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MULTIMEDIA ARTICLE - Slide show

Sinistral Portal Hypertension. A Case Report

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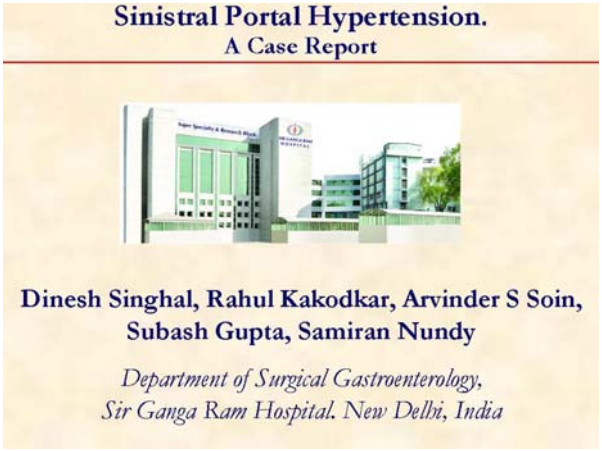
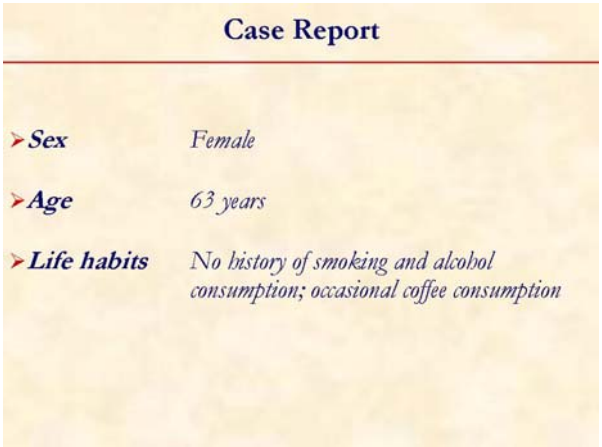
Summary

Sinistral portal hypertension is a clinical syndrome of gastric variceal hemorrhage in the setting of splenic vein thrombosis due to a primary pancreatic pathology. The distinguishing features from other forms of portal hypertension are preserved liver function and a patent extrahepatic portal vein.
The important causes include acute and chronic pancreatitis, pancreatic pseudocysts and pancreatic carcinomas. Benign pancreatic neoplasms only rarely cause sinistral portal hypertension. Splenic vein thrombosis complicates 7-20% of patients having pancreatitis or a pancreatic pseudocyst; however, bleeding occurs in only approximately 5% of patients.
The diagnosis of sinistral portal hypertension is achieved by a combination of gastroscopy, liver function tests, ultrasound examination (with Doppler) and/or contrast-enhanced CT scan of the abdomen.
A mere demonstration of sinistral portal hypertension does not warrant intervention. An expectant management is justifiable in asymptomatic patients with pancreatitis. However, concomitant splenectomy may be considered in patients undergoing operative treatment of symptomatic chronic pancreatitis if sinistral portal hypertension and gastro-esophageal varices are present.
In patients presenting with gastric variceal hemorrhage, splenectomy (with treatment for the primary pancreatic pathology, e.g. distal pancreatectomy) is curative with excellent long term results.





History (1): March 1997

- **Symptoms** Hematemesis (massive)
- **Blood** [a] Hb 7.8 g/dL, albumin 2.8 g/dL, total bilirubin 1 mg/dL, AST 32 IU/L, ALT 28 IU/L, ALP 78 IU/L, GGT 16 IU/L, INR 0.9
- **Endoscopy** Bleeding from gastric fundal varices [1]
- **US** Liver spleen normal
  Patent portal vein (16 mm) and splenic vein (10 mm)
  Dilated collaterals at splenic hilum
  No ascites
- **Presumptive diagnosis** Chronic liver disease with portal hypertension
- **Management** N-butyl-cyanoacrylate glue intravariceal injection (10 units of blood transfusion (each 350 mL) during the admission)

[a] **Reference ranges** Hb: 12-15 g/dL; albumin: 3.5-5 g/dL; total bilirubin: 0.2-1 mg/dL; AST: 0-42 IU/L; ALT: 0-60 IU/L; ALP: 39-117 IU/L; GGT: 0-64 IU/L; INR: 0.8-1.1

[1] Sarin SK, et al. Hepatology 1992; 16:1343-9.

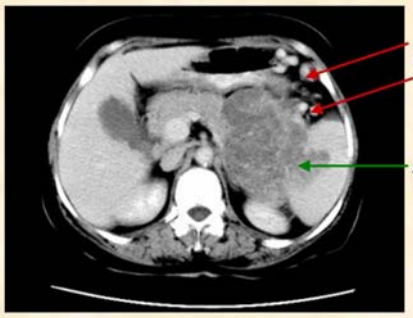
GI Surgery Review

- **Examination** No encephalopathy
  Abdomen: liver spleen not palpable
  No lump palpable, no ascites
- **Investigations** Albumin 3 g/dL, total bilirubin 1 mg/dL, AST 41 IU/L, ALT 38 IU/L, ALP 68 IU/L, GGT 17 IU/L, INR 1
- **Diagnosis** Portal hypertension with fundal variceal bleed
  Cause ?
- **Advised** Contrast-enhanced CT scan of the abdomen

History (2): February 2001

- **Symptoms** Melena
- **Blood** Hb 7.4 g/dL, albumin 4 g/dL, total bilirubin 0.4 mg/dL, AST 28 IU/L, ALT 33 IU/L, ALP 78 IU/L, GGT 14 IU/L, INR 1
- **Endoscopy** Portal hypertensive gastropathy
  No variceal bleed
  Bleeding of a duodenal ulcer
- **Management** Endoscopic ulcer therapy was performed with 3% sodium tetradecyl sulfate.

Contrast-Enhanced CT Scan



History (3): August 2001

- **Symptoms** Massive hematemesis
- **Blood** Hb 6 g/dL, albumin 3 g/dL, total bilirubin 1 mg/dL, AST 40 IU/L, ALT 34 IU/L, ALP 72 IU/L, GGT 19 IU/L, INR 1
- **Endoscopy** Bleeding gastric varices
- **Diagnosis** Chronic liver disease with portal hypertension and recurrent gastric variceal bleed
- **Management** N-butyl-cyanoacrylate glue injection (4 units of blood transfusion (each 350 mL) during the admission) and balloon tamponade
- **Follow-up** Referred to our centre for a surgical opinion due to persistent bleeding

Surgical Opinion

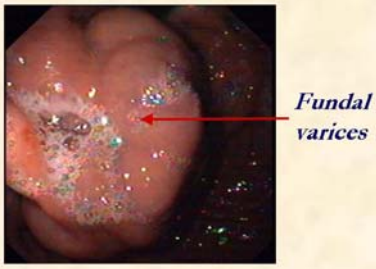
- **Further investigation**

  Serum amylase 42 U/L (reference range: 5-100 U/L)
  CEA: 1.8 ng/mL (reference range: 0-2.5 ng/mL)
  CA 19-9: 24 U/mL (reference range: 0-33 U/mL)

- **Pre-operative diagnosis**

  Pancreatic body tumor with a gastric variceal bleed due to sinistral portal hypertension (SPH) requiring surgical exploration

Endoscopic Examination: August 2001

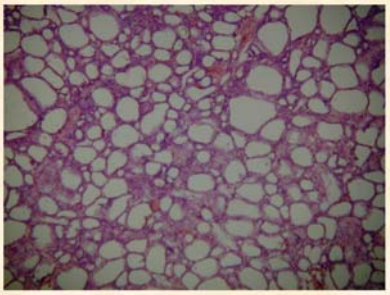


Operative Findings and Surgical Management

- Liver normal, no ascites
- Dilated collaterals along the greater curvature of the stomach
- Splenic vein thrombosis
- 6x6 cm tumor of the body and tail of pancreas adherent to the splenic hilum and gastric fundus
- Distal pancreatectomy plus splenectomy

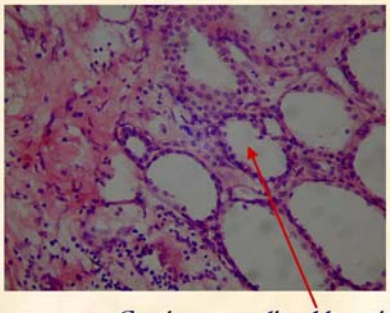
Pancreatectomy Specimen: Histopathology

Low power view



Pancreatectomy Specimen: Histopathology

High power view



Cystic spaces lined by mitotic cells

Diagnosis and Follow-up

- ***Post-operative diagnosis***

  Sinistral portal hypertension secondary to serous cystadenoma of the pancreas

- ***Follow-up***

  Uncomplicated recovery

  Discharged on the 7th post-operative day

  No evident disease 5 years after surgery

Discussion
Sinistral Portal Hypertension

Definition	*Hypertension confined to the gastrosplenic side of the portal venous system*
Synonyms	*Segmental / Left-sided portal hypertension*
Hemorrhage	*Rare, less than 1%*
Syndrome	*Gastric (fundal) varices* *Normal liver function* *Patent portal vein*
Genesis	*Splenic vein occlusion. In chronic pancreatitis, this is due to the extension of the peripancreatic fibrosis*

[2] Evans GR, et al. Am Surg 1990; 56:758-63.
[3] Loftus JP, et al. Ann Surg 1993; 217:35-40.
[4] Heider TR, et al. Ann Surg 2004; 239:876-82.
[5] Sakorafas GH, et al. Am J Surg 2000; 179:129-33.
[6] Iwasaki T, et al. Surg Today 1996; 26:442-5.
[7] Takase M, et al. Arch Pathol Lab Med 1997; 121:612-4.

Etiology
Sinistral Portal Hypertension

- *Pancreatitis (most common)*
- *Pancreatic neoplasm (very rarely cystic neoplasm)*
- *Iatrogenic (splenectomy, umbilical catheterisation, gastrectomy)*
- *Retroperitoneal fibrosis*
- *Hodgkin's disease*
- *Pancreatic transplantation*
- *Idiopathic*

[2] Evans GR, et al. Am Surg 1990; 56:758-63.
[3] Loftus JP, et al. Ann Surg 1993; 217:35-40.
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[8] Little AG, Moossa AR. Am J Surg 1981; 141:153-8.
[9] Smith TA, Brand EJ. J Clin Gastroenterol 2001; 32:444-7.
[10] Kakizaki S, et al. Hepatogastroenterology 2005; 52:1274-7.

Management
Sinistral Portal Hypertension

A mere demonstration of SPH does not warrant intervention !

Expectant	*Asymptomatic patients with pancreatitis + SPH without gastric variceal hemorrhage*
Surgical indication	*SPH with gastric variceal hemorrhage* *Symptomatic chronic pancreatitis + SPH without gastric variceal hemorrhage*
Procedure	*Splenectomy + treatment of the primary pathology (e.g. distal pancreatectomy)*

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Conclusion

- *In a patient with gastric variceal hemorrhage, if the liver function is normal and the extrahepatic portal vein is patent, the possibility of sinistral portal hypertension should be considered*

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Keywords Cystadenoma, Serous; Esophageal and Gastric Varices; Splenectomy; Splenic Vein; Thrombosis

Abbreviations SPH: sinistral portal hypertension

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