Reversible acute renal failure in a middle aged woman secondary to intravascular hemolysis caused by favism

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Case Presentation
A 58-year-old woman was referred with the complaints of nausea, vomiting and left flank pain since last night. She had a history of renal colic during previous years and in this presentation, she received medication for relieving the pain. She was discharged from the clinic with analgesic drugs and intravenous (IV) line therapy with normal saline. Urine analysis revealed, +1 blood and 4-5 red blood cells (RBCs) in microscopic urine sediment. When she presented to us, she was anuric and had severe pain in the left flank. She had a history of favism after exposure to beans a few years ago. She had consumed some food containing beans 2 days ago. On admission, she had severe left flank pain and anuria of 12 hours duration. She had dyspnea and on physical examination had end-expiratory crackles in the base of both lungs. The relevant laboratory tests on the day of admission and several days thereafter are summarized in Table 1. Her peripheral blood film showed polychromasia, anisocytosis, poikilocytosis and blister cells.

The viral and autoimmune serology was negative. On abdominal ultrasonography, the size of the right kidney was 105 mm and that of the left, 128 mm. No stone or hydronephrosis was seen. Doppler ultrasonography of renal vessels (arterial and venous) was done and normal finding was reported.

The patient’s renal functions continued to deteriorate and hemodialysis was started for ameliorating her symptoms. Her symptoms progressed and daily dialysis was done. Meanwhile, at the end of first week of admission, percutaneous renal biopsy was also performed, which is shown in Figure 1.

After three dialysis sessions, the renal function tests showed improving trend with a serum creatinine of 2 mg/dL on 12th day of admission, when she was discharged.

Given the past history of favism and the current clinical and laboratory features favoring this diagnosis, red cell enzymatic analysis for glucose 6 phosphate dehydrogenase (G6PD) levels was contemplated but not performed due to the acute presentation. However, she was advised to undergo the same after a suitable interval.

Questions
1. What do the laboratory investigations indicate?
2. What are the renal biopsy findings?
3. What is the diagnosis?
4. What is the course and prognosis of the condition?

Table 1. The results of the main laboratory investigations of the patient on the day of admission and for seven days thereafter

<table>
<thead>
<tr>
<th></th>
<th>On admission</th>
<th>Day 2</th>
<th>Day 3</th>
<th>Day 4</th>
<th>Day 5</th>
<th>Day 6</th>
<th>Day 7</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood urea nitrogen (mg/dL)</td>
<td>26</td>
<td>44</td>
<td>66</td>
<td>101</td>
<td>123</td>
<td>129</td>
<td>97</td>
</tr>
<tr>
<td>Creatinine (mg/dL)</td>
<td>2.6</td>
<td>5.5</td>
<td>6</td>
<td>8.3</td>
<td>9.4</td>
<td>10.1</td>
<td>9.4</td>
</tr>
<tr>
<td>Creatine phosphokinase (U/L)</td>
<td>161</td>
<td>122</td>
<td>145</td>
<td>123</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Lactate dehydrogenase (U/L)</td>
<td>3600</td>
<td>6760</td>
<td>5987</td>
<td>4271</td>
<td>3992</td>
<td>3060</td>
<td>2841</td>
</tr>
<tr>
<td>Total bilirubin (mg/dL)</td>
<td>1.2</td>
<td>1.1</td>
<td>0.7</td>
<td>0.5</td>
<td>0.5</td>
<td>0.7</td>
<td>0.5</td>
</tr>
<tr>
<td>Aspartate transaminase (IU/L)</td>
<td>212</td>
<td>382</td>
<td>209</td>
<td>65</td>
<td>33</td>
<td>29</td>
<td>17</td>
</tr>
<tr>
<td>Alanine aminotransaminase (IU/L)</td>
<td>127</td>
<td>286</td>
<td>239</td>
<td>173</td>
<td>114</td>
<td>94</td>
<td>52</td>
</tr>
<tr>
<td>Hemoglobin (g/dL)</td>
<td>10.5</td>
<td>9</td>
<td>8.4</td>
<td>7.9</td>
<td>8.5</td>
<td>7.5</td>
<td>8.7</td>
</tr>
<tr>
<td>Platelet count (/mm³)</td>
<td>235000</td>
<td>200000</td>
<td>137000</td>
<td>136000</td>
<td>156000</td>
<td>139000</td>
<td>192000</td>
</tr>
</tbody>
</table>

Received: 4 July 2020, Accepted: 17 August 2020, ePublished: 10 September 2020

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3. Pigment cast associated nephropathy secondary to favism.

4. Favism denotes the disease caused by the ingestion of broad beans (Vicia faba) in persons with deficiency of G6PD enzyme in the red blood cells (1-4). Because G6PD deficiency is an X linked disorder, the main clinical manifestations are seen in hemizygous males. However, in areas of high prevalence of the disorder, homozygous females can be affected as severely as males. In the heterozygous females, the disease is often milder or goes unnoticed. It can present in atypical forms also. Red cell enzyme studies during the hemolytic phase may yield normal enzyme levels. The overall prognosis of the condition is excellent (1-4).

5. The patient should be counseled about the possible precipitating factors for hemolysis, such as drugs like sulfonamides and quinolones and fava beans and to avoid these in future (3).

Authors’ contribution
All authors contributed equally to the conception and study designing, collection and interpretation of data, drafting of paper and its final approval.

Conflicts of interest
The authors declared no competing interests.

Ethical considerations
Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the author.

Funding/Support
None.

References

What advise would you give to the patient regarding prevention of such episodes in future?

Answers
1. The laboratory indices show acute kidney injury, hepatocyte damage and hemolytic anemia. There is gradual increase in blood urea and serum creatinine till dialysis was started. Liver enzymes were also elevated during first few days of admission but normalized within one week. Hepatitis virus workup was negative. There was a drop in hemoglobin but it started to rise from one week of admission. No blood transfusion was given. Platelet count remained in normal range, albeit with a transient decline in between.

2. Renal biopsy contained both cortex and medulla with upto 16 glomeruli showing minor changes. The vessels were unremarkable. The main pathology was seen in the tubulointerstitial compartment. There is moderate to severe acute tubular injury associated with interstitial edema and mild, focal interstitial inflammation. In addition, many tubular lumina contain pigment casts. These casts are bright red in colour and granular in appearance and they stain bright red with Masson's trichrome stain (Figures 1 to 4).

Figure 1. (A) Low-power view showing many normal looking glomeruli, acute tubular injury, and pigment casts in many tubular lumena. (Hematoxylin and Eosin, x100). (B). Medium-power view showing one normal looking glomerulus, acute tubular injury, interstitial edema, and pigment casts in many tubular lumena. (Hematoxylin and Eosin, x200). (C) High-power view showing one normal looking glomerulus, acute tubular injury, interstitial edema, and pigment casts in one tubular lumen. (Hematoxylin and Eosin, x400). (D) Medium-power view showing one normal looking glomerulus, acute tubular injury, interstitial edema, and pigment casts staining intense red on Masson’s stain. (Masson’s trichrome, x200).