Case Report

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How acute renal failure led to the diagnosis of aortic coarctation

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The present case report focuses on a rare presentation of aortic coarctation. A 38-year-old man with well-controlled arterial hypertension, minimal change glomerulonephritis and colitis ulcerosa was suffering from recurrent acute renal failure episodes during viral gastroenteritis. No other symptoms at rest or during physical activity were present. The workup included renal duplex sonography, which unmasked tardus parvus profile in both kidneys without any acceleration of blood flow in the renal arteries. Further examination included CT angiography, which confirmed the diagnosis of aortic coarctation. The observed narrowing of the aorta measured 4 mm and was treated with percutaneous transluminal angioplasty and stent implantation (final diameter 12 mm). After the procedure, the patient had normal blood pressure values without the need of any medication; duplex sonography showed improved renal perfusion. The present case confirms the importance of evaluation for secondary hypertension and thorough workup of acute renal failure in young patients.

Keywords: acute renal failure, aortic coarctation, narrowing of aorta, renal perfusion, tardus parvus

Abbreviations: ACE, angiotensin-converting enzyme; CT, computer tomography; RAAS, renin–angiotensin–aldosterone system

INTRODUCTION

Coarctation of the aorta accounts for 5–8% of congenital heart anomalies [1–3]. It affects the descending aorta directly after the branching of the left subclavian artery. The length and position of the narrowed segment is diverse. Similarly, clinical presentation can vary from life-threatening conditions in neonates to hypertension in adults or no clinical significance at all [4]. Hypertension in aortic coarctation should not be underestimated as it reduces life expectancy significantly [5]. Adults with aortic coarctation have a higher incidence of heart failure, dissection of the aorta, and aortic rupture, which is related to sudden death [6]. Therefore, it is important to diagnose this condition as early as possible.

Here, we present a case of a 38-year-old man diagnosed with aortic coarctation after recurring episodes of gastroenteritis accompanied by severe acute renal failure.

CASE REPORT

A 38-year-old Caucasian man presented to our institution for follow-up after several episodes of acute renal failure. During presentation, he had a normal renal function with a creatinine of 0.78 mg/dl (69 μmol/l), whereas acute renal failure with a creatinine of up to 3.43 mg/dl (303.3 μmol/l) has been recorded during gastrointestinal tract infection. According to the medical history, acute renal failure developed during repetitive episodes of gastroenteritis caused by Norovirus. The patient had a long history of colitis ulcerosa treated with budenoside 3 mg three times a day. At presentation with gastroenteritis, no signs suggested a relapse of inflammatory bowel disease. In addition to that, he had well controlled arterial hypertension for the past 10 years treated with ramipril 10 mg/day and reported tendency towards tachycardia during exercise.

In 2003, the patient was diagnosed with minimal change glomerulopathy, a condition presenting with nephrotic syndrome due to podocyte foot processes effacement at the glomerular filtration barrier. In most cases, an underlying cause of minimal change glomerulopathy cannot be identified. However, causes may include entities such as drug abuse, malignancies, infections or autoimmune disorders. This condition can be associated with acute kidney failure and the need for dialysis because of renal hypoperfusion caused by intravascular volume depletion. Although uncommon, other histopathological changes such as renal interstitial oedema, glomerular lesions and renal tubular injury may contribute to renal impairment in this condition [7]. However, renal function usually recovers with treatment, as in the present case. In fact, the patient has

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been in remission since 2007. During current presentation, patient had normal serum albumin (4.6 g/dl) and total serum protein (7.72 g/dl) and no signs of proteinuria. The patient denied smoking and consumption of alcohol or recreational drugs.

The physical examination showed the patient to be in overall healthy condition with a BMI of 25.7 kg/m^2. At the presentation, blood pressure was 128/88 mmHg with no significant differences between left and right brachial measurement – brachial–ankle index was not measured at that time. There were no signs of a heart condition or pulmonary problems. His neurological examination was unremarkable.

Laboratory results showed normal serum electrolytes, no signs of infection, normal thyroid profile, and a normal haemoglobin level with 14.4 g/dl. Aldosterone was 22 ng/l and renin 2333 pg/ml while being on angiotensin-converting enzyme (ACE)-inhibitor therapy. Analysis of spot urine showed no signs of albuminuria and/or haematuria. The electrocardiogram was normal. Echocardiography showed normal systolic function with left ventricular hypertrophy (posterior wall measuring 13 mm, septum 11 mm) and the presence of a tricuspid aortic valve – no functional or morphological valve abnormalities were observed.

During the work-up, B-mode sonography showed normal and equally sized kidneys without any structural abnormalities. Duplex mode, however, revealed a 'tardus parvus' profile of intrasegmental arteries with resistive index (resistance index) ranging from 0.30 to 0.40 and increased acceleration time with no significant differences between the kidneys (Fig. 1a). Direct blood flow measurement in the renal arteries did not show any acceleration indicative of renal artery stenosis. These findings were indicative of a possible stenosis of the pre-renal aorta. Thoracic-abdominal computed tomography (CT) scan revealed a postductal aortic coarctation with pronounced development of collateral arteries from the subclavian, thoracic, intercostal and spinal arteries (Fig. 2a and b, Fig. 3a and b). Furthermore, an aneurysm of the ascending aorta measuring 4 cm in diameter was detected.

The patient received an angiography, which showed a trans-stenotic pressure gradient of 20 mmHg. The decision to correct the aortic coarctation was made, and the stenosis was corrected with Sinus XL stent measuring 22 mm × 80 mm. The procedure was performed without any complications. Dual platelet inhibition was initiated with clopidogrel 75 mg/day for 4 weeks and life-long acetylsalicylic acid therapy with 100 mg/day.

Following the intervention, the patient's blood pressure profile normalized and no further anti-hypertensive medication was required. Renal function remained normal with a creatinine of 0.82 mg/dl (72.5 μmol/l). Renin measurement was markedly reduced with 182 pg/ml, while aldosterone concentration was 65.6 ng/l. Three months later, CT angiography was repeated and confirmed a stable stent placement with improved aortic lumen diameter of 12 mm (pre-interventional diameter 4 mm) (Fig. 2c and d). Collateral arteries and the aneurysm of the ascending aorta were present and showed no differences to the previous examination. Duplex sonography of renal arteries demonstrated a significant improvement of resistance indexes (0.45–0.50 after vs. 0.30–0.40 prior to the procedure) (Fig. 1b). After stenting of the aorta, almost normal ankle–brachial indexes were measured (right 0.95, left 0.89).

DISCUSSION

The congenital narrowing of the aorta or aortic coarctation is one of the 10 most common anatomic heart abnormalities [3]. Though, most commonly it occurs near the ductus arteriosus, it may affect other parts of the descending aorta and vary in length as well as in narrowing of the aortic

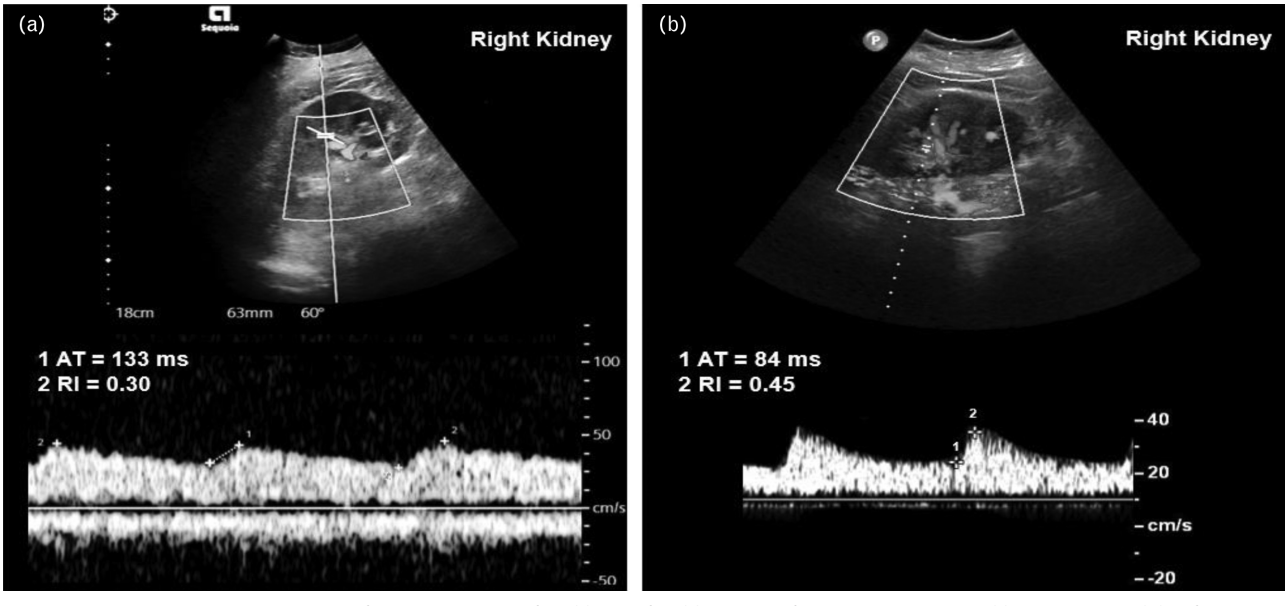

FIGURE 1 Exemplary duplex sonography images of the right kidney before (a) and after (b) stenting of the aortic coarctation. (a) Resistive index (RI) before stenting was 0.30 with an increased acceleration time (AT) of 133 ms. Without any direct findings of a renal artery stenosis in the sonography evaluation, this was indicative for a proximal stenosis of the aorta (e.g. aortic coarctation). (b) After stenting, resistive index increased to 0.45 and acceleration time was 84 ms.

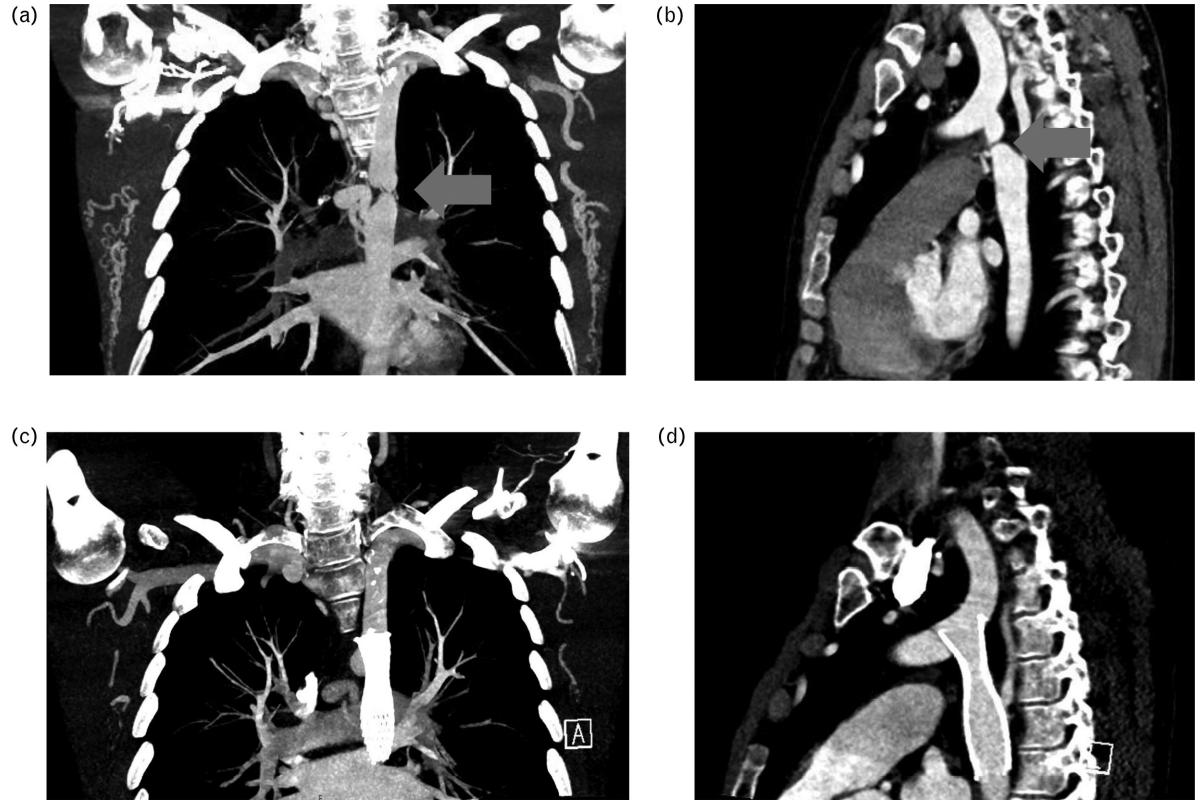

FIGURE 2 Contrast-enhanced computed tomography images of thoracic aorta before and after the treatment. (a) and (b) represent initial finding with narrowed postductal aorta segment as low as 4 mm. Red arrows present the affected part of aorta. (c) and (d) represent images at follow-up 3 months after stent placement procedure. At follow-up, an increased lumen diameter (12 mm) was observed.

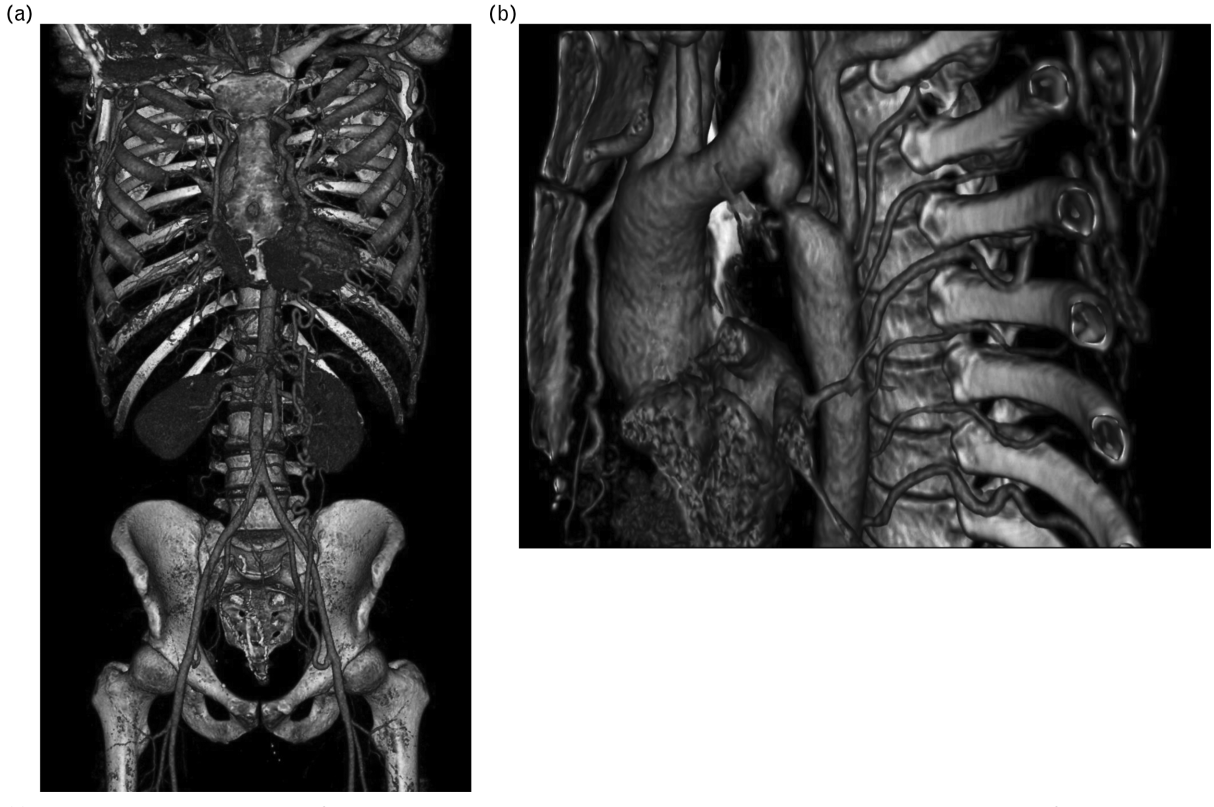

FIGURE 3 (a) Three-dimensional reconstruction of computed tomography angiography showing the collateral circulation in the presence of aortic coarctation. (b) Three-dimensional reconstruction of computer tomography with angiography showing the aortic coarctation.

lumen [2,5]. Coarctation of the aorta can be an isolated anomaly. However, it often presents in combination with other cardiac abnormalities such as bicuspid aortic valve, ventricular septum defects and transposition of the great arteries [8]. Although anatomic abnormalities differ between affected individuals, this condition shares similar pathophysiological features and sequelae of adaptive changes. First of all, the intraluminal pressure before the narrowed segment increases, causing dilation of the aortic arch and left ventricular hypertrophy over time [5]. To the same extent, any body part proximal to the stenosis is exposed to higher blood pressure whereas body parts distal to the stenosis are affected by lower blood pressure. The bodies' ability to compensate these haemodynamic changes affects how and when symptoms occur. Often, aortic coarctation is diagnosed directly after birth and is described as a life-threatening condition, thus, only 10.3% of patients are diagnosed in adulthood [9]. In adults, symptoms range from incidental findings to resistant hypertension. Unfortunately, there are still cases in which this anatomic abnormality is diagnosed after a major event such as rupture of an aortic aneurysm. Therefore, during evaluation of secondary hypertension, a precise step-by-step examination should be performed in order to exclude this rare, however, possibly life-threatening condition. Indeed, during physical examination discrepancy in blood pressure measurements between arms and legs, pulse absence in the lower limbs, colder extremities, or claudication can be observed [4,10]. The latter symptoms were not observed in our patient neither at rest nor during physical activity. The absence of significant clinical symptoms such as claudication might be associated with the abundant collateral vasculature in this patient.

One of the symptoms related to aortic coarctation is high blood pressure. In fact, our patient had a history of arterial hypertension for more than 10 years, which was well controlled with ACE-inhibition. Interestingly, there was no renal function deterioration after antihypertensive therapy with ACE-inhibitor was started. The latter and the absence of indicative symptoms have prevented the early recognition of aortic coarctation in our patient. However, recurrent acute renal failure during episodes of dehydration implied a pre-existing condition, which led to the described evaluation and discovery of the underlying cause.

As already mentioned, a decrease in lower body blood pressure in the presence of aortic coarctation may affect lower body part perfusion. Therefore, it can also impair renal blood flow. The clinical presentation may mimic bilateral renal artery stenosis [11]. Duplex sonography reveals low resistance indexes in intrasegmental arteries of both kidneys. However, increased velocity within the renal artery as the hallmark of renal artery stenosis is absent in this condition. Indeed, our patient had very low resistance indexes in both kidneys without any signs of accelerated blood flow in the renal arteries. Pioneer studies on coarctation of the aorta have shown that patients might present some degree of renal ischemia although renal function remains stable [12]. Our patient was taking ACE-inhibitor and as expected had high renin concentration. Low-normal aldosterone concentration in this case cannot exclude the presence of secondary aldosteronism. This condition leads to vasoconstriction of an efferent arteriole and ultrafiltration. Therefore, the activation of renin–angiotensin–aldosterone system (RAAS) might be one of the driving factors for the development of hypertension and maintenance of sufficient renal perfusion. If RAAS blockade is added to antihypertensive regime, acute renal failure might occur [13] as this medication cause dilation of efferent arteriole disrupting the autoregulatory mechanism. Deterioration of renal blood flow, for instance, in the case of bleeding, dehydration, or use of nonsteroidal inflammatory drugs further increases the risk of renal failure in the setting of aorta coarctation. Indeed, in the presented case, depletion of blood volume and ACE-inhibitor therapy during the episodes of gastroenteritis as well as reduced blood volume because of nephrotic syndrome in the presence of minimal change glomerulopathy were a possible cause for acute renal failure episodes. Interestingly, according to experimental studies, renal perfusion parameters are already affected in healthy individuals following changes in fluid status [14]. Therefore, in cases of aortic coarctation with decreased lower body perfusion, further decrease in circulating blood volume may aggravate acute renal failure. Similarly, this can also be observed in patients with bilateral renal artery stenosis [15]. It cannot be ruled out that aortic coarctation also affected intestinal perfusion in this patient and might have added to the susceptibility of gastrointestinal infections.

Duplex sonography of renal arteries can be indicative of anatomical abnormalities associated with the proximal aorta. Further evaluation includes electrocardiogram for signs of ventricular hypertrophy and echocardiography for signs of a bicuspid aortic valve. In the presence of morphological abnormalities of the aortic valve, a parasternal and suprasternal view with thorough examination of proximal aorta is indicated, especially in young hypertensive adults [5]. In cases of suspected abnormalities of the proximal aorta, radiological imaging should be performed with either MRI or CT angiography as in the presented case [16]. The gold standard for the diagnosis of aortic coarctation is angiography. Due to the invasive nature of this procedure, it should only be performed in combination with a possible treatment option of the abnormality.

Although aortic coarctation with little-to-no symptoms appears harmless at first sight, studies show that mortality rate for undiagnosed and untreated patients with this condition before the age of 50 is 75% [6]. These patients are prone to develop aortic aneurysm, which might lead to aortic dissection or rupture. Moreover, aortic coarctation is associated with congestive heart failure, endocarditis, myocardial infarction and intracranial bleeding [17]. Therefore, patients may benefit from a timely diagnosis and treatment of the narrowed segment. There are several treatment possibilities. In case of aortic coarctation in neonates and children, surgical treatment is the preferred choice [18]. The treatment approach in adults varies. There are several factors, which have to be addressed before the treatment. The pressure gradient over the aortic narrowing has to be evaluated and should be at least 20 mmHg. Moreover, the treatment is advised if pronounced collateral vasculature in radiographic imaging is present or if patients present with heart failure, hypertension or claudication [5]. Although in

some cases, surgical treatment is preferred, most adult patients receive a minimally invasive procedure with percutaneous transluminal angioplasty with or without stenting of the affected aortic segment [19]. The latter carries only 1% of peri-interventional risk [20].

Our patient has received minimally invasive correction of aortic coarctation with stent placement. Already 3 months later, he presented normotensive and did not require any antihypertensive medication, which significantly improved his quality of life. In order to prevent early complications or re-stenosis, patients should be monitored regularly. Of note, the re-stenosis rates after surgical treatment are relatively high and range between 20 and 30% [18]. Minimally invasive treatment for that matter achieves better results with restenosis rates around 5% [21]. In addition, patients should be controlled for re-occurrence of hypertension, early coronary heart disease and heart valve anomalies [5,17]. Therefore, life-long annual follow-up is mandatory.

Our case demonstrates that careful evaluation of patients with (excessive) renal failure should be performed – ideally by a nephrologist. Although B-mode sonography is commonly used to evaluate kidneys during and after renal failure, duplex sonography might not be performed regularly. However, it is an easy and accessible tool to evaluate renal artery stenosis in hypertensive patients and it can be indicative of a proximal aortic abnormality, as shown here. Therefore, we recommend that duplex sonography should always be performed in patients with renal failure.

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Conflicts of interest

There are no conflicts of interest.

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