Hepatopulmonary syndrome associated with schistosomal liver disease

Mohamed Al-Moamary MRCP, Ibrahim Altraif MRCP(UK)

Hepatopulmonary syndrome (HPS) is the triad of chronic liver dysfunction, abnormal arterial oxygenation and pulmonary vascular shunting (1). It is estimated to occur in 8% to 25% of liver transplant candidates (2-9). There are no reports of HPS associated with schistosomal liver disease in the literature. We report the first such case of HPS complicating schistosomal liver disease.

CASE PRESENTATION
A 55-year-old man known to have biopsy-proven hepatic fibrosis secondary to previously treated schistosoma mansoni-related liver disease. He presented in 1988 with shortness of breath, orthodeoxia, platypnea, cyanosis, marked digital clubbing and liver failure. Extensive investigation revealed no other etiology for liver disease apart from schistosomiasis. The diagnosis of hepatopulmonary syndrome was based on clinical grounds, as well as abnormal arterial blood gases and positive contrast echocardiography. The patient underwent orthotopic liver transplantation, which was initially successful, but then died of respiratory complications and multi-organ failure on day 42 post-transplant. To the authors’ knowledge this is the first report of hepatopulmonary syndrome associated with schistosomal liver disease.

Key Words: Hepatopulmonary syndrome, Schistosomal liver disease

Syndrome hépato-pulmonaire associé à la maladie hépatique schistosomique

RÉSUMÉ : Un homme de 55 ans atteint de maladie hépatique schistosomique se présente pour essoufflement, hypoxémie orthostatique, dyspnée orthostatique, cyanose, hippocratisme marqué et insuffisance hépatique. L'examen approfondi n'a révélé aucune autre étiologie de la maladie hépatique que la schistosomiasis. Le diagnostic de syndrome hépato-pulmonaire a été établi sur la base des signes cliniques, des gaz artériels anormaux et d'une échocardiographie de contraste positive. Le patient a subi une transplantation hépatique orthotopique qui a été couronnée de succès au début, mais le patient est décédé de complications respiratoires et d'insuffisance pluri-organique 42 jours après la transplantation. À la connaissance des auteurs, il s'agit du premier rapport de syndrome hépato-pulmonaire associé à une maladie hépatique schistosomique.
TABLE 1
Arterial blood gas results

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Room air</th>
<th>100% oxygen</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Supine</td>
<td>Standing</td>
</tr>
<tr>
<td>pH</td>
<td>7.4</td>
<td>7.44</td>
</tr>
<tr>
<td>PaCO₂</td>
<td>30</td>
<td>30</td>
</tr>
<tr>
<td>PaO₂</td>
<td>61</td>
<td>51</td>
</tr>
<tr>
<td>Bicarbonate</td>
<td>20</td>
<td>20</td>
</tr>
<tr>
<td>Oxygen saturation</td>
<td>93%</td>
<td>88%</td>
</tr>
<tr>
<td>Arterial-venous difference</td>
<td>48</td>
<td>–</td>
</tr>
</tbody>
</table>

Various liver diseases have been associated with this syndrome, and the presence of intrapulmonary shunt has been shown to occur in schistosomal cor pulmonale without liver failure (6). To our knowledge this is the first report of HPS associated with liver failure due to schistosomal liver disease without cor pulmonale.

Clinically the combination of platypnea, digital clubbing and spider angioma in the setting of chronic liver disease is suggestive of HPS. This should be supported by the presence of hypoxemia and orthodeoxia, a decrease of the partial arterial oxygen concentration by 10% upon standing (7).

It is essential to establish the diagnosis of HPS. For practical purposes, contrast enhanced echocardiogram appears to be the most sensitive diagnostic test to detect intrapulmonary vascular dilatation. ⁹⁹mTc microaggregated albumin scan and pulmonary angiography might be needed to confirm the diagnosis in certain cases (7-9).

The mechanism of HPS in schistosomal liver disease is probably not different from other etiologies of chronic liver dysfunction associated with HPS, but this is speculative because the mechanism of HPS is unknown. Until recently HPS was considered a contraindication for liver transplantation. However, the accumulated data have shown that HPS does not contraindicate surgery as long as intra- and post-operative oxygenation is maintained. Furthermore, severe hypoxemia has been proposed as an indication for liver transplantation (9). Krowka and Cortese (9) showed that 13 of 15 cases with HPS were transplanted successfully, and two of 15 cases had the procedure mainly for severe oxygen desaturation. This review showed resolution of hypoxemia in all but one patient who required arterial embolization.

In the case presented, severe hypoxemia was associated with HPS in a patient with schistosomal liver disease. This case demonstrates that schistosomiasis can be associated with HPS.

ACKNOWLEDGEMENTS: We thank Dr Sulaiman Al Majed for his critical comments, and Kay Finley and Jill Taylor for their expertise in typing the manuscript.

REFERENCES