

CENTRAL PRESENTATION



A 52-Year-Old Woman Presented with Progressive Jaundice

Presenter

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Particulars of the Patient:

- Name – Mrs. X
- Age – 52 years
- Sex – Female
- Marital Status – Married
- Occupation – Housewife
- Address – Kaliakoir, Gazipur
- Date of Admission – 15/03/2024
- Date of Examination – 15/03/2024



Presenting Complaints:

- Yellow discoloration of eyes and urine for 1¹/₂ months.
- Upper abdominal pain for 1¹/₂ months.
- Itching for 1 month.

History of Present illness:

Mrs. X was reasonably well 1¹/₂ months back.

Then she noticed ***yellow discoloration of eyes and urine*** which was:

- Gradually increasing
- Initially associated with ***a transient period of :***
 - ❖ Low- grade fever
 - ❖ Nausea
 - ❖ Loss of appetite
 - ❖ Malaise
 - ❖ Vomiting

History of Present illness:

- ***The vomitus was:***

- ❖ Non-projectile

- ❖ Bile stained

- ❖ Contained some food residues but no blood

- ***Her stool was:***

- ❖ Pale colored

- ❖ Non-voluminous

- ❖ Not mixed with any blood or mucus

History of Present illness:

- She also noticed **generalized itching** for the last 1 month which was:
 - ❖ Mild to moderate intensity
 - ❖ Persisted throughout the day and night
 - ❖ Not related to any food, dust, mite or any visible eruptive skin lesion

History of Present illness:

- She also complained of ***pain and discomfort in the right upper abdomen*** for 1¹/₂ months which was:
 - ❖ Localized
 - ❖ Non-radiating
 - ❖ Mild to moderate in intensity
 - ❖ Dull aching in nature
 - ❖ Not associated with taking meal or gluten containing food
 - ❖ There were no aggravating or relieving factor

History of Present illness:

- She also complained of *fatigue & generalized weakness* for the last 1 month which was:
 - ❖ Marked during exertion
 - ❖ No diurnal variation
- Her bowel and bladder habit was normal

History of Present illness:

- There was no history of:

- ❖ Abdominal swelling

- ❖ Altered level of consciousness

- ❖ Vomiting out of blood

- ❖ Abnormal movement or behavior

- ❖ Passage of black-tarry stool

- ❖ Confusion or coma

History of Present illness:

❖ Nodular swelling of any part of body

❖ weight loss

❖ Skin rash

❖ Joint pain

❖ Pigmentation of skin

❖ Weather preferences

❖ Palpitation

❖ Shortness of breath

❖ Chest pain

History of Present illness:

- She had history of taking some herbal medications

but *no history of:*

- ❖ Transfusion of blood or blood products
- ❖ I/v drug abuse
- ❖ Unprotected sexual exposure
- ❖ Sharing of needles
- ❖ Surgery or instrumentation
- ❖ Tattooing

History of Present illness:

For these above complaints, she admitted into Dept. of Surgery in this hospital and later on referred to Dept. of Hepatology for further evaluation and management.

History of Past illness:

- No significant past medical history

Personal History:

She is:

- Housewife
- Non-smoker
- Non-alcoholic

Family History:

- She lives with her spouse, one daughter and three sons
- All members are healthy
- No family history of liver disease

Socio-economic History:

- Comes from a middle-class family
- Lives in a pacca house
- Uses sanitary latrine
- Drinks arsenic free tube-well water

Obstetric & Gynaecological History

- Menopausal for last 3 years
- No history of pregnancy loss
- Pregnancies were uneventful

Treatment History:

- Took some herbal medicine for her illness
- Had a history of hospitalization in a local hospital
- Did not get improved and was referred to Dhaka Medical College & Hospital

Immunization History:

- Immunized with two dose of covid-19 vaccine
- Not immunized as per EPI schedule

General Examination

- Appearance : ill-looking
- Body built : Average
- Cooperation : Co-operative
- Decubitus : On choice
- Anemia : Absent
- **Jaundice** : **Present (++)**
- Cyanosis : Absent
- Clubbing : Absent
- Koilonychia : Absent
- Leukonychia : Absent

General Examination

- Edema : Absent
- Dehydration : Absent
- Thyroid gland : Not enlarged
- Neck vein : Not engorged
- Lymph nodes : Not palpable
- Bony : Absent
- Tenderness : Absent
- **Skin condition : Multiple scratch marks all over the body with shiny nails**

General Examination

- Pulse : 76 b/min
- Blood : 110/70 mm of Hg
pressure (No postural drop)
- Temp. : 98.4°F
- R/R : 18 breaths/min

No Other Stigmata of Chronic Liver Disease

Alimentary System :

- Lips, gum, teeth: Normal
- **Dorsal surface of tongue & oral cavity: Yellowish**
- ***Abdomen:***
 - ❖ **Inspection:**
 - Shape- Normal
 - Umbilicus- Centrally placed, inverted
 - Flanks- Not full
 - Scar mark, engorged or visible vein- Absent

Alimentary System :

❖ *Palpation:*

□ **Superficial palpation:**

- Temperature: Normal
- Tenderness: Mildly tender right hypochondrium
- Murphy's sign: Negative
- No hyperesthesia, muscle guard or lump

Alimentary System :

❖ *Palpation:*

□ Deep palpation:

➤ **Liver:** Liver was palpable, 2 cm from right costal margin along the midclavicular line

- Margin was regular
- Surface was smooth
- Mildly tender
- Firm in consistency
- Upper border of liver dullness in right 5th ICS
- Total liver span- 15 cm

Alimentary System :

❖ *Palpation:*

□ Deep palpation:

- **Gallbladder** : Not palpable
- **Spleen**: Not palpable
- **Kidneys**: Not ballotable
- **Para-aortic Lymph Node**: Not palpable
- **Urinary bladder**: Not palpable

Alimentary System :

❖ Percussion: Tympanic

- Shifting Dullness- Absent

❖ Auscultation:

- Bowel Sound- Present
- Hepatic Bruit- Absent

❖ Digital Rectal Examination : Normal

Other Systemic Examinations were Unremarkable

Salient Features

Mrs. X, 52-years old, Muslim, married, housewife, normotensive, non-diabetic, non-smoker & non-alcoholic hailing from Kaliakoir, Gazipur got admitted into this hospital with a 1¹/₂ months history of:

Progressive jaundice which was preceded by prodromal symptoms and was associated with pale stool, generalized itching for 1 month without any visible skin lesion and abdominal pain which was in the right upper abdomen, mild, dull aching in nature without any radiation, aggravating or relieving factor. She also had fatigue & generalized weakness over this period.

Salient Features

On general examination, patient was moderately icteric, multiple scratch marks, shiny nails were present and she was haemodynamically stable.

On abdominal examination, there was hepatomegaly present, 2 cm from right costal margin along the midclavicular line, margin was regular, surface was smooth, mildly tender, firm in consistency, upper border of liver dullness in right 5th ICS, hepatic bruit absent.

No other organomegaly or stigmata of chronic liver disease were present.



Problem List from Audience

Problem List

**Jaundice
1.5 months**

**Itching
1 month**

**52- years
old woman**

**Abdominal Pain
1.5 months**

**Fatigue
1 month**

Hepatomegaly

What Might Be The Diagnosis?

Provisional Diagnosis

Acute hepatitis with prolonged cholestasis

Differential Diagnosis

- Cholelithiasis
- Primary biliary cholangitis
- Drug-Induced Liver Injury (DILI)
- Autoimmune hepatitis
- Peri-ampullary carcinoma

INVESTIGATIONS

Investigations

CBC	15/03/24	06/4/24	25/4/24
Hb (g/dL)	13.2	13.7	11.8
ESR	32	44	65
RBC ($\times 10^6/\text{mm}^3$)	4.95	4.75	3.75
WBC ($/\text{mm}^3$)	10,500	12,000	10,000
Neutrophil	81%	52%	47%
Lymphocyte	12%	38%	45%
Monocyte	6%	6%	6%
Eosinophil	1%	4%	2%
Basophil	0%	0%	0%
Platelet Count ($/\text{mm}^3$)	2,50,000	2,50,000	1,80,000

Investigations

Liver Function Test	15/03/24	31/03/24	06/04/24	25/04/24
S. Bilirubin (Total) (mg/dl)	26	23.6	25.2	27.5
S. Bilirubin (Direct) (mg/dl)	-	14.14	19.5	-
S. Bilirubin (Indirect) (mg/dl)	-	9.46	5.7	-
AST (SGOT) (U/L)	962	897	661	801
ALT (SGPT) (U/L)	560	353	308	371
ALP (U/L)	145	183		329
GAMMA-GT (U/L)	180			501
Prothombin Time (Sec)	18.4			16.3

Investigations

Test	18/03/2024
S. Creatinine (mg/dl)	0.36
S. Electrolytes:	
Sodium (Na+) (m mol/L)	140
Potassium (k+) (m mol/L)	3.9
Chloride (Cl-) (m mol/L)	109
RBS (m mol/L)	5.6

Investigations

Viral markers	16/03/2024
Anti HAV IgM	Negative
Anti HEV IgM	Negative
HBsAg	Negative
Anti HCV	Negative
Anti HBc total	Negative

Investigations

Test	27/03/2024
ANA	Negative
ASMA	Negative
Anti LKM1	Negative (<titre 1:40)
AMA	Negative

Slit Lamp examination:

K-F ring absent in both eyes

Investigations

Test	Result	Reference Value
S. IgG (gm/l) (27/03/24)	↑ 20	7.67- 15.9
S. Ceruloplasmin (mg/dl) (06/04/24)	36	18-58
24 hours urinary copper (µgm/L) (29/04/24)	41.2	>100

Investigations

USG OF WHOLE ABDOMEN

- Liver** : Liver is normal in size. Parenchymal echogenicity appears homogeneous. Parenchymal echogenicity is not increased. No focal lesion is seen. Portal vein caliber and hepatic veins channels are within normal limit.
- Gall bladder** : *Gall bladder is contacted. Multiple bright echogenic*
~~Gall bladder is normal in size, shape with normal wall thickness.~~
Lumen is clear. Sometimes, casting posterior in acoustic shadow are noted in CB Lumen
- Biliary channels** : Intra & extrahepatic biliary trees are not dilated. CBD is normal in caliber.
- Pancreas** : Normal in size, shape in outline with uniform echotexture. MPD is not dilated.
- Spleen** : Normal in size with uniform echotexture. No focal lesion is seen.
- Kidneys** : Normal in size, shape and position. Cortical echogenicity is normal. Cortico-medullary differentiation is maintained in both kidneys. Pelvicalyceal systems are not dilated on both kidneys. No calculus seen.
- Urinary bladder** : Well filled. Wall thickness is within normal limit and regular in outline. No intravesical lesion is noted.
- ~~**Prostate/Uterus** : Normal in size & shape according to age~~

comment : cholelithiasis (multiple)

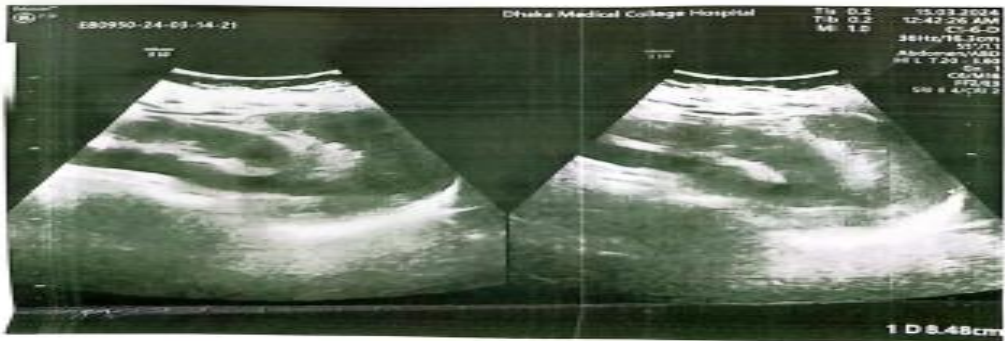
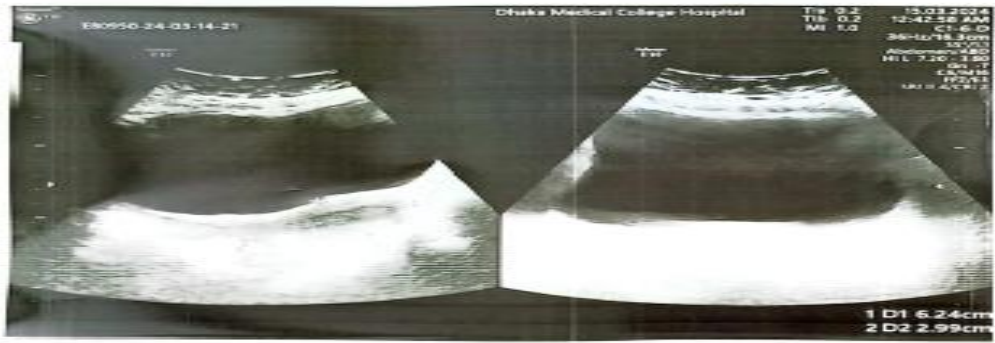
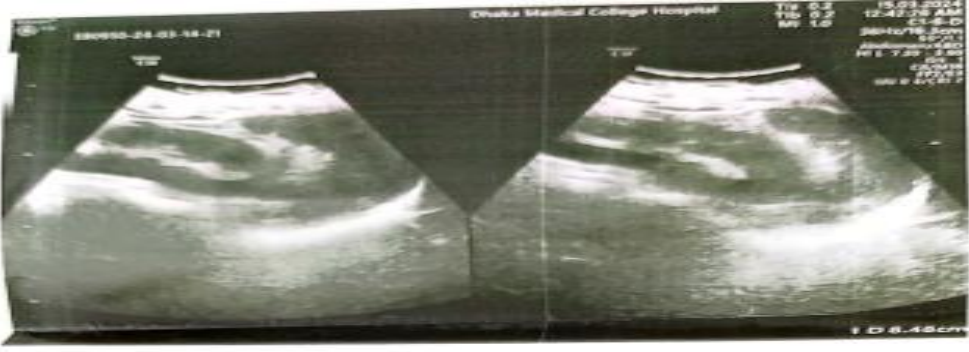
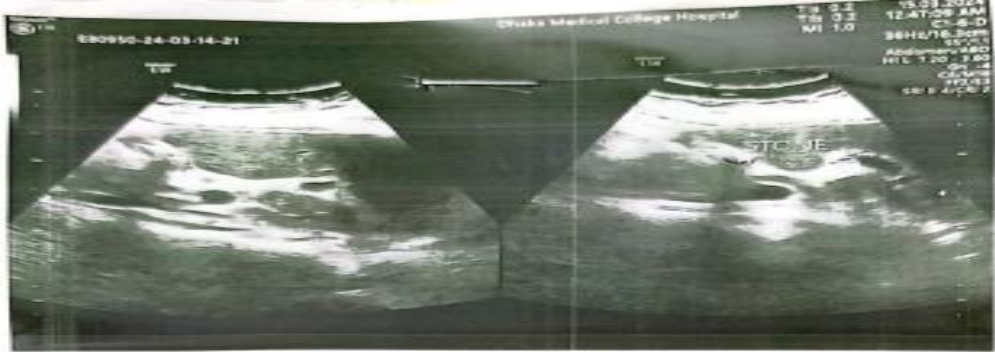
Test

20/03/2024

USG of W/A

**Cholelithiasis
(multiple)**


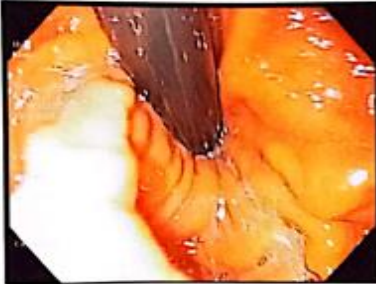

Investigations



Investigations

Test	31/03/2024
Endoscopy of upper G. I	Normal

ENDOSCOPIC PROCEDURE REPORT



Procedure : ENDOSCOPY OF UPPER G. I. TRACT

Indication : N/A

Medication : TIVA With Inj Profol.

Consultant Anaesthetist : Dr. Nawsher Alam.
Endoscopic Findings

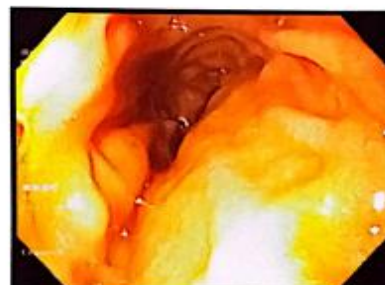
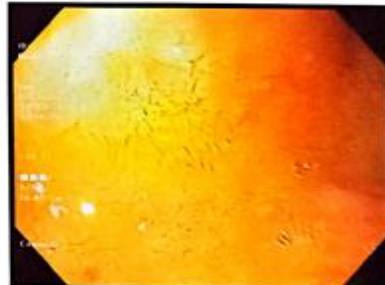

Oesophagus : Normal . No varix seen.

Stomach : Normal.

Duodenum : Bulb and post-bulbar area is normal including periampullary region.

Biopsy : Not taken.

COMMENTS : Normal Upper G.I.T. at Endoscopy.



Investigations

Test	20/03/2024
MRCP	Suggestive of hepatitis

Technique:

FS T2WI axial, coronal; MRCP.

Findings:

1. Liver: Mildly enlarged in size. It exhibits inhomogeneous parenchymal intensity. No focal area of altered intensity or abnormal enhancement is seen in the liver.
2. IVC & PV: Normal.
3. Spleen: Not enlarged. No intraparenchymal lesion is noticed.
4. **GB:** Gallbladder is contracted with uniform thickening of wall. No calculi with signal void or soft tissue lesion is seen within the lumen.
5. **Biliary channel:**
 - a. No sign of dilatation of right & left hepatic ducts and CBD. No sign of calculi with smoothly marginated signal void or soft tissue mass is seen within biliary channel.
 - b. Apparent no extra-luminal mass is present at ampullary region.
6. Pancreas: No evidence of edema, cyst or focal parenchymal lesion. Main pancreatic duct is not dilated.
7. Mesentery & peritoneum: Intensity appears to be normal.
8. Enlarged hepatic hilar, celio-mesenteric, paraaortic LNs: not present.
9. Ascites: not present.

IMPRESSION:

Suggestive of hepatitis.

Advice: Clinicopathological correlation for further evaluation please.

Investigations

Test	30/04/2024
Liver Biopsy	<p data-bbox="428 329 1085 468">Compatible with autoimmune hepatitis</p> <ul data-bbox="392 508 1105 1133" style="list-style-type: none"><li data-bbox="392 508 1105 572">■ Periportal fibrous band<li data-bbox="392 622 1105 791">■ Perisinusoidal fibrous band<li data-bbox="392 851 1105 1019">■ Infiltration of chronic inflammatory cell<li data-bbox="392 1079 1105 1133">■ Piecemeal necrosis

Specimen: Liver tissue

Clinical information: Suspected autoimmune hepatitis / primary biliary sclerosis.

Gross description:

Specimen consists of a 1.3 cm long linear piece of tissue. Submitted as such.

Microscopic examination:

Sections show a core of liver tissue. It reveals proliferation of fibrous tissue forming periportal bands. Perisinusoidal fibrosis is also present (MT stain). The portal areas and the fibrous bands contain moderate infiltration of chronic inflammatory cells. Moderate piecemeal necrosis and moderate lobular inflammation are present. Eosinophils are scanty.

No granuloma or malignancy is seen.

Diagnosis: Compatible with autoimmune hepatitis.

Adv.: Clinical correlation.

Revised Diagnostic Criteria for AIH :

In This Patient:

- Female gender = +2
- ALP:AST ratio (0.15) = +2
- S. IgG (20) = +1
- Average alcohol intake (<25g/day) = +2
- Hepatitis viral markers = +3
- Histologic findings:
 - ✓ Interface hepatitis = +3
 - ✓ Lymphoplasmacytic infiltrate = +1
- **Total Score = 14 (Probable AIH)**

Clinical feature	Score
Female gender	+2
ALP:AST ratio <ul style="list-style-type: none"> • <1.5 • 1.5–3.0 • >3.0 	+2 0 –2
Serum globulin or IgG above normal <ul style="list-style-type: none"> • >2.0 • 1.5–2.0 • 1.0–1.5 • <1.0 	+3 +2 +1 0
ANA, SMA, LKM1 <ul style="list-style-type: none"> • >1:80 • 1:80 • 1:40 • <1:40 	+3 +2 +1 0
Illicit drug use history <ul style="list-style-type: none"> • Positive • Negative 	–4 +1
Average alcohol intake daily <ul style="list-style-type: none"> • <25 g/day • >60 g/day 	+2 –2
Histologic findings <ul style="list-style-type: none"> • Interface hepatitis • Lymphoplasmacytic infiltrate • Rosette formation • None of the above • Biliary changes • Other changes 	+3 +1 +1 –5 –3 +2
Other autoimmune disease	+2
AMA positivity	–4
Hepatitis viral markers <ul style="list-style-type: none"> • Positive • Negative 	–3 +3
Aggregate score without treatment <ul style="list-style-type: none"> • Definite AIH • Probable AIH 	>15 10–15

Final Diagnosis

Sero-Negative Autoimmune hepatitis

Treatment

Treatment given:

- Tapering dose of steroid
- Azathioprine
- Calcium, Vitamin D
- Anti ulcerant

Follow-up

Investigations

Liver Function Test	10/05/24 (1st Week)	24/06/24 (6th Week)	03/11/24 (8th Week)
S. Bilirubin (Total) (mg/dl)	3.4	1.2	0.7
AST (SGOT) (U/L)	138	32	44
ALT (SGPT) (U/L)	201	36	25
ALP (U/L)	193	115	111
S. IgG (gm/l)	-	-	13.9

Take Home Message

- Autoimmune hepatitis (AIH) is an immune-mediated liver disease that can present in all ages and races and both sexes
- Disease presentation is variable, ranging from asymptomatic disease to fulminant liver failure. Acute presentations are often indistinguishable from a viral illness
- **Approximately 10-20% of AIH patients are initially seronegative for conventional auto-antibodies**

Take Home Message

- There is an absence of a specific diagnostic marker, hence diagnosis is made by exclusion of alternative liver disease and precipitants, for example, drugs and viruses
- **Response to therapy would confirm a diagnosis of auto-antibody negative AIH**

Ref: Sherlock's Diseases of the Liver and Biliary System (13th Edition)

Take Home Message

Ref: Sherlock's Diseases of the Liver and Biliary System (13th Edition) by James S. Dooley, Anna S. F. Lok, Guadalupe Garcia-Tsao, Massimo Pinzani

Acknowledgement

- Department of Surgery, Dhaka Medical College & Hospital
- Department of Haematology, Dhaka Medical College & Hospital
- Department of Radiology & Imaging, Dhaka Medical College & Hospital

THANK YOU

