulation of matrix metalloproteinase production in diabetes [26, 27]. Diabetes thus could have a "protective" effect on aneurysmatosis because of an increased arterial stiffness. Sudhir et al. [28] have shown an increased prevalence of CEA in heterozygous familial hypercholesterolaemia. They proposed that structural weakening or active lysis of connective tissue elements in the arterial wall may result from interaction with LDL-cholesterol.

In summary, the presented evidence

suggests that, apart from atherosclerosis, a combination of additional genetic, environmental and endothelial factors may cause a destructive inflammatory process in the arterial wall of susceptible persons leading to CAE and AAA. Hopefully, these pathophysiological insights will lead to new treatment concepts in the future. To date, surgical or endovascular repair is the standard therapy for end-stage AAA, smoking cessation and control of hypertension may help to slow AAA growth. CAE seems not to be a benign condition, anecdotal evidence suggests that CAE may predispose to coronary thrombus formation, spasm, spontaneous dissection, angina pectoris, myocardial infarction and possibly sudden cardiac death. An increased mortality has been reported in some studies. However, no established management is available for CAE. In particular, there are no reports on the outcome of percutaneous coronary intervention or bypass surgery in patients with coronary artery disease and CAE.

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