INTESTINAL FAILURE-ASSOCIATED LIVER DISEASE VS NON-ALCOHOLIC FATTY LIVER DISEASE IN SHORT BOWEL SYNDROME
A Case of Rapidly Progressive Hepatic Failure

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INTRODUCTION

- Intestinal Failure-Associated Liver Disease (IFALD) is a progressive disease with a high mortality rate in patients dependent on parenteral nutrition (PN).
- It is a multifactorial entity associated with a spectrum of hepatic manifestations including steatosis, cholestasis, portal hypertension choline deficiency and manganese toxicity. In patients with Short Bowel Syndrome (SBS),hepatic steatosis occurs in 40-55% ,IFALD in 5-15%.
- Risk factors include chronic PN use and length of remaining bowel.
- Early IFALD may present similarly to NAFLD, however pathogenic and prognostic differences make distinguishing these diseases crucial.
- We present a case of rapidly progressive IFALD in an adult SBS patient after PN use.

CASE DESCRIPTION

- A 65-year-old female with massive short bowel resection (< 30cm remaining) and malnutrition with a 12-year history of on-and-off PN use. PN was discontinued 6 months earlier and she was gaining weight with enteral feeding and teduglutide.
- She presented with right upper quadrant pain and nausea. Ultrasound showed cholelithiasis and hepatic steatosis. She was discharged after resolution of symptoms.

CASE DESCRIPTION(cont'd)

- In two weeks , she returned with jaundice, worsening abdominal pain, weight loss, altered mentation and asterixis. Labs showed total bilirubin 11.4 mg/d L (Direct 6.0mg/d L), ALP 139 IU/L, AST 136 IU/L, ALT 65 IU/L, NH3 182 mg/Land serum carnitine 15 µmol/L.
- She was treated for hepatic encephalopathy with lactulose, rifaximin and carnitine. Computed tomography showed moderate ascites, mesenteric edema, and edematous bowels . Diagnostic paracentesis revealed portal hypertension.
- **Liver biopsy showed cirrhosis with steatohepatitis and peri-cellular fibrosis consistent with TPN-associated liver disease in the setting of SBS-IF(Image)**. Multifocal-pneumonia with multi-organ failure led to her death.

IFALD vs NAFLD

IFALD	NAFLD
No Metabolic Syndrome	Metabolic syndrome common
Severe malabsorption mostly on PN	No Malabsorption and PN mostly not required
Low Plasma Choline levels, Choline supplementations reduces steatosis ,improves Liver tests	Normal to high Plasma Choline levels, therefore minimal difference to choline supplementation
Cholestasis highly evident with hyperbilirubinemia	Not typical
Macro and micro steatosis more common in zone 1 (periportal zone)	Predominantly macro steatosis mostly involving zone 3 (pericentral zone)
Characteristic "jig-saw" pattern fibrosis	Sinusoidal fibrosis with ballooning of hepatocytes
Rapid progression to ESLD ,cirrhosis develops within ~3-5 months after initiating PN	Longer duration approximately 10 to 20 years for cirrhosis to develop
Intestinal transplant is mainstay of treatment , Poor prognosis and Rapid onset of death within 1-4 years	No role of Intestinal transplant as no malabsorption , Comparatively better prognosis ,rapid death is rare

PATHOLOGY

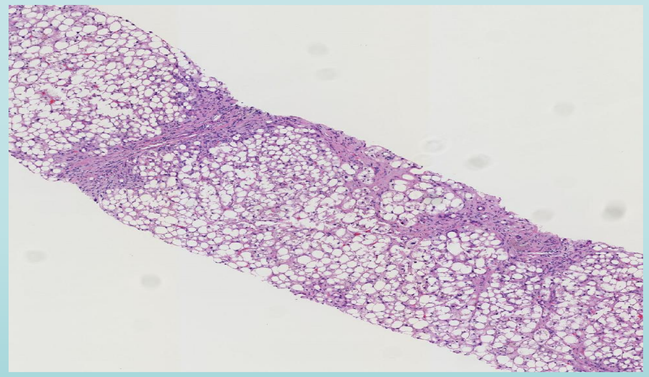

Image : Liver biopsy showing cirrhosis with marked steatohepatitis, mild lobular inflammation, ballooning hepatocyte degeneration nodular and peri-cellular fibrosis stage IV.

DISCUSSION

- This case demonstrates the potentially rapid progression of IFALD, particularly in patients with SBS.
- Clinicians should exercise high clinical suspicion of IFALD in patients with a history of PN use and SBS that present with hepatic manifestations.
- Early recognition is important to distinguish the disease from similarly presenting NAFLD.
- It is also vital to consider and treat other factors that may exacerbate hepatic disease including nutritional deficiencies.
- There may be benefit to diagnosis with liver biopsy early in the disease course to initiate prompt treatment or transplant, preventing rapid and fatal progression.