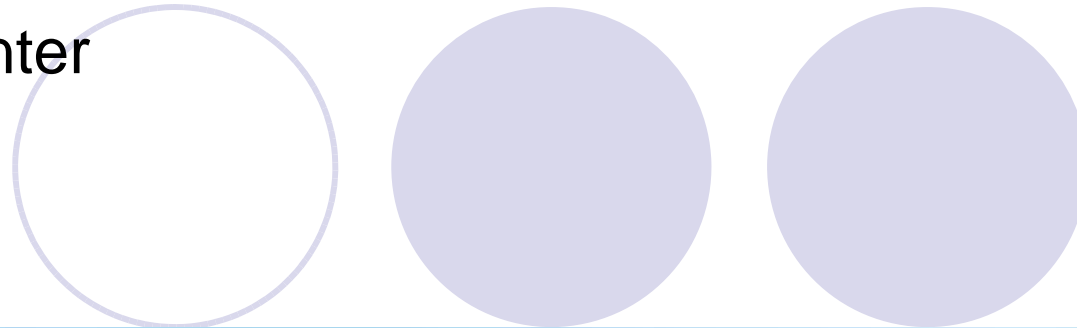


Clinical presentation

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Case summary



- 74 yo. male, resident of a nursing home facility.
- Was referred from the nursing home because of Abdominal pain, nausea, and enlargement of a known ventral hernia, confusion.
- Hx: GERD, BPH, s/p colectomy D/T diverticulosis, ventral hernia, S/P DVT, psychiatric problems

Initial presentation

- The patient was alert, afebrile, normal vital signs.
- On exam he had –
 - No significant JVP
 - enlarged abdomen, non tender, sus Ascites, +3 bilateral leg edema. No encephalopathy.
- Initial ER - CT - fatty liver – normal size, large amount of ascites, no splenomagly
- Admitted – DD: Cirrhosis vs Rt. H.F

Blood test



	11/09/2009	14/09/2009		11/09/2009	14/09/2009
glucose	76	57	Hb	16.6	16.40
Cl	105	103	RBC	5.90	6.2
Na	139	137	HCT	50	49
K	4.36	4.23			
BUN	22	28	PLT	275	382
Cr	0.81	1.51	WBC	13.1	16
			NEUT	70%	73%
T.Billi	2.7	3.4			
direct	1.4	2.3	PT	20	23
GPT	30	44	PTT	46.2	45
GGT	133	92	INR	1.83	2.13
LDH	752	940			
GOT	80	241	CRP	55	71
AlkP	194	216			
			AMONIA	11.3	263
Protein	64		B12	2000 <	
albumin	22				
globulin	42				

Peritoneal fluid

WBC	100
PH	8.5
Alb	5
amylase	18
Glu	18
LDH	133
protein	12
SAAG>1.1	

Table Causes of ascites, by SAAG level

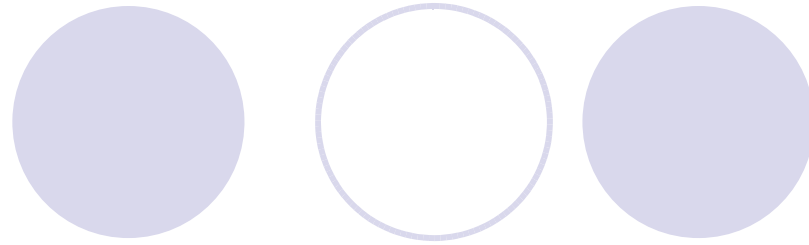
High gradient, SAAG >1.1 g/dL	Low gradient, SAAG <1.1 g/dL
<i>Liver disease</i>	<i>Infectious peritonitis</i>
Alcoholic hepatitis	HIV-associated bacterial, tuberculous, or fungal infection
Budd-Chiari syndrome	
Cirrhosis	
Fatty liver of pregnancy	<i>Malignancy</i>
Fulminant hepatic failure	Hepatocellular carcinoma
Hepatic congestion	Peritoneal carcinomatosis
Massive liver metastasis	Primary mesothelioma
Portal vein thrombosis	Pseudomyxoma peritonei
<i>Cardiac disease</i>	<i>Hypoalbuminemia</i>
Congestive heart failure	Nephrotic syndrome
Constrictive pericarditis	Severe malnutrition
Tricuspid insufficiency	
	<i>Other</i>
	Biliary ascites
	Bowel infarction
	Chylous ascites
	Familial Mediterranean fever
	Granulomatous or eosinophilic peritonitis
	Pancreatic ascites
	Vasculitis

SAAG = serum-ascites albumin gradient.

While were waiting for further tests

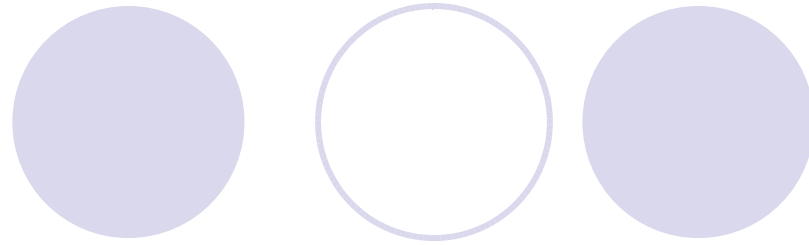
- Hepatic failure
 - acute vs chronic
 - normal size liver, no history of liver disease.
- Polycythemia – HB – 16.6, WBC – 13.
- B12 > 2000
- History of DVT
- **Suspected bud chiary d/t polycythemia vera**

Additional tests



- No history of drinking
- Serology of hepatitis b + c negative
- ANA (-), anti smooth muscle (+), AMA (-).
- Alpha feto protein (-)

Imaging studies



US

- Portal vein – slow flow.
- IVC – open but narrow
- Hepatic artery – open, Hepatic veins – could not be demonstrated
- Spleen – mildly enlarged (13.3 cm)
- Large amount ascites

CTA

- Confirms hepatic vein thrombosis – bud chiary !



MANAGEMENT

- The patient deteriorates rapidly
 - Develops encephalopathy
 - Worsening of liver and coagulation dysfunction.
- Encephalopathy – NG tube, lactulose, and finally me
- Phlebotomy
- Anticoagulation (dilemma)
- Thrombectomy failed
- after an overall 5 days admission – the patient dies.

SUBTRACTION

Se:1
Im:1 (F7/14)
Primary Angle: -1.09
Secondary Angle: 1.13

M.AVRAHAM
Study Date:16/09/2009
Study Time:14:11:28
MRN:

KVP: 80.0

UNKNOWN

C563
V6883



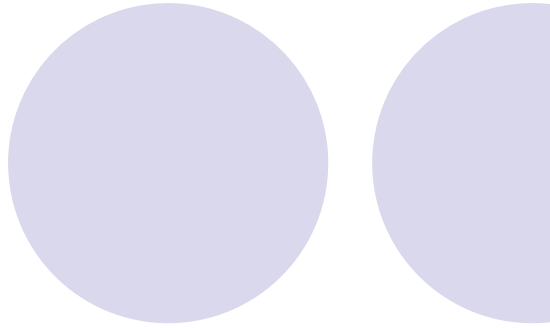


In conclusion

- The patient presented -
-
- Budd chiary syndrome presenting as fulminate acute hepatic failure.
-
- m/p d/t underlying polycythemia vera



Budd-Chiari Syndrome





Definition

- Any pathophysiologic process that results in interruption or diminution of the normal flow of blood out of the liver
- As commonly used, implies thrombosis of the hepatic veins and/or the intrahepatic or suprahepatic inferior vena cava.
- Distinguished from two other conditions that interfere with hepatic venous flow: sinusoidal obstruction syndrome (hepatic veno-occlusive disease) and right heart failure.

ETIOLOGY

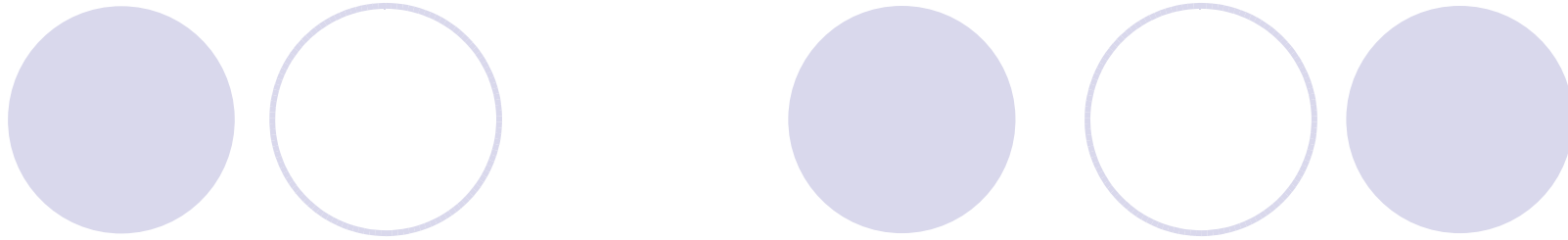


- An underlying disorder identified in over 80%.
-
- **Myeloproliferative disorders** — 50%
- **Malignancy** — 10%
- **Infections and benign lesions of the liver** —10% .
- **Oral contraceptives and pregnancy** — 20%.
- **Other hypercoagulable states** - factor V (Leiden) , factor II, APLA , Antithrombin deficiency , Protein C deficiency, Protein S deficiency, PNH
- **Behcet's disease** — Vasculitis
- **Membranous webs**
- **Miscellaneous** — SLE, MCTD, Sjögren's syndrome, IBD, hypereosinophilic syndrome, idiopathic granulomatous venulitis, sarcoidosis, protein-losing enteropathy, minimal change nephrotic syndrome, neurofibromatosis, alpha-1 antitrypsin deficiency, trauma (including laparoscopic cholecystectomy) or torsion of the liver, and rare familial cases
- **Idiopathic** — up to 20%

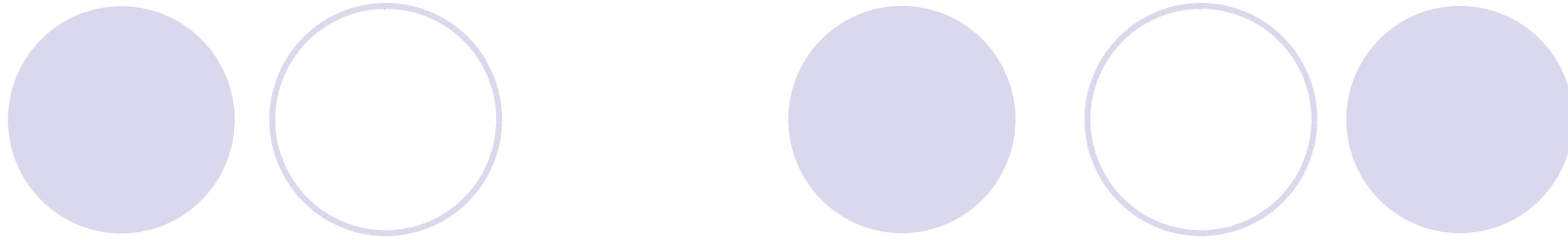
Medical therapy



- Require symptomatic therapy for fluid retention, which includes
- Stockings or repeated large-volume paracentesis, but
- Improve nutritional status.
- Therapy for the underlying condition (Patients with a history of anticoagulants such as warfarin)



- Anticoagulation — alone is unlikely to lead to sufficient
- The use of anticoagulation and diuretics alone as the primary treatment for acute or subacute Budd-Chiari syndrome with well compensated

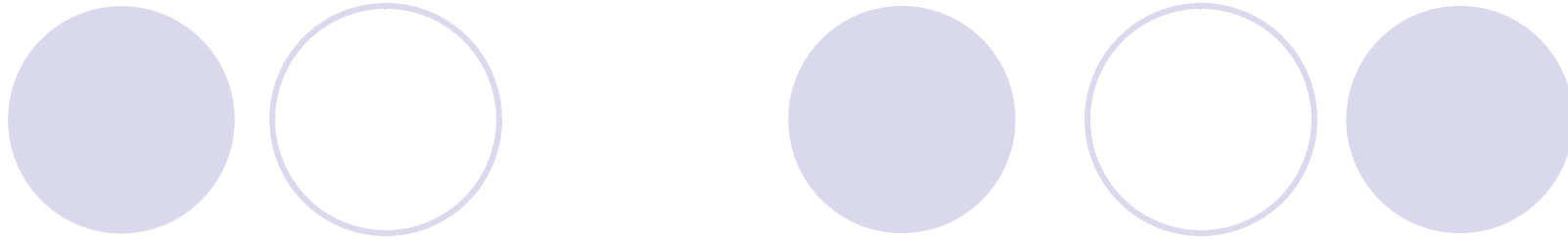


- Thrombolytic therapy — Systemic and locally
- Thrombolytic agents should not be used in the
- Thrombolytic treatment can be considered in p
three to four weeks.

Radiologic treatment



- These include angioplasty alone or with placebo



- Angioplasty — The anatomy may reveal a focal abnormality tl
- Stenting — The major problem is that reocclusion of the affec
- Transjugular intrahepatic portosystemic shunt — The rational temporizing measure prior to liver transplantation in patient

Surgical therapy



- Most surgical shunts drain the portal or mesenteric v artery to have a low pressure route by which to drain
- Surgical thrombectomy alone is usually not technical
- Surgical decompression is unlikely to be beneficial in

Liver transplantation



- May be the only option for patients with Budd-decompensated cirrhosis.

THANK YOU

