SEVERE POSTINFANTILE GIANT CELL HEPATITIS IN AN ADOLESCENT MALE RESPONDING TO IMMUNOSUPPRESSIVE THERAPY DESPITE NEGATIVE AUTOIMMUNE MARKERS

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Background: Giant cell hepatitis, characterized by extensive giant cell transformation of hepatocytes and variable inflammation, is commonly seen in newborn liver disease. Postinfantile Giant Cell Hepatitis (PIGCH) is a very rare pathologic finding in older children and adults. Thought to be due to a regenerative hepatocyte response to harmful stimuli, PIGCH is associated with drugs, viral infection, autoimmune diseases and other etiologies.

Aims: We describe a case of severe, chronic PIGCH in an adolescent male with no identifiable etiology despite exhaustive investigations. Ultimately, he responded to corticosteroids despite negative autoimmune markers.

Methods: A case of unexplained PIGCH in an adolescent male was reviewed. The literature on PIGCH was explored and summarized.

Results: A previously healthy 15-year-old male presented with a 2.5-month history of jaundice and hepatitis (ALT/AST > 2500 U/L, ALP 667 U/L, total bilirubin 64μmol/L and INR 1.1). There was no history of travel, toxins, or medication use. A thorough infectious work up was negative. Metabolic causes such as Wilson’s disease and alpha-1-antitrypsin were excluded. IgG levels were normal (12 g/L) and an extensive autoimmune hepatitis antibody panel was negative. A liver biopsy demonstrated severe lobular inflammation with abundant multinucleate giant cells and early bridging fibrosis.

Due to ongoing severe hepatitis, the patient was started on prednisone despite negative autoimmune markers and by 48 hours, his liver enzymes improved dramatically. Azathioprine and ursodiol were introduced following prednisone initiation, which led to normalization of his liver enzymes. However, follow up liver biopsies at 10 and 24 months demonstrated ongoing mild chronic hepatitis with only occasional giant cells and progression to severe fibrosis.Transient elastography values increased over two years, suggesting ongoing fibrosis. Currently the patient is asymptomatic and is maintained on budesonide, azathioprine and ursodiol. All autoimmune markers remain negative.

Conclusions: PIGCH due to an underlying autoimmune etiology accounts for approximately 40% of all reported PIGCH cases, and presents with typical autoimmune markers such as elevated IgG, ANA, and ASMA. Autoimmune PIGCH has a generally poor prognosis, with only about 20% responding well to treatment and the rest progressing to cirrhosis or death. We describe an individual with PIGCH who had no autoimmune markers but biochemically responded to steroid therapy, suggesting prednisone should be considered in patients with no identifiable cause of their PIGCH. Even though our patient had symptomatic and biochemical improvement with immunosuppressive therapy, concern remains over ongoing inflammation and...
possible progression to cirrhosis as described in most other individuals with autoimmune PIGCH.

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