case report

Hepatic amyloidosis

*Dr. raghad jawad AL Alkyshee ,
**Dr. Ali J Hussien,
***Dr Nazar ,
****Dr Kittam AL Kafajee

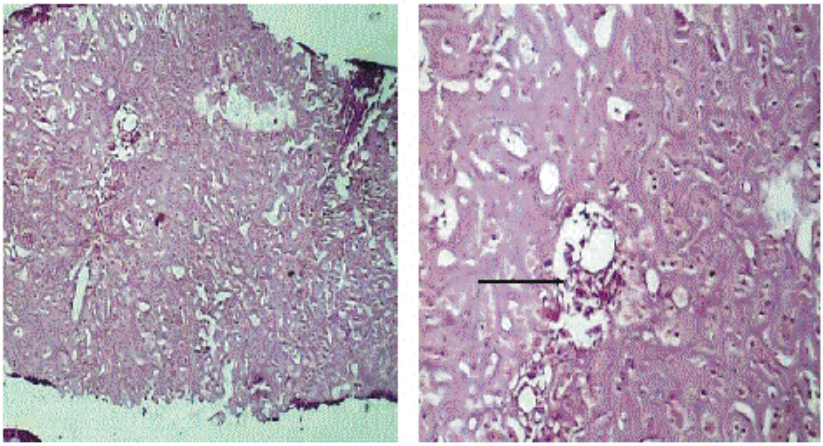
A 56 year old male patient presentwith progressive jundice of two months duration with pain in the right hypochondrial region which is contiuious dull in nature .he is free from other medical illness

On examination : patient is deeply jundice no perphral lymphadenopathy his puls ratewas 80 /min requlr and his blood pressure was 120/80 no dilated vien but mild leg edema.
Abdominal examination :hepatomegaly liver span is 20 cm firm in consistancy with splenomegaly 3 finger below costal margin
Chest and heart examination were normal
His investigation :

Fasting blood suger 6.1 mmol/L	n	6.1-9.9
Blood urea	3.5 mmol/L	3.3-7.5
Total sreum bilrubin (TSB)	382 mmol/L	5-17
Direct	310	
SGOT	35U/L	<40
SGPT	40 U/L	<45
ALP	700U/L	30-85 U/L

Prothrombin time 15.4 INR 1.2 ESR 70mm/h
S iron 25(70-180µ/dl total iron binding capacity 195 µ(250-410)
Virologlogical study for hepatitis B &C were negative
Abdominal ultrasound: gross hepatocyte normal texture normal biliary system spleen is enlarge 140 mm normal both kidney and no ascites
Chest X ray normal cardiac size and shape normal lung apart from evidence of right hemidiaghram elevation due to hepatomegaly
Liver biopsy :(fig-1) distorted liver architecture with deposition of eosinophlic hyaline extracellular material and atrophy of the hepatocyte special stain with congored is posative pictureis consistant with liver amyloidosis

*consultant in gastrinterstinal and hepatology teaching hospital
**specialist surgeon in AL karama hospital
***histopathologist in Baghdad teaching hospital
**** medical collage Baghdad university



Liver biopsy showing an abnormal homogenous material deposition causing atrophy of the hepatocytes (arrow pointing to the portal area).

After 2 weeks after liver biopsy her condition deterioated ascites developed her jundice increase albumin decrease INR become 4 he stated to have vomiting ,endoscopy was arranged which revealed esophageal vaies grade 2 portal hypertensive gastropathy prominent gastri fold by which biopsy was taken which revealed deposition of homogenous eosinophilic material in large submucosal blood vessel which eas stained positive for congored featured consisted with amyloidosis (fig-2).

There was no specific treatment was given because the patient is so tired he died after 2 months after the development of ascites.

Discussion Amyloidosis is a generic term that refers to the extracellular tissue depostion of fibrils composed of low molecular weight subunits (5 to 25 kD) of a variety of normal serum protine-1.

Gastrointestinal disease in amyloidosis results neuromuscular infiltration. In addition, an extrinsic autonomic neuropathy may also affect gut function.

Recognition of hepatic amyloidosis is difficult, since there are no path gnomonic signs of liver involvement.

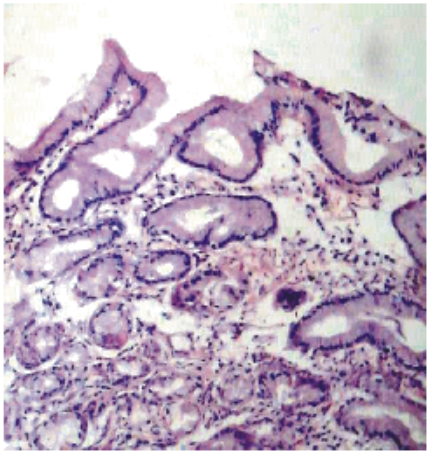
Hepatic parenchymal infiltration by amyloid was just as common in primary amyloidosis (62 per cent) as in secondary amyloidosis (59 per cent), without significant histologic or functional differences between these two forms 2.

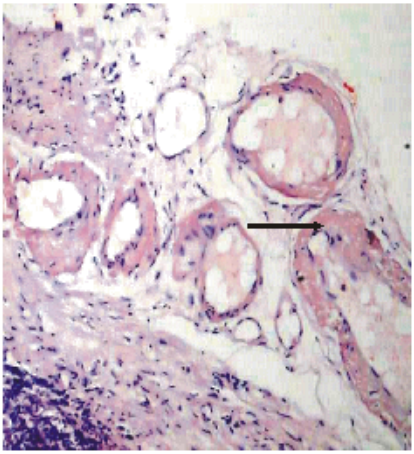
Although amyloid deposition in the liver is common in patients with systemic amyloidosis, clinical liver disease is relatively rare such patients generally have a poor prognosis , which probably reflects relatively severe disease [3,4].

Median survival was only nine months (five-year survival of 17 percent) in one of

The largest published series of 98 patient from the Mayo clinic seen between 1975 and 1997 independent predictors of shortened survival included heart failure, a total bilirubin>2mg/dL (34 micromol/L), and a platelet count >500,000/microl 5 .Hepatomegaly is noted on physical examination in 40% to 50% of patients with the more common forms of systemic amyloidosis (AL and AA) and is the most common hepatic manifestation of AL amyloidosis(16). Elevations of serum alkaline phosphatase values and hypoalbuminemia are common findings in patients with hepatic amyloidosis, with abnormalities of serum aminotransferases and elevations of serum bilirubin levels being less common(7).abnormal prothrombin times may also be present.

As there is no laboratory test capable of making a specific diagnosis of amyloidosis, a histologic diagnosis is required to confirm these suspicions. Liver biopsy has a high diagnostic yield in systemic amyloidosis. However, previous reports of significant hemorrhagic complications following needle biopsy of the liver or other orangs with amyloid deposits suggest that, when possible, diagnostic biopsies should be limited to sites accessible to local control of bleeding. Patients with hepatic amyloidosis almost invariably have involvement of other organ sites that are equally or more amenable to diagnostic biopsies. Needle aspiration of abdominal subcutaneous fat or endoscopic biopsy of stomach ,duodenum, or rectum are alternative approaches that have been recommended as having high diagnostic yield with lesser rates of life-threatening hemorrhage (9).However,this case like many cases of hepatic amyloidosis, patients present initially with hepatomegaly and liver test abnormalities and lack other clinical findings suggestive of systemic that nessisate taking liver biopsy five patients with sever cholestatic jaundice are described and an additional 20 From published reports are reviewed .hepatomagaly, usually marked, was present in 92% with ascites in 56% Of the cases . The serum bilirubin concentration was noticeably high and the serum globulin is low variety of treatment regimens was tried but median survival was only three months from the onset of jaundice (10).In such situations, liver biopsies may be appropriate if coagulation test results are in an accepeptable range and patients have not had prior evidence of bleeding dyscrasias. One review of bleeding manifestations in 100 patients with amyloidosis noted that all patients with hemorrhagic complications of diagnostic procedures had prior history of bleeding disorders, and a recent series reports a 4% bleeding rate with no fatalities following liver biopsy in 98 patients with hepatic AL amyloidosis. These findings suggest that although coagulation abnormalities complicate amyloidosis , other factors such as amyloid infiltration of blood vessel walls likely also play a major role in the propensity to hemorrhagic complications. The patient described here had cholestatic liver disease as the primary manifestation of primary systemic amyloidosis . Review of the literature suggests that prominent liver disease with cholestasis is unusual but probably under reported in patients with amyloidosis and appears to be restricted to patients with the primary fiom of amyloidosis. Nonetheless, cholestatic hepatic amyloidosis is characterized by distinct clinical,laboratory , and pathologic features, recognition of this process is critical because it identifies patients with widespread organ involvement portends a poor prognosis(10). Our patient gastric involvement with out doudenum which was notcied after the diagnosis of amyloidiosis was achived and the gastroscopy was done because of repeated vomiting .when the gastrointestinal tract is affected, the most common sites of infiltration are the descending duodenum(100 percent), the stomach and colorectum(more than 90 percent),and the esophagus(about 70 percent)(10). Patients with symptomatic gastrintestinal





Gastric biopsy showing mild chronic gastiritis with abnormal deposition of that was positive for) arrowes(eosinophilic material around blood vessels .amyloid stain

amyloidosis usually present with one of four syndromes: gastrointestinal bleeding, dysmotility, malabsorption, or protein-losing gastroenteropathy. Rare presentations include an obstructing mass and encapsulating peritonitis causing small bowel obstruction.

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