Case report of an unusual and catastrophic presentation of coral reef aorta

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Background
Coral reef aorta (CRA) is a rare condition characterized by atherosclerosis and overt calcification of the aorta leading to severe luminal stenosis of the vessel. Most patients present with hypertension and intermittent claudication at the time of diagnosis. Risk factors associated with this condition are essentially the same as those associated with atherosclerosis. However, no unique condition seems to predispose an individual to develop CRA.

Case summary
We describe the case of a patient known for rheumatoid arthritis (RA) treated with long-term systemic corticosteroids who presented with a shock of unknown aetiology and left ventricular ejection fraction of 10%. Conventional and computed tomography angiography showed a CRA with subtotal lesion of the aortic arch that led to cardiogenic shock.

Discussion
Even though the exact aetiology of her condition will remain uncertain, RA and extended use of corticosteroids likely played a role in the development of this severe form of CRA.

Keywords
Atherosclerosis • Coral reef aorta • Shock • Corticosteroids • Rheumatoid arthritis • Case report

Introduction
Coral reef aorta (CRA) is a rare condition characterized by atherosclerosis and overt calcification of the aorta leading to severe luminal stenosis of the vessel. Qvarfordt et al.¹ first described it in 1984 and few cases have been reported in the literature since then. The presentation of this disease depends largely on the disease extent and on which aortic branches are involved. Most patients have hypertension and intermittent claudication at the time of diagnosis, but abdominal angina, renal dysfunction, anuria and lower extremity, and intra-abdominal thromboembolic events may also occur.² Risk factors associated with generalized atherosclerosis such as hypertension, tobacco use, and advanced age are well known, but no unique condition

Learning points
• Coral reef aorta (CRA) is a condition of extreme atherosclerosis and calcification of the aorta that can present with severe post-aortic valve obstruction.
• Rheumatoid arthritis and long-term use of corticosteroids are risk factors for atherosclerosis and can possibly play a role in the development of CRA.
seems to predispose an individual to develop CRA. However, women tend to be more affected than men in a ratio of 1.6:1. Definitive treatment is surgical and most commonly requires endarterectomy and/or bypass surgery.

Timeline

| Date       | Event Description                                                                 |
|------------|-----------------------------------------------------------------------------------|
| 04 July 2017 | Emergency consultation: patient with a shock of unknown etiology and stabilized with intravenous fluids and vasopressors and inotropes |
| 04 July 2017 | Echocardiography showed a left ventricular ejection fraction of 10% and moderate mitral regurgitation. Aorta was not well visualized on this modality |
| 08 July 2017 | Angiography showed a very calcified subtotal occlusive lesion of the aortic arch. It was not possible to pass a guidewire through the lesion and therefore not possible to image the coronary arteries as planned |
| 08 July 2017 | Computed tomography angiogram showed extensively calcified atheromatous aorta with a subtotal stenosis just distal from the left subclavian artery consistent with a diagnosis of medial reef aorta |
| 09 July 2017 | Patient palliated and death occurred after it was deemed impossible by the cardiovascular and thoracic surgeon to perform a bypass surgery |

Case presentation

We present a rare case of a 67-year-old woman who presented initially with left sided heart-failure and presumed cardiogenic shock. Cardiovascular risk factors included hypertension and dyslipidaemia. Her past medical history also included rheumatoid arthritis (RA), which had been treated with corticosteroids for the past 40 years. Additional medication included aspirin, diltiazem, hydrochlorothiazide, and atorvastatin.

On questioning, she reported two episodes of syncope during the last 3 months, both lasting less than a minute with a rapid return to normal state. More recently, she had noticed oedema in the lower extremities and a shortness of breath, for which she attended the emergency department. Physical exam showed hypotension at 82/58 mmHg, tachycardia at 152 b.p.m., peripheral vasoconstriction, and relatively cold extremities. Jugular vein distension was also noted at 7 cm H₂O above sternal angle. Cardiovascular exam was notable for a third heart sound, but did not show any murmur. Chest auscultation was normal. Moderate peripheral oedema was also noted.

Laboratory findings showed elevated lactate at 3.5 mmol/L (0.6–2.2), creatinine at 90 μmol/L (58–110)–patient’s baseline was 55 μmol/L, ALT at 1658 (0–37), AST at 1024 (0–32), total bilirubin at 75 μmol/L (2.8–17.0), and mildly elevated white blood cells at 14.8 × 10⁹/L (3.8–10.6) and neutrophils at 13.2 × 10⁹/L (1.4–6.6). Cardiac enzymes were marginally elevated with high-sensitivity troponin T at 64 ng/L (0–14). An electrocardiogram demonstrated sinus tachycardia, left ventricular hypertrophy using Cornell voltage criteria, and no ischaemic changes or Q waves. Transthoracic echocardiography performed upon arrival showed diffuse severe hypokinesia with a left ventricular ejection fraction (LVEF) of 10% and moderate mitral regurgitation. The aortic arch was not well seen because of patient morphology. Chest X-ray showed a right lower lobe opacity highly suspicious of pneumonia, cardiomegaly, and an atheromatous aorta. The working diagnosis after the emergency consultation was a mixed septic and cardiogenic shock with possible adrenal insufficiency.

The patient was treated with piperacillin/tazobactam 4.5 g intravenously every 8 h and ciprofloxacin 400 mg intravenously every 12 h to treat sepsis. Hydrocortisone 50 mg was administered intravenously every 8 h to combat any potential adrenal insufficiency from long-term steroid usage. Fluid resuscitation, inotropes, and vasopressors including norepinephrine 0.08 μg/kg/min and dobutamine 5 μg/kg/min were required to treat hypotension. Diuresis was achieved using furosemide 40 mg intravenously every 8 h.

Cardiac catheterization was deferred because of renal dysfunction, and there was only a mild elevation of troponin. Clinical improvement was witnessed for 48 h but sudden deterioration occurred on the third day of her hospital admission. Decision was made to proceed to diagnostic coronary angiography and left–right heart catheterization study.

A right femoral approach was selected for coronary angiography with placement of a 5 Fr sheath. A guidewire was advanced in the usual manner but resistance was felt in the aortic arch. This was impossible to overcome. Aortography was performed and revealed a subtotal aortic arch stenosis, with an extremely reduced blood flow through the stenosis. Therefore, a severe post-aortic valve obstruction was the likely explanation of the patient’s symptoms, and it was felt that trying to better define her coronary anatomy by using a right...
radial approach to perform coronary angiography would unnecessar-
ily delay the management of her condition.

A computed tomography (CT) angiogram of the aorta was per-
formed. This showed an extensively calcified atheromatous aorta
with a subtotal stenosis just distal from the left subclavian artery.
These radiologic features were consistent with a diagnosis of CRA.
The CT also showed marked calcification of the left coronary artery,
but stenosis quantification was not possible given that the image was
acquired without contrast.

After discussion with a cardiovascular and thoracic surgeon, no
surgery was possible for this patient because of the extent of vascular
calcification and the haemodynamic profile of the patient. It was then
decided with the patient and family to provide palliative care. When
active treatments were stopped, the patient rapidly deteriorated her
condition and died in the next 12 h.

Discussion

Coral reef aorta is a rare condition affecting between 0.6% and 1.8%
of the population. In a case series of 21 patients suffering from this
condition, 81% had hypertension, 57% were smokers with a mean of
32.2 package-years, 48% had dyslipidaemia, and 14% had type 2 dia-
etes. Although most patients with CRA have classic atherosclerosis
risk factors, the pathophysiology leading to the development of
such extensive calcification remains uncertain. The symptoms
and clinical presentation depend on the disease extent and
involvement of aortic branches. While the majority of patients
present with classic manifestations of peripheral artery disease,
acute kidney injury, and congestive heart failure have also been
reported in the literature.

This patient had RA that had been treated with prednisone mono
therapy for over 30 years. Rheumatoid vasculitis, a relatively rare con-
dition, can affect the aorta. This condition is difficult to discriminate
clinically from atherosclerosis. Furthermore, RA is associated with
significant inflammation and an elevated cardiovascular risk and
The use of corticosteroids in the treatment of RA further elevates the risk through a deleterious effect on blood pressure, as well as on lipid and glucose metabolism. The side effects of corticosteroids on cardiovascular risk appear to be dose related. In this patient, it is likely that a combination of RA associated inflammation, corticosteroid therapy, and elevated cardiovascular risk was responsible for the striking disease observed within the aorta.

The treatment of CRA is largely surgical. Transaortic endarterectomy is considered the treatment of choice, although thoracoabdominal bypass grafting, extra-anatomic bypasses, laparoscopic plaque removal, and aortic stenting have been published. The vast majority of patients undergoing surgical correction underwent treatment in an elective setting with a reported perioperative mortality of 8.7–11.6%. The only report of emergency aortic surgery was associated with death, Day 2 post-operative secondary to multi-system organ failure.

Ultimately, the exact aetiology of the severe calcification process affecting the aorta of this patient will remain uncertain in the absence of autopsy. Multiple factors may lead to extensive calcification of the aorta and CRA. In this patient, inflammation caused by RA and long-term corticosteroids may have played a role. The vast majority of patients with CRA do not have thoracic involvement and there is only one case in the literature of CRA leading to congestive heart failure with reduced LVEF. To our knowledge, our patient is only the second case of proximal CRA. It may also be the first case of CRA ending with heart failure with reduced LVEF and shock, but in the end, the definite cause of acute compromise and death will remain uncertain. A large myocardial infarction leading to severely depressed LVEF, although statistically much more common than a subtotal aortic stenosis, initially appeared to us less likely in the absence of markedly elevated troponins and compatible symptoms. Nonetheless, it remains a possibility that we have to strongly consider, and the aortic calcification might have been a bystander to the acute presentation given that it most likely has developed over a considerable period of time.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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