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Cirrhosis of Liver (Metabolic Etiology)

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Abstract

The exact prevalence of cirrhosis is not known because the disease is often silent. Nearly 30% to 40% of cases are discovered at autopsy, indicating that in a substantial proportion of people, the disease goes undetected during life. Signs and symptoms: Organs commonly affected by haemochromatosis are the liver, heart, and endocrine glands. Haemochromatosis may present with the following clinical syndromes. Cirrhosis of the liver: Varies from zonal iron deposition] to fibrosis (cirrhosis). Diabetes due to selective iron deposition in pancreatic islet beta cells leading to functional failure and cell death. Cardiomyopathy, Arthritis (calcium pyrophosphate deposition in joints), Testicular failure; Slate grey discoloration of the skin, Joint pain and bone pain. Routine treatment in an otherwise-healthy person consists of regularly scheduled phlebotomies (bloodletting). When first diagnosed, the phlebotomies may be fairly frequent, perhaps as often as once a week, until iron levels can be brought to within normal range. The major etiology of liver cirrhosis in Indian remains HCV. Our survey revealed the prevalence of NASH-related LC in Japan and the frequency of HCC. Future changes in etiology must be considered in establishing preventive or educational strategies, as well as in developing new treatment strategies.

Key words: Ferritin, Hemochromatosis, Inflammation, Iron overload, Percent saturation, Transferrin, Total iron binding capacity, Iron chelating agents, hepatic iron index

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Introduction

The term "cirrhosis" is derived from the Greek work kirrhos, meaning "tawny" and referring to the tan color of the liver. Cirrhosis is a histologic diagnosis, based on three essential criteria: diffuse disease, presence of fibrosis, and replacement of normal architecture by abnormal nodules. [1]

The initial step is deposition of fibrous tissue at sites of liver cell necrosis, which in the initial stages links portal and central areas or one portal/central area with another. [2] As the disease advances, the fibrous bands become wider and denser. [3] These nodules, are called "pseudolobules" because the normal lobular architecture is lost. Depending upon their size, the defect is termed as "macronodular" (>3 mm in diameter) or "micronodular cirrhosis." [4]The histologic abnormality should involve the entire liver, since localized defects such as focal nodular hyperplasia do not constitute cirrhosis. [5]

Background

Iron overload, also known as haemochromatosis, indicates accumulation of iron in the body from any cause. The most important causes are hereditary haemochromatosis (HHC), a

genetic disorder, and transfusional iron overload, which can result from repeated blood transfusions.[6] A 35 year old female patient by name Mrs. XX, who is a house wife resident of kavali came to general medicine OPD at Narayana Medical College, Nellore with chief complaints of abdominal distension since 2 months. The patient was apparently normal 2 months ago Then she developed abdominal distension which is insidious in onset and progressive in nature with no aggravating and relieving factors. History of passage of black colored stools is present. No history of vomiting. No history of diarrhoea or constipation. No history of altered sensorium. No history of passage of blood in stools. No history of blood transfusions. No history of itching over the body. No history of fever. No history of alterations in sleep pattern. No history of cough or burning micturition.

Past history was positive for similar complaints 10 years back for which she had taken treatment. The patient had history of jaundice at the age of 15 years and she had taken medications for it following which it subsided. She had a second episode of jaundice at the age of 19 years for which she had taken ayurvedic treatment. She is not a known case of Diabetes Mellitus, Hypertension, Tuberculosis, Epilepsy, thyroid disorders, Coronary Artery Disease or Cerebral vascular accident. Personal history is she consumes mixed diet with history of decreased appetite since 2 months. She has history of passage of yellow coloured urine and history of passage of black coloured stools. No history of consumption of stored food. No history of similar complaints in the family members. In the Prenatal period regular antenatal checkups were done, no anomalies were detected during scans, no fever, iron folic acid supplementation were taken by mother. On general examination; weight 44 kgs and height of 1.47 m with BMI of 19.8 and Patient is moderately built and poorly nourished.

On examining for signs of liver cell failure

Patient had alopecia, icterus and muscle wasting,ankle edema. There is no hepatic encephalopathy, fetor hepaticus, parotid enlargement, spider naevi, loss of sexual hair, clubbing palmar erythema, asterexis, menstrual abnormality, dupytrens contracture. No signs of chronic cholestasis obstructive jaundice, (scratch marks and pruritus) and no stigmata of Tuberculosis.

On General Examination

Pallor, Icterus, Pedal edema are present and Cyanosis, Clubbing, Lymphadenopathy areabsent. The skin of thePatient is darkly pigmented

Vitals

B.P. 90/60 mm of Hg recorded on Right arm in supine posture, Pulse:78 bpm low volume regular rhythm no vessel wall thickening no radio radial and no radiofemoral delay, Respiratory rate:-20 cycles per min thoraco abdominal type, Temperature :- 97.5 degree Fahrenheit.

Systemic Examination

Oral Cavity:- lips, angle of mouth, teeth. tongue, oral mucosa, palate, tonsils, oral cavity have a darkened appearance.
Abdomen:- (INSPECTION) Abdomen is uniformly distended. The abdomen is globular shaped. Flank fullness is present. The umbilicus is Horizontally split. The skin over abdomen is smooth and glistening. The movement of abdomen with respiration is equal in all quadrants. There are no visible pulsations. There are dilated veins over the Right upper quadrant. There is no caput medusae. No visible peristalsis. No hernial orifices. No scars and sinuses. Ascitic fluid tap mark present over the Right side of abdomen.
Palpation:- No local raise of temperature. Girth of abdomen at the level of umbilicus is 84cms. The distance between xiphisternum and umbilicus is 18 cms. The distance between umbilicus and pubic symphysis is 17cms. The dist between anterior superior iliac spine and umbilicus is 19 cms on both sides. There is no tenderness guarding and rigidity and Liver span - 7cms
Organomegaly:- Spleen is palpable along with notch, smooth surface with consistency firm to hard, moving with respiration + and lower pole of spleen, directed towards Right illiac fossa. The Spleen span is 19 cms below the costal margin. There is no tenderness
Percussion:- Horse shoe shaped dullness, fluid thrill, shifting dullness are present
Auscultation:- Bowel sounds Present, No bruit, No venous hum, No succussion splash, No friction rub.
Other Systems; CVS:- S1 S2 +, Rs :- BAE + Normal vesicular breath sounds heard and CNS:- NFND
Investigations; CBP:- Haemoglobin- 8.8gm/dl, Total WBC count- 3700cells/cumm, Neutrophils :- 76%, Lymphocytes :-14%,Eosinophils :- 3%, Monocytes :- 7%, Basophils :- 0%, E.S.R.- 46mm/1 hr and Platelet count- 1,13,000 per cumm.

Liver Function Tests: Total bilirubin - 12.19 mg/dl, Direct bilirubin - 10.54mg/dl ,S.G.O.T. - 93 U/L,S.G.P.T. - 36 U/L, Serum alkaline phosphtase - 629 U/L, Total protein - 5.8 g/dl, Serum albumin - 2.2 g/dl, Globulin - 3.6 g/dl and A/G ratio - 0.6.
Electrolytes:- Sodium :- 132 meq/L , Potassium :- 3.7 meq/L, Chloride :- 98 meq/L, Random plasma glucose :- 95 mg/dl, Serum urea :- 26mg/dl
Haemogram:- Hb - 9.2 gm/dl, P.C.V. - 28 vol%, R.B.C. Count - 3.8 millions/cumm, M.C.V. - 79 Femto Litres , M.C.H. - 26 Pico grams, M.C.H.C - 32 %, R.D.W. - 27 %, Total count W.B.C. - 4500 cells/cumm, Neutrophils - 74%, Lymphocytes - 20%, Eosinophils - 5%, Monocytes - 5%,Basophils - 0%, Platelets count - 1,05,000 per cumm, E.S.R. - 30 mm/ hr, R.B.C.- Moderate anisopoikilocytosis, predominantly Microcytic Hypochromic, cells with some normal cells. Ovalocytes pencil shaped cells. Rare NRBC and Polychromasia seen. No immature cells and no haemoparasites seen,W.B.C.- TC and DC within normal limits. No toxic granules, Platelets - Decreased and Impression - Hypochromic Microcytic anaemia with thrombocytopenia
Ascitic Fluid Analysis:- Macroscopic Examination: Volume - 30ml, Colour - yellow and Appearance - clear fluid
Microscopic Examination: T.C. - 65 cells/cumm, D.C - Polymorphs - 2%, Lymphocytes - 55%, Mesothelial cells - 43%
Proteins: Total proteins - 1.57 gm/dl and Albumin - 1%
Ascitic fluid for A.D.A. 12 Units/L
S.A.A.G.:-1.2
Complete Urine Examination:- Colour :- Yellow, Appearance:- Clear, PH :- 6.0, Specific gravity :- 1.020, Sugar:- Nil and Albumin:- Nil
Microscopic Examination:- Pus cells:- 1-2 cells/ HPF, EP cells :- 1-2 cells/ HPF, R.B.C. :- Occasional, Crystals:- Nil, Casts:- Nil, Yeast:- Nil, Bacteria :- Nil and Others:- Nil
Impression:-
1. *Ultrasound Abdomen:*
Shrunken liver with coarse echotexture and surface irregularity and multiple eterogeneously
hypoechoic areas in parenchyma S/O Regenerative or dysplastic nodules.
2.Portal venous thrombosis at the level of porta.
3.Splenomegaly with multiple *Gamma Gandy bodies*, dilated and tortuous splenic vein and multiple splenic hilar collaterals.
4.Gross ascites
Upper Gastrointestinal Endoscopy:- Impression:-
Oesophageal varices, G.E. Junction and Duodenal Ulcers and Forrest III
Iron Studies:- SERUM IRON:- 45.8 micmol/L, Serum Ferritin:- 698 micgrams/L, T.I.S.P. :-90% (Transferrin Iron Saturation Percentage), Serum T.I.B.C.:- 40.3 micmol/L, Serum Transferrin:- 125mg/L

Discussion

Signs and symptoms: Organs commonly affected by haemochromatosis are the liver, heart, and endocrine glands. Haemochromatosis may present with the following clinical syndromes.[7] Cirrhosis of the liver: Varies from zonal iron deposition[1] to fibrosis (cirrhosis). Diabetes due to selective iron deposition in pancreatic islet beta cells leading to functional failure and cell death. Cardiomyopathy, Arthritis (calcium pyrophosphate deposition in joints), Testicular failure; Slate grey discoloration of the skin, Joint pain and bone pain. [8]
Diagnosis: Selective iron deposition in pancreatic islet beta cells is seen, There are several methods available for diagnosing and monitoring iron loading including:

- Serum ferritin
- Liver biopsy
- HFE
- MRI

Serum ferritin testing is a low-cost, readily available, and minimally invasive method for assessing body iron stores.[9] However, the major problem with using it as an indicator of iron overload is that it can be elevated in a range of other medical conditions unrelated to iron levels including infection, inflammation, fever, liver disease, renal disease, and cancer. Also, total iron binding capacity may be low, but can also be normal. Assessment of the hepatic iron index (HII) is considered the "gold standard" for diagnosis of haemochromatosis.[10]

Magnetic resonance imaging (MRI) is emerging as a noninvasive alternative to accurately estimate iron deposition levels in the liver as well as heart, joints, and pituitary gland. The typical physical findings are Patients with cirrhosis have a hyperdynamic circulation with tachycardia, a bounding pulse, and warm extremities.[11] An interesting finding is the presence of orthodeoxia, or increased breathlessness (with decrease in PaO2) in sitting or standing position but not while lying down. This defect is due to shunting of blood through collateral vessels, which are present in greater numbers in the lower lobes of the lungs. Signs of

early encephalopathy should be investigated, including poor recall. [12]

Treatment

Routine treatment in an otherwise-healthy person consists of regularly scheduled phlebotomies (bloodletting). When first diagnosed, the phlebotomies may be fairly frequent, perhaps as often as once a week, until iron levels can be brought to within normal range. Once iron and other markers are within the normal range, phlebotomies may be scheduled every other month or every three months depending upon the patient's rate of iron loading. Each session typically draws from 450 to 500 cc. For those unable to tolerate routine blood draws, there is a chelating agent available for use. The drug desferoxamine binds with iron in the bloodstream and enhances its elimination in urine and faeces. Typical treatment for chronic iron overload requires subcutaneous injection over a period of 8–12 hours daily.] Two newer iron chelating drugs that are licensed for use in patients receiving regular blood transfusions to treat thalassaemia (and, thus, who develop iron overload as a result) are deferasirox and deferiprone.

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