CASE REPORT

RADIOLOGICAL FEATURES AIDING IN EARLY DIAGNOSIS OF OBLITERATIVE PORTAL VENOPATHY: A CASE REPORT

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ABSTRACT

Obliterative portal venopathy (OPV) is one of the major causes of non-cirrhotic portal hypertension. This condition is erroneously diagnosed as cirrhotic portal hypertension secondary to chronic liver disease. Histologically characterized by obliteration of small portal venous branches and a resultant increase in splanchnic circulation causing portal hypertension. Liver functions are well preserved. Symptoms primarily occur due to variceal bleeding. OPV has distinct early and advanced stages: its late stage is marked by hepatic de-compensation which poses a diagnostic hurdle mimicking cirrhotic hepatic de-compensation on imaging. Although OPV is diagnosed on biopsy however certain imaging characteristics can be attributed to this condition along with lab and history co-relation which are discussed in detail in the case reported here.

Keywords: Obliterative portal venopathy(OPV),non-cirrhotic portal hypertension, Hepatocellular carcinoma (HCC).

Introduction

Non-cirrhotic portal hypertension can result from multiple causes at pre-hepatic, hepatic, and post-hepatic levels.[3] Among many obliterative portal venopathy commonly occurs. Previously more documented in south-east Asia now also reported frequently in west and America.[3] This entity is diagnosed on histology specimen of liver core biopsy with specimen size at least up to 20 mm characterized by portal tracts with fibrosis, sclerosis of portal vein branches along with thrombosis. There is associated regenerative nodular hyperplasia.[1] OPV is itself caused by immunosuppressed conditions like HIV, certain drugs like didanosine,[1] and hyper-coagulable and pro-thrombotic states. On imaging, there is marked splenomegaly, gastric varices and less often with ascites. Most of the time it goes unnoticed and the patient is misdiagnosed as a case of cirrhotic portal hypertension because of the overlap of imaging features. However, a good correlation with history and labs can warrant early biopsy to diagnose the condition.

Case Report

This was a biopsy proven case of obliterative portal venopathy(OPV). A fifty-five-year-old female patient from a rural area got initial treatment by quack for hemmorhoidal bleeding which is a rare presentation of OPV. Later she developed left upper quadrant dull pain and heaviness. Her ultrasound reveals chronic hepatic parenchymal disease with portal hypertension and marked splenomegaly. Her treating gastroenterologist performed an upper GI endoscopic examination revealing grade I, II, and III gastric and lower esophageal varices and band ligation was done. Despite

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these findings, her LFTs were within normal range. She was HBV and HCV negative. A laborious lab workup was done to rule out the cause of CLD. This includes ANA, ASMA, and AMA, along with serum ceruloplasminand alpha-1 antitrypsin levels revealing nothing. Her CBC shows decreased hemoglobin 7 g/dL (N 11 - 14.5) and platelets 64000 (1,50,000-4,50,000) and decreased WBC counts 2900 (N 4000-11000). These findings were suggestive pancytopenia reflecting hypersplenism. A liver core biopsy was performed with a 1.2 x 0.1 cm specimen size showing loss of portal veins, dilated sinusoids, and focal congestion. There were no features of cirrhosis. The only variation in thickness of trabeculae suggests nodular regenerative hyperplasia. All these features were suggestive of obliterative portal venopathy.

With the above-described record, she was referred to our department for Computed tomography examination for further co-relation. Tri-phasic CT examination of the abdomen was performed using a Canon 164 slice helical Computed tomography scanner. CT study reveals normal sized liver. Liver margins were smooth with atrophy of the IVth segment and widened gallbladder fossa. (Fig.3b) Extra-hepatic portal vein calcifications were noted. (Fig.1) Arterial phase shows lengthened, a tortuous hepatic artery in the hilum. (Fig.2) No intra-hepatic arterial lesion was noted. Portal venous phase shows dilated tortuous, calcified extra-hepatic portal vein. (Fig.1,4). The intra-hepatic main portal vein branches were not visualized at routine and MIP (maximum intensity projection) images. Linear hypodense areas suggesting intra-hepatic portal vein thrombosis (Fig.3a). These features are radiological clue which supports the histological diagnosis of obliterative portal venopathy. IVC and hepatic veins were normal. Lower esophageal and gastric varices were viewed. (Fig.3a,4) Varices were also noted at the splenic hilum. There was marked splenomegaly measuring up to 18 cm. (Fig.4) Mild ascites was noted. Additionally dilated veins were also noted in the bilateral adnexal region suggesting pelvic congestion. Patient lost to follow up.

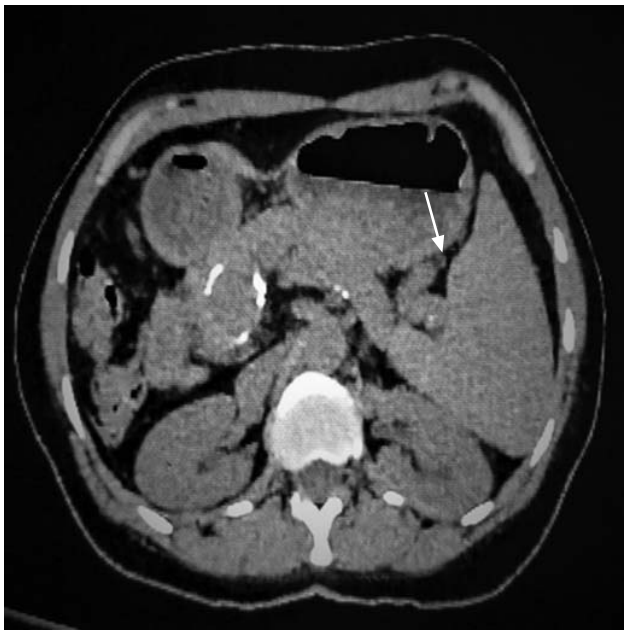

Figure 1: Plain image shows dilated tortuous, calcified extra-hepatic portal vein (arrow).

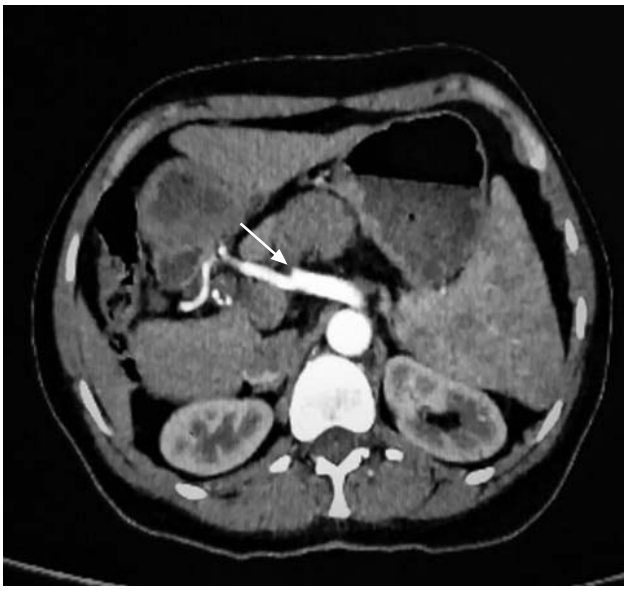

Figure 2: Arterial phase tortuous hepatic artery in the hilum (arrow)

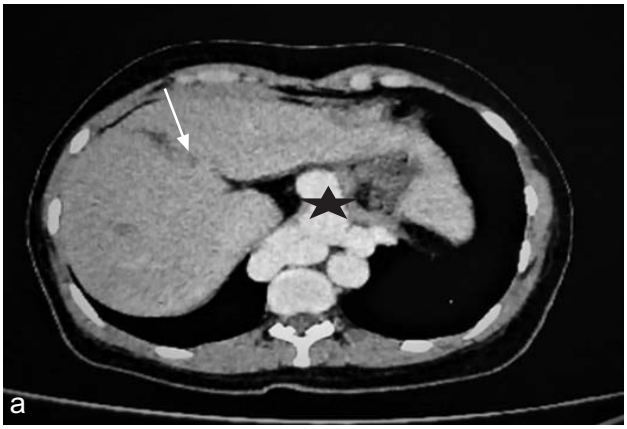

Figure 3a: Axial portal venous MIP image shows thrombosed intrahepatic portal vein (arrow) and non-opacification of intrahepatic portal veins. Esophageal varices (star).

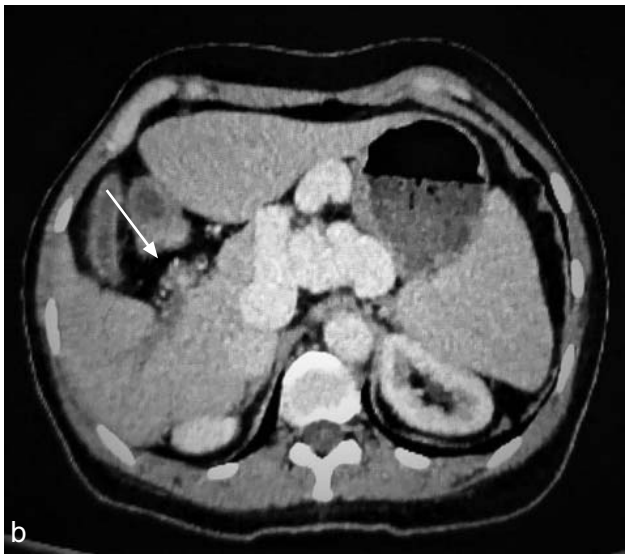

Figure 3b: Widened gall bladder fossa (arrow)

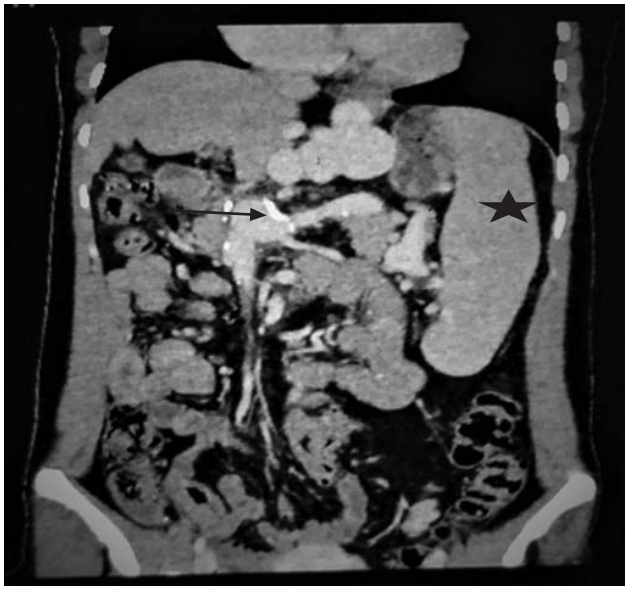

Figure 4: Coronal MIP image.
Dilated tortuous, calcified extra-hepatic portal vein (arrow). Enlarged spleen (star)

Discussion

Obliterative portal venopathy (OPV) was previously known as idiopathic portal hypertension or hepatic sclerosis or non-cirrhotic portal fibrosis.[3] Recently a new trend is proposed for this condition Porto-Sinusoidal Vascular Disease (PSVD) by European Association for Vascular Diseases (VALDIG).[1]
It primarily affects young adults of 30-40 years. Despite the severity of portal hypertension in this condition, the overall 10-year survival rate is 86% - 95%.[3] Among many causes of this condition mostly encoun-tered one is a hypercoagulable state. In one of the reported cases of obliterative portal venopathy (OPV); the cause was a prothrombotic state because of the patient taking oral contraceptive pills.[2]
In our case no other significant history was found as a leading cause of OPV except for probable pro-thrombotic state during pregnancies , as patient was gravida 7 para 5 with early neonatal death of twins. Pregnancy is a hyper-coagulable state per se aug-mented by harsh dry environment and undocumented blood loss. The patient was a non-smoker, non-alcoholic. She had no personal or family history of liver disease or any other genetic condition. No immu-nosuppressive state. There was no other significant drug history.
Computed tomography findings in this case pointing towards the obliterative portal venopathy are complete non-visualization of intrahepatic portal vein branches in the MIP portal venous phase. There were some hypoattenuating intra-hepatic areas suggesting intra-hepatic portal vein thrombosis. In contrast extrahepatic portal vein was massively dilated and calcified sug-gesting chronicity. This CT finding is an established one and is published in various articles.[3,1,7] Most common reported finding however is the rapid tapering of medium-sized portal vein branches and abrupt cut-off with or without thrombosis.[3] Our case shows a slightly advanced stage of the disease. Portal vein calcification is also commonly reported in OPV as in our case. Segment IV atrophy was found along with normal sized caudate lobe in contrast to cirrhosis where there is hypertrophy of caudate and left lobe. Well-preserved liver size and smooth liver margins also favours OPV. OPV is associated with both gastric and ano-rectal varices. Although bleeding from ano-rectal varices are not quite common but is confused with hemorrhoids as in our case. Spleen is more mar-kedly enlarged in OPV as compared to cirrhotic portal hypertension along with hypersplenism which is reflected by pancytopenia and decreased platelet counts.
Ascites is a rare finding although can develop in advanced stages of OPV as in our case. Liver func-tions are well preserved until very late in the disease process. There is a low but definite chance of hepatic

decompensation and development of HCC as reported by Homsi and friends.[4]

Conclusion

Non- visualization of intra-hepatic branches of the portal vein in the background of normal liver is a predictor of a cause other than cirrhotic liver disease causing portal hypertension. It is for the benefit of a patient if the radiologist keenly views the portal vein and its intrahepatic branches on MIP images to early pinpoint the cause in the context of normal LFTs and other lab findings.

Conflict of Interest: Declared none by authors

References

1. Glatard, A. S., Hillaire, S., d Assignies, G., Cazals-Hatem, D., Plessier, A., Valla, D. C., &Vilgrain, V. Obliterative portal venopathy: findings at CT imaging. Radiology 2012; **263(3):** 741-50.

2. Ashraf M. F., Trifonova, R, Batool, A. Obliterative Portal Venopathy Caused by Oral Contraceptive Pills: A Case Report. Journal of Medical Cases 2021; **12(11):** 446-50.

3. Rajesh S, Mukund A, Sureka, B, Bansal K, Ronot, M, Arora A. Non-cirrhotic portal hypertension: an imaging review. Abdominal Radiology 2018; **43(8):** 1991-2010.

4. El Homsi M, Fiel M. I., Taouli B. Hepatocellular carcinoma in obliterative portal venopathy. Diagnostic and interventional imaging 2021; **102(2):** 115-6.

5. Gioia S., Riggio O, Nardelli S, d Amati, G., Ridola L. Identifying Patients at High Risk of Developing Non-Cirrhotic Portal Hypertension. Hepatic Medicine: Evidence and Research2021; **13:** 105-09.

6. Kmeid M, Liu X, Ballentine S, Lee H. Idiopathic non-cirrhotic portal hypertension and porto-sinusoidal vascular disease: review of current data. Gastroenterology Research 2021; **14(2):** 49-53.

7. Arora A, Sarin S. K. Multimodality imaging of obliterative portal venopathy: what every radiologist should know. The British Journal of Radiology 2015; **88(1046):** 20140653.

8. Cazals-Hatem D, Hillaire S, Rudler, M., Plessier A, Paradis V, Condat B, Valla, D. C. Obliterative portal venopathy: portal hypertension is not always present at diagnosis. Journal of hepatology 2011; **54(3):** 455-461.

9. Aggarwal S, Fiel M I, Schiano T D. Obliterative portal venopathy: a clinical and histopathological review. Digestive diseases and sciences 2013; **58(10):** 2767-76.

10. Patel K R, HarpavatS, Khan Z, Dhingra, S, Quintanilla N, Firan, M, et al. Biliary atresia patients with successful Kasai portoenterostomy can present with features of obliterative portal venopathy. Journal of pediatric gastroenterology and nutrition 2020; **71(1):** 91-98.