MONOCLONAL IMMUNOGLOBULIN DEPOSITION DISEASE: A RARE CASE OF ACUTE LIVER FAILURE

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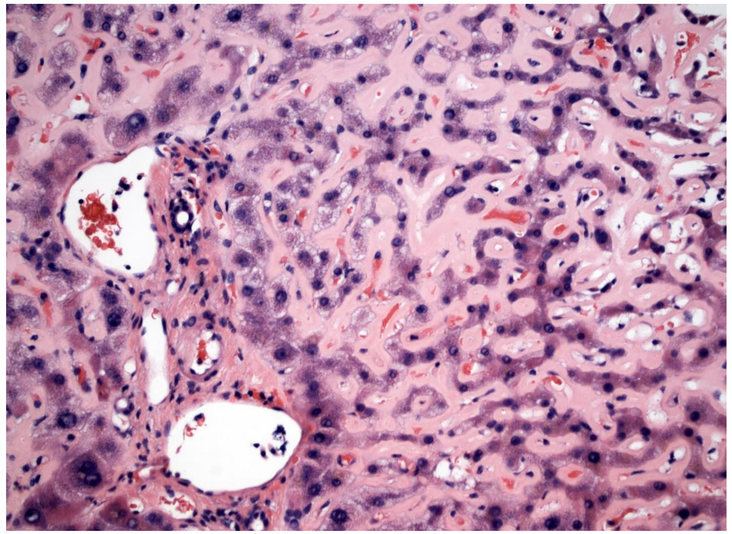
Introduction

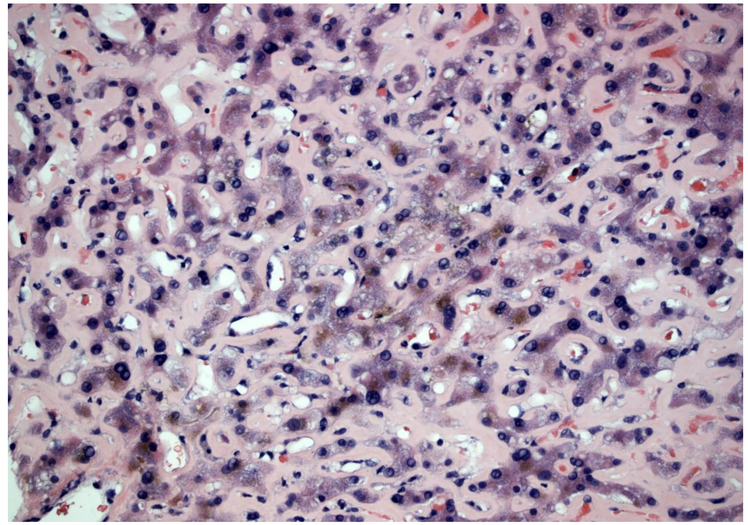
- Light chain deposition disease (LCDD) is a characterized by deposition of immunoglobulin light chain in extracellular tissue.
- Classically it involves the kidneys, however, can rarely affect other organs such as liver.
- Its manifestation is mostly non-specific and presents as mild cholestatic liver injury and , acute liver failure is extremely rare.

Case Description

- An 83-year-old female with MGUS was admitted to the hospital for dyspnea and anasarca. Her initial labs were remarkable for mild, predominantly cholestatic, pattern of liver injury.
- Additional workup was negative for common etiologies of liver injury, with normal appearance of liver and new-onset diastolic cardiac dysfunction.
- Initial diagnosis was acute liver injury in the setting of recent heart failure and treatment with aggressive diuresis was commenced.
- However, she progressed to acute liver dysfunction and multi-organ failure manifesting as encephalopathy, jaundice (T.Bili 16.25) and coagulopathy (INR 1.3).
- Her Alk-Phos was 1116and AST, ALT were 289 and 95, respectively.
- Liver biopsy revealed deposition of pink hyaline material expanding the sinusoids.
- This was initially suspected to be amyloid, until a positive PAS and negative Congo-Red stain established the diagnosis of LCDD.
- SPEP revealed a M-protein light chain with elevated κ/λ ratio of 16.6.
- Chemotherapy was rejected considering poor prognosis and patient was discharged on hospice care.

Case Description





Pale pink hyaline material (negative for Congo Red stain) expanding the sinusoid with focal yellow bile pigment (consistent with hepatocellular cholestasis). There is no significant portal or lobular inflammation.

Discussion

- LCDD results from excessive production and deposition of monoclonal light chains into various organs including liver.
- Concurrent plasma cell dyscrasias and multiple myeloma are often seen in these patients.
- High suspicion of disease and biopsy without delay is imperative for diagnosis, which typically show granular, κ-LC predominant deposits in the perisinusoidal basement membrane which unlike amyloid, appear unorganized and stain positive with PAS and negative with Congo-Red.
- At present, there is no consensus on LCDD treatment, however, autologous stem cell transplant following bone marrow conditioning with high dose melphalan and bortezomib based induction chemotherapy with or without systemic glucocorticoids have been reported to treat hepatic and systemic LCDD with some success.