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A Rare Case of Post Infantile Giant Cell Hepatitis- Case Report

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Abstract: ***Giant cell hepatitis is characterized by formation of multinucleated hepatocytes and inflammation of the liver parenchyma. It is rare in adults and associated with autoimmune disorders, hematological diseases, medications and rheumatological diseases. We report a case of 47 year old female who presented with jaundice and fever. Trucut biopsy from liver was performed. Microscopic findings favored a diagnosis of Post infantile giant cell hepatitis with autoimmune hepatitis.***

Keywords: giant cell hepatitis, autoimmune hepatitis, liver inflammation, jaundice, liver biopsy

1. Introduction

Giant cell hepatitis is a condition characterized by inflammation and large multinucleated hepatocytes in the hepatic parenchyma. Giant cell transformation of hepatocytes along with extramedullary hematopoiesis is a common response in the newborn liver diseases. Postinfantile giant cell hepatitis is a rare disorder in adults. It is an unusual regenerative or degenerative hepatocytes response to various noxious stimuli, characterized by the presence of multinucleated cells in liver with generally dismal clinical outcome.

2. Case Details

47 year old female presented with jaundice and fever for one month duration. On examination patient had icterus, fever, and enlarged liver and it was tender on palpation.USG abdomen showed chronic liver disease, focal lesion in right lobe of liver (segment VIII) and hepatomegaly. Antinuclear antibody was positive.Her bilirubin levels were 1.1 mg/dL. LDH, ferritin, ceruloplasmin levels were normal. She underwent trucut biopsy from right lobe of the liver and sample was sent for histopathological examination.

We received two linear cores of yellowish to grey white tissue, each measuring two centimeter in length. Micrpscopy showed linear cores of liver tissue with retained architecture. Individual hepatocytes shows ballooning degeneration with patchy lobular necrosis and mutinucleated giant cell formation of hepatocytes. Portal area shows intense inflammation lymphocytes and plasma cells. Interphase hepatitis noted in >50% of the portal area. Pseudorosette formation noted. Reticulin staining showed fibrous portal expansion.Viral markers were negative.

With clinical features, serological findings and histopathological morphology, final diagnosis of Post infantile giant cell hepatitis with autoimmune etiology (Modified Ishak activity index-score 15/18, fibrosis staging-3/6) was given.

Microscopy:

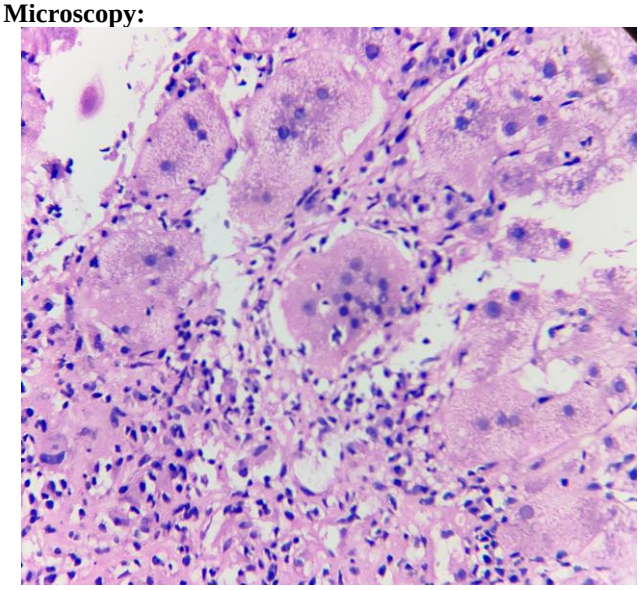

Figure 1: Giant cell formation of hepatocytes

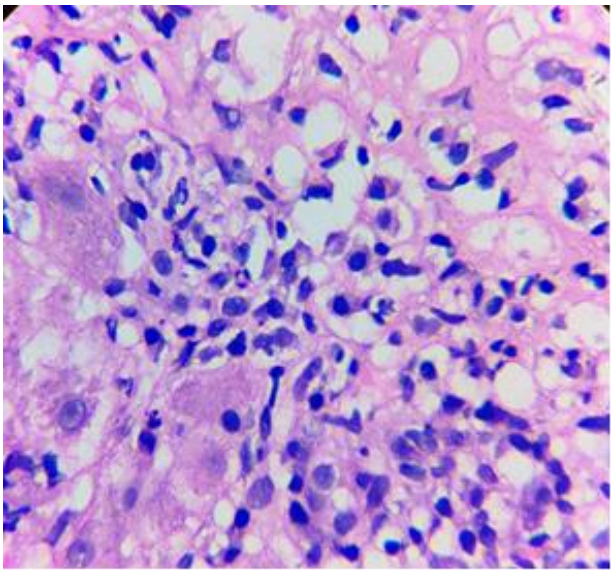

Figure 2: Inflammatory infiltrates composed of lymphocytes and plasma cells

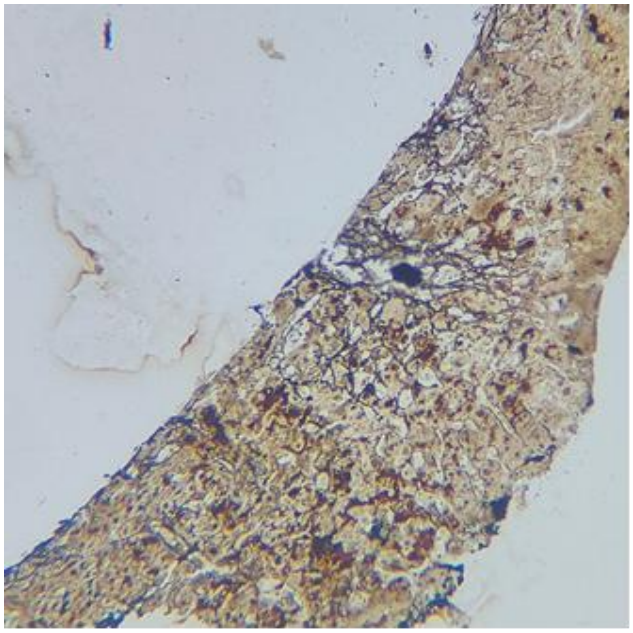

Figure 3: Reticulin staining

3. Discussion

Post infantile giant cell hepatitis is very rare in adults (0.1%–0.25% of all hepatic diseases). Various etiologies associated with postinfantile giant cell hepatitis includes medications, autoimmune disorders and infections

"Certain medications, including methotrexate, 6-mercaptopurine, clometacine, amitriptyline, chlordiazepoxide, p-amino salicylic acid, vinyl chloride, chlorpromazine, herbal medicines, amoxicillin-clavulanate, and doxycycline, can potentially cause giant cell hepatitis. These drugs may damage liver cells (hepatocytes), leading to degeneration and the formation of abnormally large liver cells (giant hepatocytes) in susceptible individuals."

Autoimmune hepatitis, particularly type 1 with ANA positivity, is a significant cause of giant cell hepatitis, accounting for 40% of autoimmune-related cases. However, the exact process behind giant cell formation remains unclear.

Two theories attempt to explain this phenomenon:
a) Hepatocytes fuse together.
b) Nuclear proliferation occurs without cell division.

Several factors may contribute to giant cell formation:
a) The autoimmune disease itself.
b) Immune complexes and vascular damage, which can disrupt hepatocyte nutrition.

The progression of the disease varies greatly: Some patients experience normalization of liver histology, while others progress to cirrhosis and liver failure. The severity of the underlying liver disease dictates the prognosis. Unfortunately, the clinical course is often severe, leading to rapid cirrhosis.

Hepatitis A, B, C, E Epstein-Barr virus (EBV), HIV, Cytomegalovirus, and a potentially unidentified paramyxolike virus have been found to be associated with giant cell hepatitis.

Postinfantile giant cell hepatitis presents a varied clinical spectrum, ranging from acute hepatitis to mild chronic liver disease characterized by jaundice, rapid progression to cirrhosis, subacute liver failure, and fatal liver failure. In adults, giant-cell hepatitis is often a progressive and fatal disease process, with a survival rate of approximately 50% without orthotopic liver transplantation. The high mortality rate is primarily due to severe liver failure or sepsis resulting from aggressive immunosuppression. This disease has a significant impact on patients, emphasizing the importance of timely diagnosis and appropriate management to improve outcomes.

Gross examination of liver biopsy specimens may reveal a uniformly dark green to grayish brown color, with the liver often appearing shrunken, although in acute cases it can be enlarged. Microscopically, the diagnostic hallmark of giant cell hepatitis is the presence of giant cells, typically occupying more than two-thirds of the parenchyma, with giant cell transformation most prominent in zone 3. These cells often contain 4-20 centrally located nuclei. Additional microscopic features include periportal lymphocytic infiltrate (primarily T lymphocytes), massive necrosis, bridging necrosis, activated perisinusoidal cells, bilirubin stasis, Mallory hyaline bodies often accompanied by neutrophilic infiltrate, and severe fibrosis. These characteristic findings are indicative of giant cell hepatitis, a severe and potentially progressive liver disease.

The formation of multinucleated hepatocyte syncytia in giant cell hepatitis is not fully understood. Proposed mechanisms include increased nuclear proliferation without cell division and membrane fusion between hepatocytes. This giant cell change represents an atypical regenerative response to liver injury in adults.

4. Conclusion

In conclusion we report a rare case of post infantile giant cell hepatitis with typical histomorphological features. Post infantile giant cell hepatitis is a microscopic diagnosis, so the histopathological examination is essential for diagnosis and treatment of the patient.

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