Jaundice In Patient With Myelofibrosis

Wolfson Medical Center
Gastroenterology Department
Dr. Vera Dreizin
The case presented

- Born 1930, 1+1, she arrived in Israel at the age of 15 years.
- She lives with her daughter, independent in daily work.

Background:
- Myelofibrosis since 1999. Last BMB 2008
- Hypothyroidism
- Hysterectomy
- Osteoporosis
- Relapsing polychondritis

Treatment:
Eltroxin, Lanton, Fosalan, Prednison: ⚠️

Brach: ⚠️
הצגת מאקרה

ב 09/04/2009-21:12 אושפזה עקבlek דלקת סמפונות חירפת, חוס נמרץ,بونוходит צויר ריאטי מונגרו בצלום חודש.
שופלה ב-Augmentin, ואיינפליציתות. שחררה בצמצר כליל טוב.
שוחרה בצמצר כליל טוב.
ב 09/05.07 אושפזה שוב עקב דלקת הדרדרות בمعنى הכליל, חולה,
בצקת היקפיות והחדרות האמוקה.
הצגת מקרה

赈ית מקורה

- בצורתה - בתוכה מלאת, ללא חום, לחם דם 140/80, דופק 120
- סדיר, צהוב עמידה בלהמיות ובעור.
- משקל 38 ק"ג, גובה 153 ס"מ.
- ריאות - נקיות, קולות לב סדירים, אוזנה סיסטובית 6/2 בוחד.
- הצלב.
- בטן רך, רגישות דיפוזית בכל הל gratuito, ללא סימני גירוי צפתי, בכדי
- נמוך כ - 5 ס"מ מתוח לקשה הצלובות, סחוס נמוך בצלובות.
- בצקת גומתית בשוכרים.
<table>
<thead>
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<th>01.04.09</th>
<th>12.04.09</th>
<th>22.04.09</th>
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<th>08.05.09</th>
<th>09.05.09</th>
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<tbody>
<tr>
<td>Hb</td>
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<td>14.6</td>
<td>13.8</td>
<td>13.8</td>
<td>14.3</td>
<td>14.6</td>
<td>15.5</td>
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<tr>
<td>PLT</td>
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<td>159</td>
<td>138</td>
<td>6</td>
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<td>3</td>
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<td>WBC/NEUTR</td>
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<td>31.3/21</td>
<td>25.7/17.3</td>
<td>72.2/31.4</td>
<td>56.3/33.4</td>
<td>79.1/15.3</td>
<td>85.8/37.6</td>
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<tr>
<td>T.BILIRUBIN</td>
<td>0.84</td>
<td>0.8</td>
<td>1.2</td>
<td>11.85/7</td>
<td>16.2/8.71</td>
<td>23.9/12.3</td>
<td>29.4/15.6</td>
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<tr>
<td>ALP</td>
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<td>106</td>
<td>218</td>
<td>527</td>
<td>622</td>
<td>602</td>
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<tr>
<td>ALT</td>
<td>14</td>
<td>18</td>
<td>28</td>
<td>133</td>
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<td>130</td>
<td>89</td>
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<tr>
<td>AST</td>
<td>21</td>
<td>22</td>
<td>37</td>
<td>122</td>
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<tr>
<td>ALBUMIN</td>
<td>3.69</td>
<td>3.18</td>
<td>2.9</td>
<td>2.31</td>
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<tr>
<td>CREATININ</td>
<td>0.75</td>
<td>0.8</td>
<td>0.62</td>
<td>0.61</td>
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<td>PTT/INR</td>
<td></td>
<td></td>
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<td>40.7/1.46</td>
<td>45.7/1.7</td>
<td>53.5/2.06</td>
<td></td>
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<tr>
<td>FACTOR v</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>59%</td>
<td>72%</td>
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<tr>
<td>FERRITIN</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>3340</td>
</tr>
<tr>
<td>FIBRINOGEN</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td>83.7</td>
</tr>
</tbody>
</table>
הצגת מקרה

בדיקות נוספות:

- HBV, HCV, HAV, CMV, EBV שלילי
- AMA, ASMA, ANA שלילי
- בדיקת חזה ובטן: עים פלואורלי בכסיס הא-basket, ובו מוגדל עצם הקשיות בבסיס הא-basketidental
- בדיקת CT חזה ובטן: עין פלואורלי במגנט החזה, ובו מוגדל עצם הקשיות בבסיס הא-basketidental
- לבד 24 ס"מ מהגן עד האגן הקשיות בסביבת היפודנсимוס קטעים
- המרובים מפורים enough. טחול מוגדל מעט, ללא מימיתولة
- עצות לשחרۇת ושכרולים
האישה נפטרה ב 10/05/2009 – כ – 3 ימים לאחר אשפוזה
悝ום המקרה

בת 78 עם מיוגליפרוציות ברקע שאושפזה בתמונה של דלקת חירפה בסמפונות, טופלה בעצרת אנטיביוטיקה ופרדניזון, ושוחררה לביתה לאחר שבוע של אשפוז. אושפזה שוב כשבועיים בתמונה של צהבת, פטוס ספלנומגליה. בבדיקת CT בטן הודגמו נגעים קטנים ומרובים בכבד. צהבת הבינה קלינת מעבדתית של צהבת, הפוט ספלנומגליה. כמשר בבדיקת בטן הודגמו נגעי קטייס ומורובים בכבד.
הצגת מקרה
Postmortem liver biopsy:

A piece of liver tissue with granulomas in multiple areas, surrounded by a palisading mass and the appearance of a large number of Langhans type cells.

In the Ziehl Neelsen + PAS stain, there were a large number of acid-stable tags.

No fungi were found.
Liver and Tuberculosis

- Primary hepatic tuberculosis
- Hepatic injury by hepatotoxic anti-Tb drugs
- Tuberculosis that develops in patients with chronic liver disease
Primary hepatic tuberculosis
Primary Liver Tuberculosis

- TB is a worldwide health problem with a high prevalence in developing countries.

- During the late 1980s and early 1990s, the number of reported cases of TB increased in developed countries and those increases were largely due to immigration from countries with a high prevalence of TB, drug abuse and infection with HIV.

- Abdominal TB is uncommon, comprising 3.5% of extra pulmonary TB. Hepatic TB is considered very rare among abdominal TB patients.
Hepatic TB is encountered more frequently in Asian countries.

Hepatic involvement was found clinically in 50-80% of all patients dying from pulmonary TB and in up to 91% on autopsy.

There are many case reports of hepatic TB mimicking other conditions with hepatic and non-hepatic manifestation.

M:F ratio is 2:1

Age range 11-50 years
Primary hepatic tuberculosis – con.

- **Presenting common symptoms and sings**

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RUQ pain</td>
<td>65-87%</td>
</tr>
<tr>
<td>Non-specific symptoms (fever, anorexia, weight loss)</td>
<td>55-90%</td>
</tr>
<tr>
<td>Non specific pain abdomen</td>
<td>50%</td>
</tr>
<tr>
<td>Jaundice</td>
<td>20-35%</td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>70-96%</td>
</tr>
<tr>
<td>Slenomegaly</td>
<td>25-55%</td>
</tr>
</tbody>
</table>
Primary hepatic tuberculosis – cont.

Hepatic tuberculosis presents in three forms.

- Diffuse hepatic involvement seen along with pulmonary or miliary tuberculosis. Despite the diffuse involvement of the liver - symptomatic liver disease is often absent.

- Diffuse hepatic infiltration without recognizable pulmonary involvement, also known as granulomatous TB hepatitis.

- A focal/local tuberculoma or abscess formation

Primary hepatic tuberculosis – cont.

- The presence of jaundice suggests biliary involvement.
- Jaundice or biliary stasis is due to either porta hepatis nodes causing biliary compression or to pericholangitis. There might be direct involvement of biliary epithelium or rupture of tuberculous granuloma into the bile ducts as well.
- Massive miliary spread to the liver may cause acute liver failure as well as septic shock with multiorgan failure.
Primary hepatic tuberculosis – con.

- Tubercle bacilli reach the liver by hematogenous dissemination.
- The portal of entry in the case of miliary tuberculosis is through the hepatic artery whereas in the case of focal liver tuberculosis it is via the portal vein.
- Irrespective of the mode of entry, the liver responds by granuloma formation.
- Tuberculous granulomata are most frequently found in the periportal areas (zone 1 of Rappaport) but may occasionally occur in perivenular areas (zone 3) as well.

Both caseating and non-caseating granulomas are seen.

In focal tuberculosis, various granulomas may coalesce to form a large tumor like tuberculoma.

A tuberculoma which has undergone extensive caseation and liquefactive necrosis may form a tubercular abscess.
## Presenting lab investigations

<table>
<thead>
<tr>
<th>investigation</th>
<th>percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Deranged LFT</td>
<td>30-80%</td>
</tr>
<tr>
<td>Abnormal chest X-ray</td>
<td>75%</td>
</tr>
<tr>
<td>Histopathological examination</td>
<td></td>
</tr>
<tr>
<td>epithelioid granuloma formation</td>
<td>80-100%</td>
</tr>
<tr>
<td>caseation</td>
<td>33-100%</td>
</tr>
<tr>
<td>AFB stain</td>
<td>60%</td>
</tr>
<tr>
<td>PCR assay</td>
<td>88%</td>
</tr>
</tbody>
</table>
Primary hepatic tuberculosis – cont

- The final diagnosis of hepatic TB - local or diffuse, rests on histopathological evidence of caseating granuloma or demonstration of acid fast bacilli on smear or culture of biopsy specimen.
- The aim should be to demonstrate the presence of tubercle bacilli in the liver tissue, either directly or by culture.
Primary hepatic tuberculosis – cont.

- Cumulative mortality for hepatic tuberculosis ranges between 15 and 42%.

The factors associated with adverse prognosis are:
- age <20 years,
- miliary tuberculosis,
- concurrent steroid therapy,
- AIDS,
- cachexia,
- associated cirrhosis and liver failure.
TB and Hematologic diseases
Liver tuberculosis in hematologic patients

[Malignant hemopathies combined with latent tuberculosis (a report of 2 cases)]. Vutr Boles 90-3: (4)26; 1987

[Article in Bulgarian]
Naplatanova N, Georgieva R, Kűrnolski I.

Abstract

- Two cases of a combination of a malignant blood disease with latent tuberculosis are presented. One of the patients was with acute leukemia and the other one was with non-Hodgkin's malignant lymphoma (Lymphosarcoma). The blood disease dominated the clinical picture and determined the severity of the course and the lethal outcome. The post mortem examination revealed hepatosplenic form of miliary tuberculosis in both patients. In one of the patients caseous tuberculosis of the bronchopulmonary, peribronchial and periportal lymph nodes was found, too. The tuberculous process had a latent course without characteristic manifestations but it also led to worsening of the patients' condition.
Liver tuberculosis in hematologic patients

[Miliary tuberculosis developed after cured lymphogranulomatosis]. [Article in Russian]
Kirpicheva GN, Alfionova OIu.
Liver tuberculosis in hematologic patients

Liver changes in reactive haemophagocytic syndrome.
Tsui WM(1), Wong KF, Tse CC. Author information: (1)Institute of Pathology, Queen Elizabeth Hospital, Hong Kong.

- There were reported the findings of 12 fatal cases of RHS in which histological materials of the liver are available for study. The underlying diseases of these patients included lymphoma/leukemia (6 cases), disseminated undifferentiated carcinoma of the ovary (1 case), disseminated nasopharyngeal carcinoma complicated by tuberculosis (1 case), adenovirus pneumonia (1 case), pneumococcal pneumonia (1 case), typhoid fever (1 case), and possible drug intoxication (1 case). Ten patients had involvement of the liver by the underlying disease process which contributed to the marked hepatic derangement.
Liver tuberculosis in hematologic patients


Abdominal tuberculosis: a frequent diagnostic challenge.

Ibrahim EM, Anim JT, Al-Idrissi H, Al Mohaya S, Al Dossary J, Grant CS.

A case of abdominal tuberculosis is described in a young male who presented with non-specific symptoms together with CT scan findings that simulated abdominal lymphoma.
Liver tuberculosis in hematologic patients


[Article in French]

Bartoli JM, Poncet M, Harle JR, Moulin G, Distefano D, Weiller PJ, Kasbarian M.

1Service central de Radiologie, CHU Timone, Marseille.

Abstract

The authors report a case of hepatic tuberculosis on a pre-existing Hodgkin disease with hepatic localisations. They point out how echographic and CT imaging are non specific in this disease and so insist on the diagnostic interest of puncture-biopsy under guidance of the hepatic nodules.
There is a high index of suspicion to diagnosis of abdominal tuberculosis.

- age <20 years,
- miliary tuberculosis,
- concurrent steroid therapy,
- AIDS,
- cachexia,
- associated cirrhosis and liver failure.