Original article

Splenomegaly and left sided portal hypertension

D.Voros,[1] E. Mallas,[1] A. Antoniou[2] E. Kafantari,[1] S. I. Kokoris,[3] B.Smyrniotis,[1] G. A. Pangalis[3]

SUMMARY

Background: Left-sided portal hypertension (L.S.P.H.) is usually associated with splenic vein occlusion or arteriovenous fistulae of splenic vessels ; however there have been some case reports of L.S.P.H. in patients with splenomegaly, without vein occlusion or fistulae.

In the present study we include a number of patients with haematological diseases and an enlarged spleen without thrombosis in order to study the circulation of the splenic area.

Patients and methods: During a two and a half year period 53 patients with haematological diseases and splenic enlargement were investigated by esophagogastroscopy and ultrasonography (US) of the splenic area focused on the perisplenic circulation.

Results: Thirty four out of 53 patients of the study, underwent both endoscopy and US investigations, 9 underwent endoscopy only and 10 underwent only US examination. Fourteen out of 34 patients (i.e. 43%) had endoscopic findings of varices or congestive gastropathy and 15 out of 34 (i.e. 44%) had abnormal circulation around the spleen in the ultrasound examination. Among the 19 patients who underwent either endoscopy or ultrasonography only there were 8 patients with positive findings (4 in the endoscopy group and 4 in the US group).

A possible explanation may be that the spleen receives through its enlarged splenic artery an increased volume of blood, which leads to an enlargement of the splenic vein. The vein remains open but cannot accommodate this increased blood volume; this causes impaired venous drainage and finally the blood drains through the short gastric veins or retroperitoneal collaterals.

Conclusions: We concluded from our study that patients with splenomegaly due to haematological disorders carry a high risk to develop left sided portal hypertension.

Key words: Left Sided Portal Hypertension, Portal Hypertension, Sinistral Portal Hypertension, Splenomegaly, Variceal bleeding, Esophagogastric varices

[1]2nd Dept. of Surgery, [2]1st Dept. of Radiology, 1st Dept. of Internal Medicine (Section of Haematology), [3]Medical School, University of Athens, Greece

Author for correspondence:

D. Voros, 110 Vas. Sophias Ave., 115 27 Athens, Greece,
Tel.: 30 210 7706152, Fax: 30 210 7785 780,
e-mail: diovoros@med.uoa.gr

INTRODUCTION

Left sided –otherwise called sinistral or segmental portal hypertension– and bleeding esophagogastric varices are usually associated with splenic vein occlusion or arteriovenous fistulae of the splenic vessels [1-4]. This condition attracted more attention in recent years because it is easily diagnosed, when suspected, and it can be cured by splenectomy. According to observations made by our group in the past, two out of six patients with left-sided portal hypertension, who finally underwent splenectomy for bleeding gastroesophageal varices, had massive splenomegaly due to an underlying haematological disorder without any signs of obstructive process of the splenic vein or an arteriovenous fistula[5].

Based on the aforementioned observations we decided to study the association of left-sided portal hypertension with splenomegaly in a large series of patients with haematologic disorders without evidence of liver disease or other conditions resulting in systemic portal hypertention, in order to investistigate further these observations and to find if there is more concrete evidence of left- sided portal hypertension developing as a result of splenomegaly.

PATIENTS AND METHODS

During a two and a half year period, we studied 53

patients with a variety of haematological diseases and splenic enlargement. We used a simple inclusion criterion: splenomegaly due to hematological disorders detected by imaging techniques. The exclusion criteria included: the history or the diagnosis of splenic vein obstruction and the clinical or laboratory evidence of cirrhosis. All patients with abnormal values of plasma proteins, raised transaminases, raised bilirubin, raised alkaline phospatase, γGt, prolonged prothrombin time, or liver pathology in US or CT investigations of the liver were excluded.

All patients were informed of the study design and they accepted to participate. In each patient the following laboratory tests were performed: full blood counts, liver chemistry tests, upper abdominal ultrasound and computed tomography studies. The presence of esophagogastric varices was investigated by using upper gastrointestinal endoscopy and ultrasonography of the spleno-portal vascular axis (by either one or both methods).

Ultrasound examination was performed using a commercially available unit (GE upgraded 450). All patients were examined after a 12hour fast, in supine and lateral oblique positions. The real time examination included size and texture of left and right liver lobes and three dimensions of caudate lobe. The portal vein diameter (normal values 11±2mm) was measured on the sagittal view during a deep inspiration with the patient in supine position. The splenic vein measurement was performed in transverse view at the level of the superior mesenteric artery, the patient in supine position (normal 6±1mm). The spleen was measured in right lateral oblique position and maximum anteroposterior and anterolateral diameters obtained to calculate Splenic Index. The Splenic Index (SI) is calculated by measuring (a) the longest dimension of the spleen and (b) the shortest dimension perpendicular at the hilum of the spleen on the intercostals scan through the greatest area. The SI is equal to the aXb and should be between 13-38 cm^2. The Color Doppler application was used for the presence and direction of flow (hepatopetal or hepatofugal). Velocities, volumes and flow were calculated using the software of the unit as well as resistive and pulsatility indices of portal vein, splenic vein and hepatic artery. All measurements were repeated three times and afterwards mean values were calculated.[6-9] Real Time and Doppler findings of liver lesions, lymph nodes, ascites, pleural effusion and portosystemic collaterals were noted if present.

Angiography was not considered in any patient because of its invasive nature. Liver biopsies were not performed for the same reason.

Table 1: Patient characteristics, Haematological Disease and Size of Spleen

Age (yr)	66.5 (range: 32-83)	
Sex	Men	26
	Women	27
Underlying hematological disease:		
Lympoproliferative Disorders		*27*
Chronic lymphocytic leukaemia		10
Hairy cell leukaemia		4
Non-Hodgkins lymphomas		13
Myeloproliferative Disorders		*22*
Chronic myelogenous leukaemia		4
Myeloschlerosis with myeloid metaplasia		9
Polycythemia vera		3
Essential Thrombocytemia		3
Chronic myelomonocytic leukaemia		3
Others		*4*
Mean length of spleen (cm)	20.1 (range: 17.5- 26.5)	

RESULTS

Our patient characteristics, such as age, sex, heamatological desease and mean splenic axis, are listed in table 1.

The positive findings of each type of investigation are shown on table 2. Thirty- four out of 53 patients studied underwent both esophagogastroscopy and ultrasound investigations, 9 patients underwent only upper gastrointestinal endoscopy and 10 only US examination. Fourteen out of the 34 patients (i.e. 43%) had varices of first or second degree in the gastric fundus or in the lower esophagus[1] or they had congestive gastropathy of different severity (Fig. 1a, b), and these were regarded as positive endoscopy findings. Fifteen out of the 34 patients (i.e. 44%) had abnormal collateral circulation around the spleen and this was defined as a positive finding (Fig. 2). Six patients (17.5%) had abnormal findings in both examinations. Among the 19 patients who underwent either endoscopy or ultrasound only, there were 8 patients with positive findings, 4 in the endoscopy group and 4 in the US group, (in the endoscopy group, splenic vein

[1] *First degree varices: small that flatten with air insufflation. Second degree: larger that not flatten with air insufflation.*

thrombosis had been excluded by previous CT).

The results of the ultrasound examination in 19 patients with positive findings are summarized on table 3. It is very interesting, besides the presence of collaterals, that the flow direction in the portal and splenic vein was hepatopetal (towards the liver). This finding is not usually associated with liver disease and systemic portal hypertension.

Among the 53 patients studied, five (9.5%) had a history of upper G.I. bleeding treated conservatively, while one of them had duodenal ulcer in the recent gastroscopy.

Five of the 53 patients underwent splenectomy (haematological indication) during the study period. In 3 of them the intraoperative examination revealed large collateral veins around the spleen (Fig. 3) although the rest of the portal net and the liver appeared normal. The liver of these patients was also normal. One of these patients who underwent splenectomy had varices diagnosed during the gastroscopy before the splenectomy. A new endoscopy performed four months after splenectomy revealed that the varices had disappeared.

DISCUSSION

This prospective clinical study revealed that a large number of patients with splenomegaly, without splenic vein thrombosis, have collateral venous circulation around the spleen verified by ultrasound examination or they have an endoscopic picture of varices or congestive gastropathy on gastroscopy, suggesting left-sided portal hypertension. The current knowledge of the pathophysiology [1,3,4,7] may explain why splenomegaly without splenic vein thrombosis can lead to left-sided portal hypertension. The spleen recieves through its enlarged splenic artery an increased volume of blood that leads in the beginning to an enlarge-

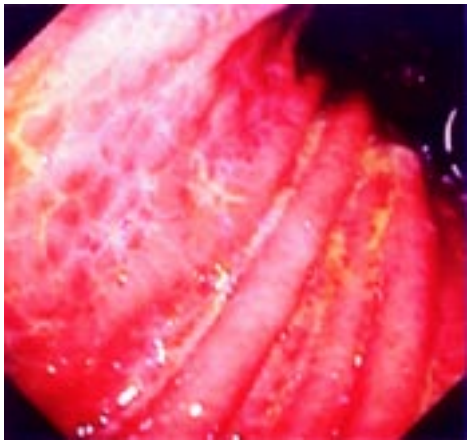

Fig. 1a.Typical portal gastropathy, "snakeskin" appearance.

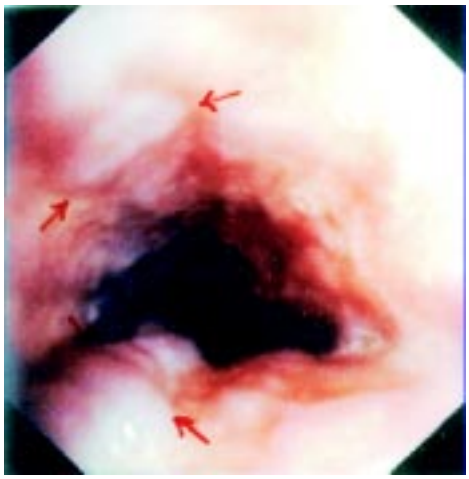

Fig. 1b.Oesophageal varices first to second degree (arrows).

Table 2: Findings in 53 Patients with Hematological Diseases and Splenomegaly

Investigation	*Positive finding during:*		*Both investigation*
	Endoscopy	U/S	
Endoscopy			
+	14 (43%)	15 (44%)	6 (17.5%)
U/S			
n=34			
Endoscopy only			
n=9	4 (44%)	–	
U/S only			
n=10	–	4 (40%)	
Total n=53	18 (34%)	19 (36%)	

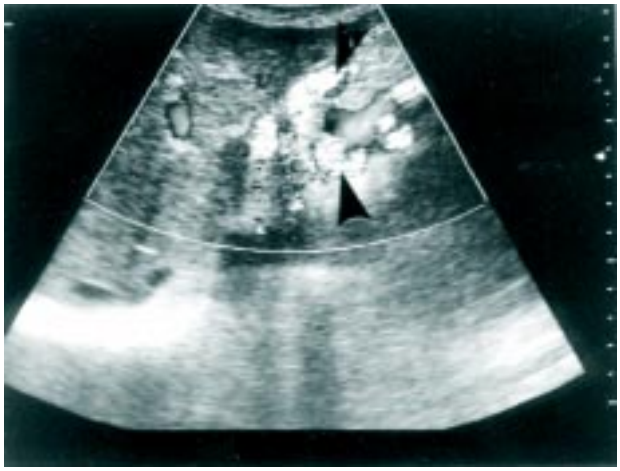

Fig. 2. Perisplenic collaterals (arrows). Multiple serpigineous venous collaterals at the splenorenal space.

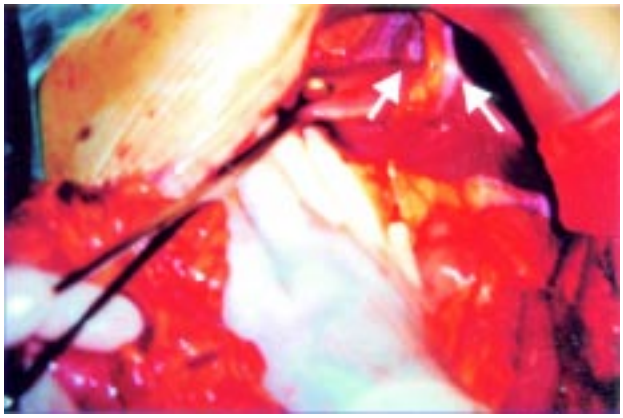

Fig. 3. Big collateral veins (arrows) seen around the upper pole of the spleen. The rest portal venous system had normal appearance.

ment of the splenic vein. The vein remains open but cannot accommodate this increased volume of blood and therefore there is impaired venous drainage. The final result is that the blood drains through the short gastric veins. Similar findings were presented by Aufses[10]. Wanless et al[11] suggested a differnt mechansm. There hypothesis was focused on the nodular regenerative hyperplasia of the liver in hematological disorders as a cause of portal venopathy. We did not perform liver biopsies to our patients considering the liver biopsy as an invasive investigation unsuitable for our research protocol. On the other hand it is interesting that in five of our study patients who underwent splenectomy for their haematological problem, no liver pathology was found. It is also mentioned in our results that no hepatofugal circulation was detected by the Triplex ultrasonography.

We must stress the low frequency of upper G.I. bleeding in these patients in contrast to those with splenic vein thrombosis.

Left-sided portal hypertension in cases of splenomegaly due to heamatological diseases has not been investigated extensively. There are a few case reports in the literature of splenic vein thrombosis in lymphoma[12].

Aufses has reported[10] a few patients with haematologic diseases (myeloid metaplasia, polycythaemia vera) and enlarged spleen, who developed left-sided portal hypertension without splenic vein thrombosis and were operated for variceal bleeding.

The left-sided portal hypertension as a result of splenic vein obstruction (thrombosis or from outside pressure) was presented at first by Greenwald and Wash in 1939,[13] who suggested splenectomy as the treatment of choice. The pathophysiology of the collateral venous circulation around the spleen and the development of esophagogastic varices with the possibility to bleed were described later in details by others.[1,3,14] Evans et all.[14] believe that splenic vein thrombosis produces an outflow block and prevents the outflow of blood from the spleen.

Table 3 Ultrasound parameters in 19 patients with abnormal findings.

	No of patients	mean values	Normal values
Length of spleen	19/19	19.8cm	11±2 cm
Splenic Index	18/19	272.6cm^2	13-38cm^2
Collaterals	19/19		–
Splenic vein diam	18/19	10.7mm	6±1 mm
Portal vein diam	13/19	17.1mm	11±2 mm
Portal vein flow direction	19/19	hepatopetal	hepatopetal
Portal vein blood flow	17/19	2145 ml/min	1200±572 ml/min

Splenic blood, unable to drain through the occluded splenic vein, flows into the vasa brevia and other collaterals. Gastric or esophageal varices may then develop from the increased left-sided splanchnic pressures.

The risk of bleeding from esophagogastric varicies in cases of splenic vein thrombosis has been estimated up to almost 50% [2,15]. Prophylactic splenectomy is not recommended except for certain cases presenting with chronic pancreatitis and known splenic vein thrombosis[3,4]. The problem of left-sided portal hypertension as a result of splenic arterio-venous fistula has not received a lot of attention, probably because of its rare appearance [16,17]. The indications for splenectomy may be similar to those of splenic vein thrombosis

It may be interesting to extend the investigation of left-sided portal hypertension and splenomegaly in other groups of patients with splenomegaly such as patients with splenomegaly due to hereditary haemolytic disorders (thalassemia) and maybe to exclude the possibility of liver disease by a more certain method as percutaneous biopsy.

We concluded from our study that patients with splenomegaly due to haematologic disorders carry a high risk to develop left-sided portal hypertension.

This group of patients with heamatological diseases and splenomegaly should be examined for left-sided portal hypertension and, our opinion, should be considered for splenectomy, when bleeding varices are present and liver pathology has been excluded by all means, including biopsy.

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