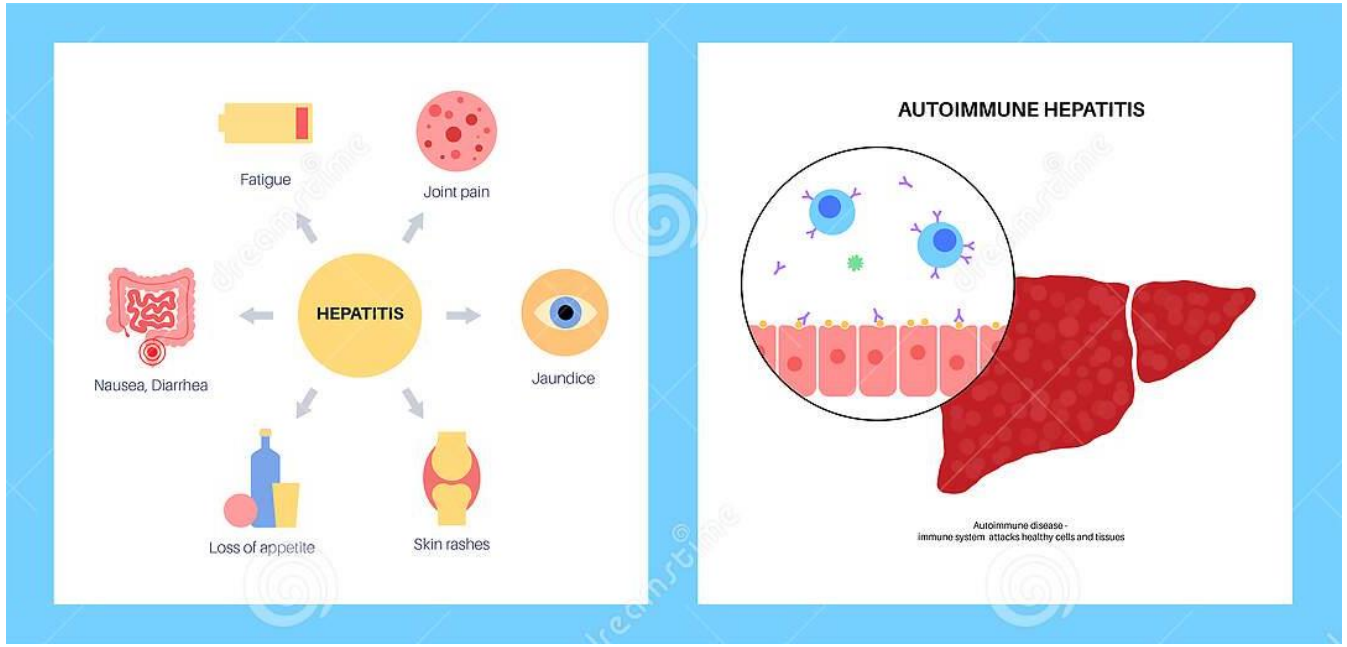
In the Name of GOD



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SUMS

Important clues:

- Recent HAV hepatitis
- Nonspecific symptoms
- ? Chronic liver dx
- ? Extrahepatic manifestations
- FHX of autoimmune dx

The most probable diagnosis:

Clinical presentations:

Variable: Asymptomatic …… Fulminant liver failure.

1. Acute onset: resembling that of viral hepatitis: 1/3
2. Insidious onset: AIH-1 > AIH-2: 1/3
3. Complications of cirrhosis and portal hypertension: 10%
4. Acute liver failure: AIH-2 > AIH-1
5. Incidental finding of raised hepatic aminotransferases
6. Triggered by HAV infection

Clinical presentations:

- Children with AIH often present before puberty
- Relapsing course with flares and spontaneous remissions: delayed or missed diagnosis.
- A more aggressive phenotype in children
- AIH should be suspected in all children with symptoms & signs of liver disease.
- AIH should always be suspected when known causes of acute hepatitis are excluded
- It is essential to inquire about symptoms and signs of the associated autoimmune diseases.
- Physical examination:
  - Jaundice
  - Enlarged liver or spleen
  - Ascites, coagulopathy, and encephalopathy in decompensated disease

Associated AI disorders:

- A family history of autoimmune disease is frequent (40%)
- 20% of patients: associated AI disorders: at diagnosis or during follow-up:
  - Thyroiditis/ Hypoparathyroidism
  - IBD
  - hemolytic anemia
  - Vitiligo
  - coeliac disease
  - insulin-dependent diabetes
  - Behcet disease/ Sjögren syndrome
  - Glomerulonephritis
  - idiopathic thrombocytopenia
  - urticaria pigmentosa
  - Addison disease

Associated immunodeficiency:

- AIH-2 can be part of the APECED:
  - Hypoparathyroidism
  - Addison disease
  - mucocutaneous candidiasis
  - Ectodermal dysplasia
- Other immune-dysregulated states associated with AIH:
  - IPEX
  - common variable immunodeficiency
  - hyper-IgM syndrome
  - Partial IgA deficiency: 40% of patients

AIH-1 vs AIH-2

	AIH-1	AIH-2
Prevalence	2/3	1/3
Age	higher	younger
Female	75%	75%
Acute hepatitis	++	+
Acute liver failure	+	+++
Insidious onset	++	+
Complication of chronic liver disease	+	+
Associated immune disease	+	+
IBD	++	+
Family history of AI dx	++	++
Refractory to treatment withdrawal	+	+++
Cirrhosis	+++	+

Minocycline:

a known trigger of an “auto- immune-like hepatitis”

- Minocycline-induced AIH-like liver injury:
  - usually occurs acutely within 2 years after drug initiation: range of 3 days to 6 years.
  - Jaundice, lethargy, anorexia, and abdominal discomfort
  - Signs of serum sickness (fever, rash, joint pains)
  - Over 90% of patients: ANA positive with hypergammaglobulinemia
  - Only 25%: ASMA positive
  - ALKM positivity has not been described
  - Histologically: mimics classical AIH: cirrhosis at presentation has not been reported.
  - Treatment: stop minocycline + corticosteroids