

Gastrointestinal and Liver Pathology at Rush

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Case of the month – February 2009

Contributed by Drs. Ajay Patel and Shriram Jakate

Diagnosis: Adult giant cell hepatitis, variant of autoimmune hepatitis.

Sections of the explanted liver show multinucleated hepatocytes with cholestasis and focal acute inflammation. There is also chronic inflammation and a plasmacytosis within the portal tract. Thus, the diagnosis is adult giant cell hepatitis variant of autoimmune hepatitis.

Giant cell hepatitis is a well known entity in the newborn (known as neonatal giant cell hepatitis) which is a common response of newborn liver to a variety of insults including extrahepatic biliary atresia, paucity of intrahepatic bile ducts, chromosomal and metabolic disorders, and viral infections. It is very rare in adults, causing less than 0.25% of all hepatic disease. About 100 cases have been reported in the literature within the last 20 years. Given the rarity and its typical association with the newborn, it has also been referred to as “postinfantile” or “synctial” giant cell hepatitis. The pathogenesis of the giant cells is thought to be different in the two groups. In newborns, it is thought to be a normal response of immature hepatocytes with a high regenerative capacity to a variety of insults. In adults, it is thought to be an abnormal and idiosyncratic response of hepatocytes with a low regenerative capacity which ultimately leads to cell death. Possible causes in the adult include viral infections, cholestatic disorders, a novel paramyxovirus-like virus, and autoimmune hepatitis. Microscopically, it is characterized by multinucleated hepatocytes and hepatitis. Although this patient was seronegative, surrogate autoimmune markers were present including the history of hemolytic anemia and elevated immunoglobulins. Also, other possible causes such as viral infections or drug were ruled out. Clinically, this entity is progressive and often fatal with a survival rate of 50% without transplant. It can be treated with corticosteroids and if necessary, liver transplant.

References:

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