

## Incidental Leriche Syndrome in Horseshoe Kidney Disease: A Non-Classic Couple

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### INTRODUCTION

Atherosclerotic disease is the leading cause of death in the U.S.<sup>1</sup> Leriche syndrome (LS) is a rare variant of the atherosclerotic occlusive disease, also known as aortoiliac occlusive disease, because it commonly produces total occlusion of the aorta, below renal arteries, and/or both iliac arteries.<sup>2</sup> LS was first mentioned by Robert Graham of London in 1814 and named by René Leriche, a famous French surgeon in 1923.<sup>3</sup>

LS risk factors are hypertension, hyperlipidemia, smoking, and diabetes.<sup>4</sup> It usually presents as a late atherosclerotic disease due to reduced blood flow to the renal artery, producing significant stenosis with accelerated hypertension or acute renal injury.<sup>5</sup> It also could affect the iliac vessels producing a wide range of manifestations including: sexual dysfunction, intermittent claudication of lower limbs, bilateral buttock claudication, and absent femoral arterial pulses,<sup>6</sup> especially when the occlusion is greater than 50% of the arterial lumen.<sup>7</sup>

Horseshoe kidney disease (HSK) is defined as a fused renal parenchyma with uncrossed ureteral systems,<sup>8</sup> and even though it is mostly benign, abnormal urine drainage predisposes to urologic diseases,<sup>9</sup> including hydronephrosis, nephrolithiasis, urinary tract infections, chronic abdominal pain, and urogenital cancer.<sup>10,11</sup> HSK has an incidence of 1/600 and male predominance 2:1.<sup>12</sup>

Both conditions, although rare causes of acute abdominal pain, are potentially fatal and should be suspected due to the concomitant risk factors in this scenario. We present a case report describing a patient with abdominal pain and progressive intermittent claudication. Due to the physical findings, altered blood-urine panel, and antecedent of horseshoe kidney disease, hospital admission and imaging were ordered.

### CASE REPORT

A 61-year-old Latin American man presented to the emergency room complaining of progressively severe abdominal pain. His medical history included poorly controlled hypertension (HTN), type 2 diabetes mellitus, recurrent urinary tract infections, and horseshoe kidney. He also reported a 40 pack-year smoking history. The patient also complained of dysuria, and two months progressive lower limb pain walking short distances.

Vital signs included: blood pressure 120/80 mmHg, heart rate 116

bpm, temperature 36° C, oxygen saturation 98%, respiratory rate 26 rpm, and capillary refill 2 seconds. Physical examination revealed pallor skin, hypogastric tenderness but no guarding or rebound sign, and percussion elicited pain over the left flank; no lower limb edema was found. Initial blood workup showed: hemoglobin 14.9 g/dl, hematocrit 44.9%, glucose 149 mg/dl, creatinine 1.6 mg/dL, urea 34 mg/dl, total cholesterol 185 mg/dl (LDL 135.5 mg/dl), elevated transaminases (ALT 134 U/L, AST 207 U/L), and potassium 5.3 mEq/L. Urinalysis revealed abundant bacteria and leukocytes. Abdominal x-rays showed bilateral kidney stones (Figure 1).

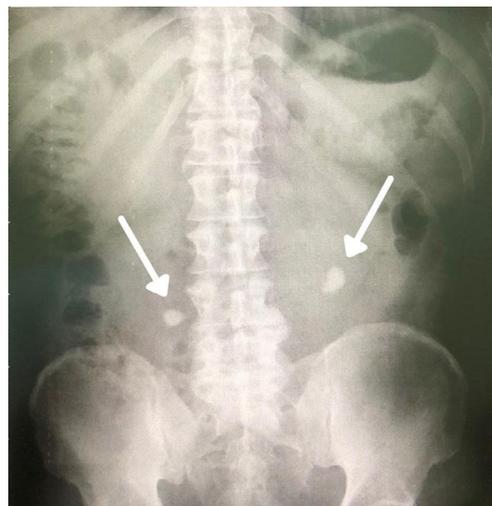


Figure 1. Abdominal x-ray showing bilateral kidney stones.

The patient was admitted for hospitalization under the clinical suspicion of acute pyelonephritis. Treatment with broad spectrum antibiotics was started. Bedside electrocardiogram showed atrial fibrillation and images suggestive of prior myocardial infarction. Amiodarone 200 mg/day for rhythm control and aspirin 100 mg/day as antiplatelet therapy were started.

The urology service was consulted due to nephrolithiasis and a possible lithotripsy procedure. Contrast abdominal computed tomography (CT) and a renal angiography were ordered, and horseshoe kidney and atherosclerotic plaques occluding arterial lumen were found (Figures 2, 3, and 4).



Figure 2. Contrast abdominal CT showing the horseshoe kidney disease.

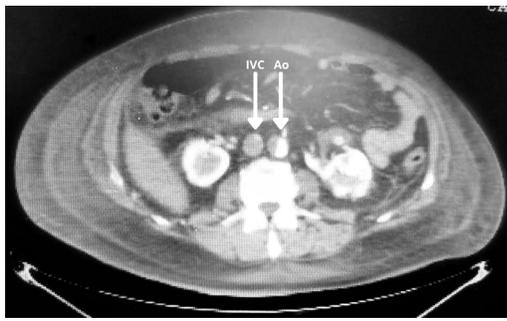


Figure 3. Contrast abdominal CT showing 50% occlusion of abdominal aorta below the renal arteries. IVC: inferior vena cava, Ao: Aorta.

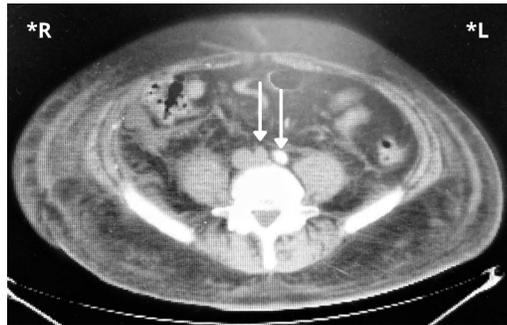


Figure 4. Contrast abdominal CT showing full occlusion of right common iliac artery (right side arrow) and the presence of a minor plaque within the left common iliac artery (left side arrow). R: right, L: left.

Later, the patient developed shortness of breath, fever, and productive cough. RT-PCR for SARS-CoV-2 and blood cultures were negative. Chest CT demonstrated alveolar infiltrates, pleural effusion, and a thrombus allocated in the right pulmonary artery. Transthoracic echocardiography revealed left ventricular ejection fraction of 27%, left-sided dilation, and pulmonary hypertension (43 mmHg). Low-molecular-weight heparin was started.

During the following days, the patient's respiratory symptoms markedly improved, but he frequently complained of nocturnal right leg pain. We identified edema in the right leg and no peripheral pulses. The patient died shortly after due to a massive aortic thromboembolic event.

## DISCUSSION

Our patient complained of intermittent claudication for months but sought no medical opinion, and 24 hours prior to his death, physical examination revealed absent arterial pulses in the lower limbs. Both findings were a consequence of complete arterial obstruction caused by LS.

Mortality risk was considered in the patient, since 63% of intermittent claudication cases have a 10-year mortality trend.<sup>13</sup> Death has been reported as a consequence of thrombosis of the ascending aorta.<sup>14</sup> Other systemic complications following systemic embolization included sudden vision loss due to acute retinal artery occlusion,<sup>7</sup> ischemic stroke, and skin necrosis,<sup>15</sup> which our patient did not present during hospitalization.

Although conventional angiography is considered the gold standard for evaluating LS, noninvasive imaging techniques can identify vascular anatomy with great accuracy and increasingly have become the first choice technique, especially computed tomography angiography (CTA) with three-dimensional (3D) reconstruction which provides more information about distal permeability.<sup>16</sup> In our case, a contrast abdominal CT was clear enough to demonstrate the vascular occlusion,

even though it was accidental. Regarding LS treatment, aortobifemoral bypass has shown better long-term results but in the short-term endovascular reperfusion may be more advantageous,<sup>17</sup> which our patient could not undergo due to his hemodynamic-respiratory instability.

HSK often presents with nephrolithiasis, urinary tract infections, and chronic abdominal pain,<sup>10,11</sup> such as in our patient. Among the pediatric population, abdominal pain and urinary tract infections are also the most common manifestations.<sup>18</sup> Renovascular HTN in the setting of HSK is unusual,<sup>19</sup> but HTN might accelerate atherosclerotic disease, since it is a risk factor for vasculopathy and our patient had controlled poorly.

As a part of the approach for horseshoe kidney, imaging to analyze kidney anatomy and its vessels were made for our patient, showing the classic presentation of fusion of the kidney's inferior poles,<sup>10</sup> and nephrolithiasis bilaterally. Diagnosis usually is made accidentally when performing noninvasive imaging for another reason.<sup>10</sup> Surgical treatment has been used in these patients, such as shockwave lithotripsy, ureteroscopy, percutaneous nephrolithotomy, and laparoscopy.<sup>20</sup> Lithotripsy was a possible plan due to nephrolithiasis, but aortoiliac atherosclerotic plaque came across in imaging studies.

## CONCLUSIONS

Patients' comorbidities and cardiovascular risk factors may accelerate the development of atherosclerotic disease.<sup>21</sup> Intermittent claudication is a first indicator of peripheral occlusive arterial disease, but it may be overlooked during anamnesis. In our case, the use of contrast abdominal computed tomography was useful to diagnose Leriche Syndrome. HSK, despite being a rare condition, predisposes nephrolithiasis and urinary tract infections,<sup>10</sup> which are common diseases.<sup>22,23</sup> Imaging studies in a patient with acute abdominal pain should be analyzed thoroughly, focusing not only on solid organs but also in small vascular details to avoid missing rare and potentially deadly syndromes.

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