Section: Ophthalmology

Case Report

NEW CLUES TO CATCH AN OLD CULPRIT - CENTRAL RETINAL VEIN OCCLUSION IN A CASE OF BUDD CHIARI SYNDROME

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ABSTRACT

Background: Budd-Chiari syndrome (BCS) is a rare heterogeneous liver disease that is characterised by obstruction of the hepatic venous outflow tract, which may occur anywhere leading to an hypercoagulable state sometimes even involving the retina. We present this case due to its rarity and the need for regular fundus evaluation in such patients. **Materials and Methods:** This report describes a case of a 47 year old female who was already diagnosed with Budd Chiari syndrome presented with sudden onset visual loss. **Result:** Fundus examination of right eye was normal and left eye showed ischemic central retinal vein occlusion with cystoid macular edema. **Conclusion:** A patient with vein occlusion should be evaluated for systemic parameters and for possibility of hypercoagulable nature.

INTRODUCTION

Budd- Chiari syndrome (BCS) is a rare congestive hepatopathy caused due to blockage of hepatic veins. The impaired drainage causes congestion in liver which leads to damage to hepatocyte and finally leads to fibrosis and end stage liver disease. The obstruction can be thrombotic or non-thrombotic. It could be anywhere along the venous course from the hepatic venules to junction of the inferior vena cava (IVC) to the right atrium. It is the hypercoagulable state that has caused central retinal vein occlusion in our patient. We present this case due to its rarity and the need for regular fundus evaluation in such patients.

CASE REPORT

A 47-year old female, with ascites and hepatomegaly (decompensated liver disease), who is already a known case of Budd-Chiari syndrome, presented with a one week history of left eye sudden onset and profound decreased visual acuity. Specifically, she had no history of superficial or deep venous thrombosis and no history of thromboembolic events. Her serum alanine aminotransferase (ALT) was 256 U/L (upper limit of normal < 50U/L). Abdominal sonographic imaging revealed an enlarged liver with ascites. Her blood reports revealed protein C deficiency, suggesting primary Budd-chiari syndrome. She had already undergone direct intrahepatic portocaval stent. She was started on oral anticoagulants in the past 6 months. Visual acuity at presentation was 6/6 in the right eye, and in 1/60 the left eye. Anterior segment examination showed grade1 nuclear sclerosis in both eyes. Fundus examination of right eye was normal and left eye showed ischemic central retinal vein occlusion with cystoid macular edema.

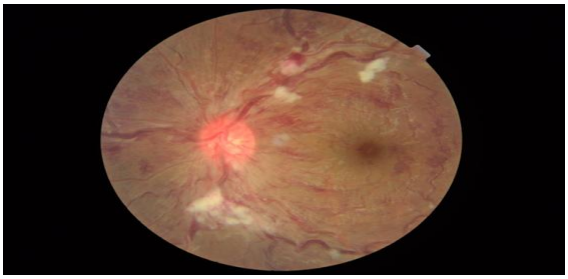

Figure 1: Fundus Image of Left Eye Revealed Ischemic Retinal Vein Occlusion (White Arrows) with Cystoid Macular Edema

SD-OCT of the left eye shows cystoid macular edema with subretinal serous detachment.

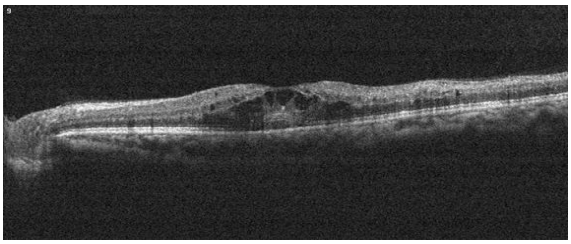

Figure 2: Sd-Oct of Left Eye Showing Cystoid Macular Edema with Serous Retinal Detachment

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DISCUSSION

BCS is a thromboembolic disease with obstruction in the hepatic venous system due to non cardiac causes. This is usually a triad with ascites, hepatomegaly and abdominal pain. It is a rare disorder but affects middle age male or female with equal preponderance.[1]

This hepatoreno occlusive disease is mostly due to obstruction by thrombus. Based on the pathology, the disease is classified as primary if obstruction is endovascular and secondary if there is obstruction from outside. Primary BCS is mainly due to thrombus caused due to hypercoagulable states like myeloproliferative disorder, protein c or s deficiency, hyper homocysteinemia, factor 5 Leiden mutation, presence of anticardiolipin antibodies and antithrombin 3 deficiency.[2] Secondary BCS can be caused due to compression from tumors that arise from adjacent structures.[3]

The obstruction of the venous flow from liver to the right atrium causes increased portal vein and hepatic sinusoid pressures. This causes ascites in the abdomen and collateral venous flow through alternative veins leading to esophageal, gastric and rectal varices. Obstruction also causes compression of hepatocytes leading to centrilobular necrosis and peripheral lobule fatty change due to ischemia. This circardial changes causes renal and liver failure which can be potentially lethal.

Behcets disease (BD) is a rare chronic multisystem inflammatory disorder affecting the eyes and other organs. BCS has been reported as a rare complication of BD. The vascular involvement is reported to be 7-29%.[4] Endothelial dysfunction resulting from vascular inflammation is considered to be an important factor of thrombosis.[5] Myeloproliferative neoplasm (MPN) is the most common etiology of BCS and advances in genetic technology helps us to diagnose latent MPN as well. BCS patients are now treated by endovascular intervention and anticoagulation. The survival rates have improved with these advances. Liver transplantation is reserved for select indications.[6]

On the other hand not much reports have been published regarding the ocular manifestations of BCS. The cause of vein occlusion in our case is due to the hypercoagulable nature of the disease. Apart from regular blood monitoring for hypercoagulable states and maintaining their coagulation profile with oral anticoagulants, these patients need regular monitoring of possible thrombosis in other sites especially retinal vein occlusions. This case is presented due to its rarity and also to emphasise the need for regular eye examination.

CONCLUSION

To conclude a patient with vein occlusion should be evaluated for systemic parameters. The patient should be tested for the causes of hypercoagulable nature.

REFERENCES

1. Rajani R, Melin T, Björnsson E, Broomé U, Sangfelt P, Danielsson Å, et al. Budd-Chiari syndrome in Sweden: epidemiology, clinical characteristics and survival – an 18-year experience. Liver International. 2009;29(2):253-9.
2. Garcia-Pagán JC, Valla D-C. Primary Budd–Chiari Syndrome. New England Journal of Medicine. 2023;388(14):1307-16.
3. Aydinli M, Bayraktar Y. Budd-Chiari syndrome: etiology, pathogenesis and diagnosis. World J Gastroenterol. 2007;13(19):2693-6.
4. Kuzu MA, Ozaslan C, Köksoy C, Gürler A, Tüzüner A. Vascular involvement in Behçet's disease: 8-year audit. World J Surg. 1994;18(6):948-53; discussion 53-4.
5. Lee YJ, Kang SW, Yang JI, Choi YM, Sheen D, Lee EB, et al. Coagulation parameters and plasma total homocysteine levels in Behcet's disease. Thromb Res. 2002;106(1):19-24.
6. Sharma A, Keshava SN, Eapen A, Elias E, Eapen CE. An Update on the Management of Budd–Chiari Syndrome. Digestive Diseases and Sciences. 2021;66(6):1780-90.

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