Malakoplakia of Liver Diagnosed by a Needle Core Biopsy

A Case Report and Review of the Literature

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Although malakoplakia of the genitourinary tract and colon is reported frequently in the literature, malakoplakia that occurs primarily in the liver is rare, and only 4 cases have been described thus far. To our knowledge, this is the first case of malakoplakia of the liver diagnosed by a needle core biopsy. This case occurred in a 19-year-old man with small bowel ileus following Klebsiella pneumonia.

PATHOLOGIC FINDINGS

Histologic sections of the liver needle core biopsy specimen showed cirrhotic hepatic parenchyma with bile ductular proliferation and portal inflammatory infiltrates composed of lymphocytes, neutrophils, and a few scattered eosinophils. Rare isolated bile ductules were associated with neutrophils consistent with cholangitis. Scattered within periportal and portal regions were small, round-to-oval targetoid structures consistent with Michaelis-Gutmann bodies (Figure, A). These stained positively with periodic acid–Schiff (with and without diastase) (Figure, B), Von Kossa calcium (Figure, C), and colloidal iron stain (with and without hyaluronidase) (Figure, D), but were negative for copper and α1-antitrypsin. Immunohistochemistry with CD68 (Figure, E) demonstrated these targetoid bodies within scattered histiocytes and Kupffer cells. Electron microscopy confirmed the presence of Michaelis-Gutmann bodies (Figure, F) within histiocytes and failed to show any evidence of mucopolysaccharidosis. The hepatocyte cytoplasm was neither pale nor reticulated as would be expected in mucopolysaccharidosis.

COMMENT

Malakoplakia, a Greek term meaning soft (malako) plaque (plakia), is an unusual inflammatory process. It was originally described in the early 1900s. Although the original article was published by Michaelis and Gutmann in 1902, its discovery is best attributed to their senior colleague, Von Hansemann, even though his article was published a year later.

Malakoplakia was first reported and is most commonly seen in the bladder. It has since been described in other locations, such as the colon, upper and lower genitourinary tract, gynecologic regions, upper and lower respiratory tract, spleen, joints, and brain. It has rarely been reported to occur in the liver. This case is the fifth reported case of malakoplakia of the liver and the first to be diagnosed by a needle core biopsy (Table). Two other cases involved the liver through extension from the kidney and urinary tract.
Malakoplakia is usually associated with *Klebsiella* and *Escherichia coli*, although gram-negative and gram-positive cocci and acid-fast bacilli have also been identified. Recently, malakoplakia has been reported to occur in association with unusual organisms such as *Rhodococcus equi* and paracoccidioidomycosis infection. The most plausible pathogenetic explanation for the development of malakoplakia centers on the defective function of macrophages that phagocytose the bacteria but are unable to kill or digest them. Intracytoplasmic phagolysosomes continue to enlarge and coalesce with subsequent degenerative change of both matrix and peripheral hyperemia. The lesions of malakoplakia may be solitary, multifocal, or present as a large mass.

Microscopically, malakoplakia is characterized by aggregates of histiocytes, the Von Hansemann cells, with Michaelis-Gutmann calcospherites. Surrounding inflammatory cells usually consist of lymphocytes and plasma cells. There are 3 phases in the histopathologic evolution of malakoplakia, as described by Smith. The early phase consists of Von Hansemann histiocytes and plasma cells in an edematous stroma. The granulomatous phase displays many lymphocytes and plump histiocytes with typical Michaelis-Gutmann calcospherites. The fibrosing or healing phase is characterized by interwoven fibroblasts and strands of thickened collagen admixed with histiocytes. Michaelis-Gutmann bodies are sparse.

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Reports of Malakoplakia of Liver

<table>
<thead>
<tr>
<th>Source, y</th>
<th>Age/Sex</th>
<th>Underlying Disease</th>
<th>Diagnostic Material</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moldavski and Rustamov, 1984</td>
<td>34/Female</td>
<td>Miliary tuberculosis with hepatic pseudocyst</td>
<td>Autopsy</td>
</tr>
<tr>
<td>De Saint-Maur and Gallot, 1990</td>
<td>68/Female</td>
<td>Infected polycystic liver</td>
<td>Segmentectomy</td>
</tr>
<tr>
<td>Robertson et al, 1991</td>
<td>54/Female</td>
<td>Liver abscess following immunosuppression for systemic lupus erythematosus</td>
<td>Wedge biopsy</td>
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<tr>
<td>Boucher et al, 1994</td>
<td>43/Male</td>
<td>Perforated colonic diverticulum</td>
<td>Segmentectomy</td>
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<tr>
<td>Present case, 2000</td>
<td>19/Male</td>
<td>Small bowel ileus following Klebsiella pneumonia</td>
<td>Needle core biopsy</td>
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linergic agonists would correct the lysosomal deficiency and prove therapeutically beneficial.\textsuperscript{10}

In summary, this case occurred in a 19-year-old man with a cirrhotic liver of uncertain origin, who subsequently developed malakoplakia of the liver following sepsis that resulted from \textit{Klebsiella} lobar pneumonia. It is the fifth reported case in the medical literature of malakoplakia that occurred primarily in the liver and, to our knowledge, the first to be diagnosed by a needle core biopsy. A surgical procedure, which comparatively has more potential for complications than a needle core biopsy, may be avoided in some cases. Complete workup and diagnosis of malakoplakia are possible even in specimens as small as those obtained with a needle core biopsy.

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\textbf{References}