CASE REPORT

BUDD CHIARI SYNDROME

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ABSTRACT

BACKGROUND: Budd-Chiari Syndrome is a rare syndrome characterised by post hepatic obstruction leading to various complications **OBJECTIVE:** To describe a case of chronic Budd-Chiari Syndrome, stable for more than two decades without any major complications **METHODS:** Detailed history, physical examination and laboratory investigations. **CONCLUSION:** Commonly, Budd-Chiari Syndrome can lead to back pressure changes and cirrhosis of liver with its sequel. However, sometimes, the obstruction is too small or partial enough to cause some complications but not the full fledged syndrome leading to a stable state

Key Words: Budd-Chiari Syndrome, Cirrhosis of liver

INTRODUCTION

Budd-Chiari Syndrome is a syndrome of post hepatic obstruction to the outflow channel of liver. Obstruction can occur anywhere from hepatic sinusoids to hepatic veins to inferior vena cava upto the right heart. Causes of Budd-Chiari Syndrome includes any hypercoagulable states like pregnancy, anti-phospholipid syndrome, smoking, hereditary thrombophilias, amongst many. Symptoms vary from being asymtomatic to development of cirrhosis of liver and its complications. Diagnosis is by ultrasound angiography to look for hepatic sinusoids and MRI of liver with hepatic veins and inferior vena cava. Treatment is to relieve the congestion of liver, either by bypass or by stent placement.

CASE REPORT

A 50 year old post menopausal female from lower socioeconomic strata presented to the outdoor patient department of Sir SayajiRao General Hospital, Vadodara from tribal area near Ujjain, Madhya Pradesh. She presented with chief complaints of abdominal distension for the last 20 years and on and off abdominal pain also for the last 20 years. She also had on and off swelling of both lower limbs for the last 20 years, dilated veins over the back and front of abdomen for last 15 years and anorexia and early satiety for 20 years. She had a past history of yellowish discoloration of urine and sclera 20 years ago, which was related to the onset of these symptoms. She never had symptoms of liver decompensation including malena, hemetmesis or bleeding per rectum. She has hypopigmented patches over skin of cheeks, back and soles since childhood. Her family and personal history were insignificant. She was 10 years post menopausal during presentation. She conceived four times and had three alive children and one intrauterine fetal death at 7 months. There was no history of blood transfusion.

During presentation, she was vitally stable. No parotid enlargement, hair loss, spider naevi, dupuytren contracture or palmer erythema was noted. On per abdomen examination, she had dilated and tortuous veins present over upper part of abdomen

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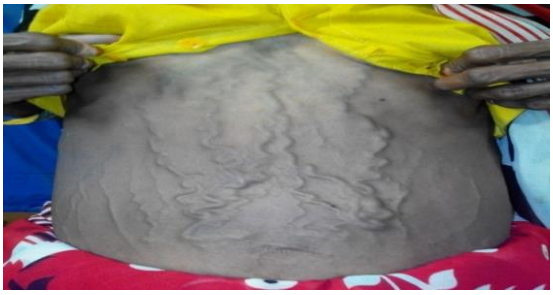
and upper back. Umbilicus was shifted downwards and inverted. On milking the veins over abdomen, they filled upwards from below. Rest of the system examination was within normal limits.



INVESTIGATIONS

PARAMETER	VALUE
Haemoglobin	12.00 gm%
Total Counts	6000 per cu. Mm
Differentials	70 / 28 / 01 / 01
Platelet counts	1.31 lakh per cu. Mm
ESR	22 mm in one hour
Smear	Normocytic normochromic RBCs
Blood urea	50 mg/dl
Serum creatinine	0.8 mg/dl
Bilirubin Total	1.4 mg/dl
SGPT	20 U/L
SGOT	42 U/L
ALP	91 IU/L
Serum sodium	136 mmol/L
Serum potassium	4.5 mmol/L
Total protein	8.0 gm/dl
Serum albumin	4.8 gm/dl

Chest X-Ray	Normal
RBS	122 mg%
ECG	Grossly within normal limits
Serum TSH	4.61
Serum ANA	Negative

ASCITIC FLUID ANALYSIS	
Total cells	320 per cu. Mm
Differentials	60 / 40
Total proteins	3.6 gm/dl
Sugars	94 mg/dl

Prothrombin time	14.80
INR	1.060
Activated partial thromboplastin time	30.00
HIV	Negative`
HBsAg	Negative
HCV	Negative

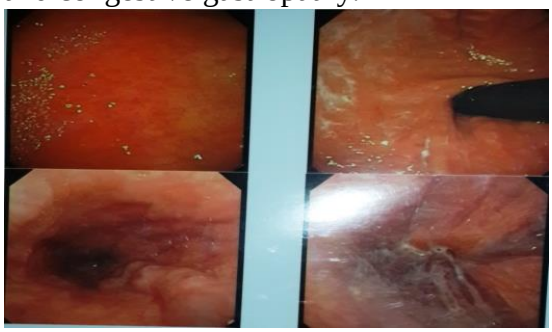
PARAMETER	VALUE	NORMAL VALUE
Protein C level	0.66 units/ml	0.55 – 1.11 unit/ml
Protein S level	0.92 units/ml	0.60 – 1.13 unit/ml
Anti thrombin III level	0.23 g/L	0.19 – 0.31 g/L
Factor V laden	4.6 unit/L	2.0 – 10.0 unit/L

Ultrasound abdomen: Liver span 126 mm with altered surface irregularity suggestive of cirrhosis of liver. Spleen 122 mm and moderate free fluid in abdomen.

Ultrasound liver screening: Occlusion of terminal inferior vena cava (IVC). Entire IVC shows reversal of flow. Both iliac veins show flow reversal. Right hepatic vein patent, dilated and sole outflow channel of the liver. Left hepatic vein patent but its ostium is occluded. Flow drains into right hepatic vein through a prominent collateral running over liver surface.

Contrast enhanced CT abdomen: enlarged caudate lobe and left lobe of liver with surface nodularity represents cirrhosis of liver. Multiple homogenously enhancing nodules of varying sizes in both lobes represents regenerating nodules. Marked narrowing of intrahepatic IVC seen. Middle hepatic vein is not visualised. Right and left hepatic veins visualised. Intra-hepatic veno-venous collaterals seen. Multiple abdominal wall, paraspinal and perioesophageal collaterals seen. Findings suggestive of Budd-CHiari Syndrome.

Upper gastrointestinal endoscopy is suggestive of grade II oesophageal varices and congestive gastropathy.



DISCUSSION

As stated above, Budd-Chiari Syndrome can present as an acute condition or a chronic condition. Chronic Budd-Chiari Syndrome is mostly a benign condition which after years of stagnant asymptomatic disease can present as a decompensated cirrhosis of liver. However, there are cases where the disease process is too trivial to cause the back

pressure changes and hence the development of liver parenchymal complications and its sequel is delayed for decades together or do not manifest at all. As per the texts, a case of chronic Budd-Chiari syndrome should be operated even after the cirrhosis is set in, in form of bypass grafting called TIPS (Transvenous intrahepatic portosystemic shunts) or in form of stenting and making an artificial window bypassing the obstruction. When the obstruction is diffuse or elongated, as in this case which involves the inferior vena cava, it becomes difficult to insert a stent. Treatment of such a case would rest only on prevention of decompensation of liver. The message here by highlighting this patient is simply that all cases of decompensation of liver (mild) with cirrhosis of liver does not require active intervention and only close follow up with patient education should be done.

REFERENCES

1. Tilanus HW. Budd-Chiari syndrome. Br J Surg [Internet]. 1995 Aug [cited 2017 May 12];82(8):1023–30. Available from: http://doi.wiley.com/10.1002/bjs.1800820807
2. Ferral H, Behrens G, Lopera J. Budd-Chiari Syndrome. Am J Roentgenol [Internet]. 2012 Oct [cited 2017 May 12];199(4):737–45. Available from: http://www.ajronline.org/doi/10.2214/AJR.12.9098
3. The Budd–Chiari syndrome: a review.
4. Hepatic vein thrombosis (Budd–Chiari syndrome).
5. Etiology, management, and outcome of the Budd–Chiari syndrome.
6. Rajani R, Melin T, Björnsson E, Broomé U, Sangfelt P, Danielsson A, et al. Budd-Chiari syndrome in Sweden: epidemiology, clinical characteristics and survival - an 18-year experience. Liver Int [Internet]. 2009 Feb [cited 2017 May 12];29(2):253–9. Available from: http://www.ncbi.nlm.nih.gov/pubmed/18694401
7. Budd-Chiari Syndrome: Practice Essentials, Background, Pathophysiology [Internet]. [cited 2017 May 12]. Available from: http://emedicine.medscape.com/article/184430-overview
8. Aydinli M, Bayraktar Y. Budd-Chiari syndrome: etiology, pathogenesis and diagnosis. World J Gastroenterol [Internet]. 2007 May 21 [cited 2017 May 12];13(19):2693–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/17569137
9. Plessier A, Valla D-C. Budd-Chiari Syndrome. Semin Liver Dis [Internet]. 2008 Aug 23 [cited 2017 May 12];28(3):259–69. Available from: http://www.thieme-connect.de/DOI/DOI?10.1055/s-0028-1085094
10. Wang Z-G, Jones RS. Budd-Chiari syndrome. Curr Probl Surg [Internet]. 1996 Feb [cited 2017 May 12];33(2):81–211. Available from: http://linkinghub.elsevier.com/retrieve/pii/S0011384096800013