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An Unusual Cause For ExtraHepatic Portal Vein Obstruction– A Case Report

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Abstract

Extra Hepatic Portal Vein Obstruction is defined as“a vascular disorder of liver,characterized by obstruction of the extra-hepatic PV with or without involvement of intra-hepatic PV radicles orsplenic or superior mesenteric veins”. Etiological factors differ among pediatric and adult populations.Despite extensive history and laboratory work-up,many cases have been labelled idiopathic.However in our patient we were able to identify a unusual cause for EHPVO.

Keywords:ExtraHepatic Portal Vein Obstruction, Systemic lupus erythematosus

Introduction

Extrahepatic portal vein obstruction is a vascular disorder of the liver, which can results in obstruction and cavernous transformation of the portal vein with or without involvement of intrahepatic portal vein, splenic vein, or superior mesenteric vein.Portal vein obstruction due to chronic liver disease,neoplasm, or post surgery is considered a separate entity and is not the same as extrahepatic portal vein obstruction

Case report

A 13 year old female patient was admitted with abdominal distension and dragging abdominal pain, and multiple oral ulcers for one years and hematemesis and passing melena for past 2 days.

She was apparently normal before one year and she had no history of abdominal surgery, trauma or any drug intake. On general examination the patient had short stature pallor and thin built. She had multiple oral ulcers and angular stomatitis.

VITALS-BP-100/60 ,PR-112/min, regular,RR-18,Spo2-99% in room air Cardiovascular system-Apex at 5th Left intercostal space medial to mid clavicular line.S1 and S2 heard and systolic murmur of Grade 2/6 at apex and left sternal border. Respiratory system- Bilateral normal vesicular breath sounds, no added sounds Abdomen – Spleen palpable 12 cm below costal margins, firm in consistency, non tender, splenic notch felt, moves with respiration and no splenic bruit . Liver span- 5 cm. Central nervous system – clinically normal.Fundus- normal, No K-F ring.

Investigations-

CBC-Hb-4.4 g/dl,TC-1,100per microliter , PLATELET-77,000 per microliter, ESR-110mm/hr

RFT- UREA-45 mg/dl , S.CREATININE-0.9 mg/dl

LFT-TOTAL BILIRUBIN-0.9mg/dl,DIRECT-0.4mg/dl, INDIRECT-0.5mg/dl,SGOT-49 IU/L,SGPT-36IU/L. TOTALPROTEIN-8.1g/dl,S.ALBUMIN-5.0g/dl, S.GLOBULIN-3.1g/dl

URINE ROUTINE-albumin-trace, no deposits

HIV-NON REATICE, HBsAg and Anti HVC- Negative

PERIPHERALSMEAR-PANCYTOPENIA,NO ATYPICAL CELLS, RETICULOCYTE COUNT-0.5%

BONEMARROW-ERYTHROID HYPERPLASIA WITH MEGALOBLASTS AND MICRONORMOBLAST.MYELOID SERIES RELATIVELY REDUCED.NO BLAST OR ATYPICAL CELLS

XRAY CHEST- NORMAL

USG-

LIVER- NORMAL ECHOTEXTURE AND SIZE
POTRAL VEIN – CAVERNOUS MALFORMATION OF PORTAL VEIN WITH MULTIPLE PERIPANCREATIC AND SPLENIC HILAR COLLATERALS
SPLEEN- 19 CMS

OGD SCOPY-

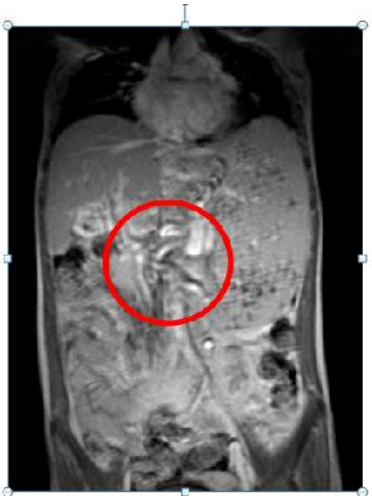
GRADE III – 2 COLUMN ESOPHAGEAL VARICES.

TESTS FOR AUTOIMMUNITY-

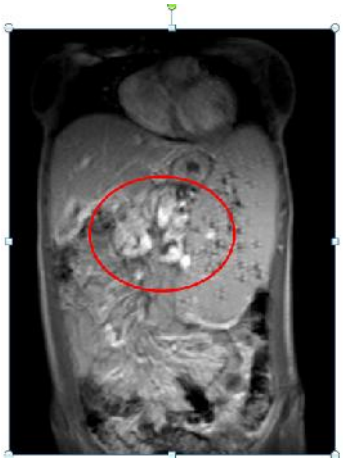
ANA – Positive
Anti- ds DNA- Positive
Anti-Smith- Positive
Other auto antibodies including lupus anticoagulant and anti-cardiolipin for APLAs- Negative

MRI ABDOMEN-

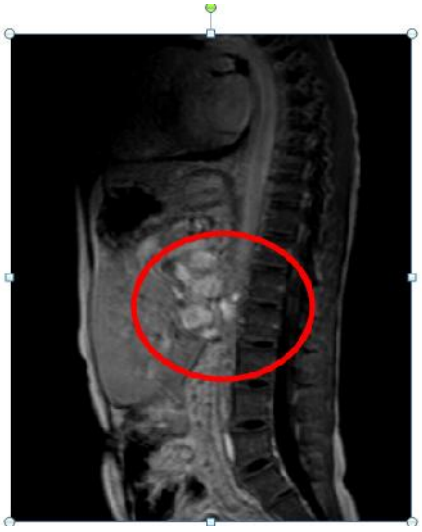
SPLEEN- 19.5 cm, with gamma gandy bodies, splenic infarct of size- 5x3 cm, Splenic Vein is dilated , tortuous and 8mm
PORTAL VEIN- is reformed distal to confluence with cavernous transformation of portal vein. There are multiple gastrosplenic, splenic hilar, linorenal collaterals
IMPRESSION- EHPVO WITH CAVERNOUS



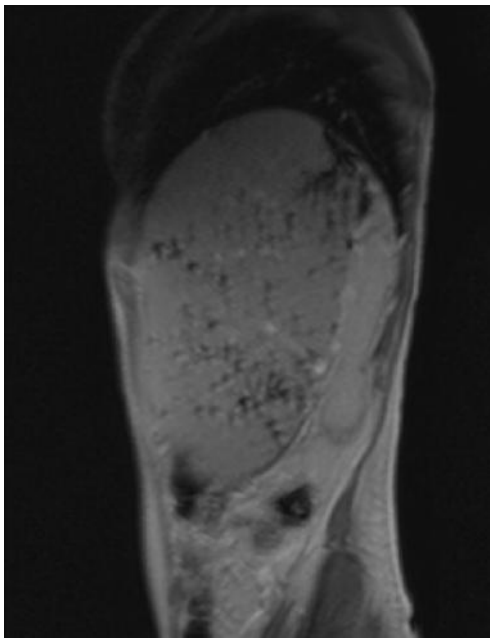
CORONALVIEW-PORTALVEINOBSTRUCTIONAND SPLENOMEGALY



CORONAL VIEW-CAVERNOUS MALFORMATION OF PORTALVEIN AND SPLENOMEGALY



LATERAL VIEW-CAVERNOUS MALFORMATION OF PORTAL VEIN AND SPLENOMEGALY



LATERAL VIEW-MASSIVE SPLENOMEGALY WITH GAMMA GANDY BODIES

Discussion

Cohen et al. reported that most patients with PVT are cirrhotics with primary or metastatic cancer; however, nontumoral and noncirrhotic PVT is the second most frequent cause of portal hypertension, worldwide.This constitutes approximately 5%–10% in the western world and 40% in the Indian subcontinent. Thrombophilic conditions are seen in 60% of PVT patients while local predisposing factors account for 30%. Usually there is more than one factor responsible for PVT. Despite an intensive workup, idiopathic groups still form 30% of PVT. Among the thrombophilic states, primary myeloproliferative disorders are more common causes of PVT.One should be aware of occult myeloproliferative disorders (MPD).Genetic variations in thrombin activatable fibrinolysis inhibitor (TAFI) gene, has been recently described as a risk factor forPVT.Much less commonly acquired disorders are anti phospholipid yndrome and paroxysmal nocturnal hemoglobinuria (PNH).Factor V Leiden mutation is the most common inherited thrombophilia predisposing factor to PVT followed by Protein C deficiency.The role of protein S and antithrombin (AT) III deficiency has not yet been confirmed in PVT.Thus factors associated with PVT discussed in literature are-

1.Systemic risks factors-

A.Inherited

- Factor V Leiden mutation-6%–32%
- Protein C/S deficiency –
  - Protein C deficiency- 0%–26%,
  - Protein S deficiency- 2%–30%
- Factor II mutation 14%–40%
- Antithrombin deficiency-0%–26%.

B.Acquired

- MPD-30%–40%
- Antiphospholipid antibody syndrome (APLA-S)-6%–19%
- PNH
- Hyperhomocystinemia-12%–22%
- Oral contraceptives and pregnancy
- Hyperhomocysteinemia.

Local factors-

Focal inflammatory lesions

- Diverticulitis
- Pancreatititis
- Tuberculus lymphadenitis
- Inflammatory bowel disease (IBD)
- Cytomegalovirus (CMV) infection

Injuries to a portal venous system by

- Surgery
- Transplantation
- Trauma
- Vascular procedures/tips.

Cirrhosis

Conclusion

In our case initially work up was directed towards establishing myelodysplastic syndrome. However the patient had no evidence in bone marrow. So further work up was done to establish the thrombophilic state. In our patient SLE was diagnosed based on Revised American College of Rheumatology Classification Criteria forSystemic Lupus Erythematosus as she met 4 out of 11. She was started on steroids and Esophageal variceal ligation was done.

There are several factors that increase the thrombosis risk in SLE patients. The most important risk factors are the Antiphospholipid Anti bodies (APLAs) and increased Inflammation and disease activity.In SLE Inflammation may affect several steps in blood coagulation like initiation, propagation, and regulation.It has been shown that inflammation induces the expression of tissue factors, an important step of the initiation of coagulation. The presence of inflammation reduces the fibrinolytic activity through upregulation of the production of plasminogen activator inhibitor (PAI). Further, the anticoagulant effect of the *protein C pathway is impaired due to downregulation of thrombomodulinand a decrease of protein S*. This might explain the reason of the occurrence of thrombosis early in lupus patients. Therefore, SLEis can be considered an independent risk for thrombosis and can predispose to EHPVO and SLE presenting as EHPVO without other classical cutaneous feature and signs is uncommon.

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