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Venous thrombosis of the portal system. Etiology, diagnosis and treatment – analysis of 225 cases

Piotr Małkovski,[1] Bogdan Michałowicz,[1] Jacek Pawlak[1], Włodzimierz Otto,[1] Elżbieta Leowska,[2] Olgierd Rowiński,[3] Rafał Paluszkiewicz,[1] Paweł M. Paczkowski,[1] Krzystof Zieniewicz[1]

[1]Department of General Surgery and Liver Diseases [2]Department of Nuclear Medicine [3]Department of Radiology, Medical Academiy, 02-097 Warsaw, Banacha 1a, Poland.

The authors present the material of 225 patients, treated since 1975, with various forms of portal system venous thrombosis (PSTV), of various origin and etiology. The largest group (120 patients) were the young people suffering from portal hypertension due to pre-hepatic venous obstruction of uncertain etiology, lasting since childhood. The next group consisted of 75 patients with liver cirrchosis coexisting with PSTV. In other cases PSVT was diagnosed as coincident with: Budd-Chiari Syndrome (8 cases), liver tumors (9 cases), chronic pancreatitis (3 cases), and polycythemia (2 cases). In 3 cases PSVT developed postoperatively and in 5 was the result of oral contraceptives. The course of the disease depended on extensivity and dynamism of thrombosis, but consequently led to the development of portal hypertension. The most effective diagnostic procedures were: computed tomography (CT) and ultrasonography Doppler flowmetry (SG), detecting PSVT in 96 % and 95 % of cases, respectively. Bleeding esophagal varices required either sclerotherapy (152 cases) or surgical treatment-decompressive shunts (26 cases) or »non-shunts« procedures (20 cases). In the cases of recent thrombosis, without bleeding varices, thrombolytic therapy was effective in all 6 cases.

Key words: portal vein: thrombois

Introduction

Portal system venous thrombosis (PSVT) was believed to be a rare phenomenon. Not more than 1000 cases of this pathology have been reported in the literature since 1901.[1–6] Recently, thanks to the development of noninvasive diagnostic imaging procedures (Doppler ultrasound and dynamic CT-scan) the diagnosis of PSVT is being established much more frequently.[3,5,7–10]

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Liver cirrhosis is recognised as the most common condition coexisting with PSVT.[1,3,6] Decrease of the portal blood flow is probably the main factor enhancing the development of thrombosis.[4,6,9] The frequency of PSVT in cirrhotic patients is estimated by different authors at 5 to 25 per cent.[1,3,6,9] The clinical features of liver cirrhosis coexisting with PSVT (»double block« of portal flow) do not essentially differ from those of portal hypertension caused by cirrhosis alone. The dominating symptoms are: large esophageal varices with high bleeding tendency and progressive impairment of liver function.[1,6,9]

The other conditions predisposing to PSVT are liver tumours (often accompanying liver cirrhosis), carcinoma of the gallbladder, pancreas and stomach, inflammatory processes within the abdominal cavity (pancreatitis, peritonitis, Crohn disease).[3,9,11–13] In liver tumours thrombosis involves mainly the intrahepatic branches of the portal vein,[12,14] while in the other cases its' main trunk and splenic vein.[13] The less frequent causes of PSVT are trauma, including iatrogenic lesions at the time of surgery (most commonly splenectomy)[15,16] and hypercoagulation conditions (puerperium, polycythaemia, paroxysmal nocturnal hemoglobinuria and congenital antithrombin III deficiency).[16,17] More recently the coexistence of PSVT with Budd-Chiari syndrome was reported.[16,18] The increasing incidence of PSVT in young women using oral contraceptives is also noteworthy.[7,16,18,19]

In a considerable number of cases of PSVT the etiology remains unknown.[16,20] It concerns most of patients treated for portal hypertension due to pre-hepatic obstruction of portal flow, second, after cirrhosis, reason of gastroesophageal varices.[21,22] According to some authors the role of neonatal umbilical infection, resulting in umbilical and portal thrombosis is overestimated.[16,21] Retrospective investigations, only in some cases of pre-hepatic block, have proved it's relationship to umbilical infection or thrombosis.[21,22] Anyway, the etiology of pre-hepatic portal obstruction remains unclear.[16,21]

In the symptomatology of PSVT, apart from the symptoms of primary disease (if present) the clinical features of portal hypertension dominate. These include extensive collateral venous circulation, gastroesophageal varices, and splenomegaly accompanied sometimes by abdominal pain, ascites and jaundice. Their intensity depends upon the degree of hemodynamic disorders of the portal venous system.[1,6,9,13,16,20]

Material and methods

From 1975 to 1993 the diagnosis of PVST was established in 225 patients treated in the Department of General Surgery & Liver Diseases of Warsaw Medical Academy. There were 100 females and 125 males aged from 17 to 74 (mean 38 years). In 197 cases the reason of admission was active or controlled bleeding from esophageal varices, accompanied in 35% of patients by symptoms of liver function impairment (jaundice, ascites, coagulopathy). Some of these patients had a long history of portal hypertension. Other indications for admission were: suspected liver tumour (9 cases), chronic pancreatitis (3 cases), upper abdominal pain (6 cases), ascites of unknown origin (6 cases), splenomegaly alone (3 cases) and ultrasonographic suggestion of portal vein obstruction (1 case).

All actively bleeding patients were treated by esophageal balloon tamponade and/or emergency sclerotherapy with simultaneous i. v. infusion of Vasopressin. As soon as the bleeding was controlled and the patients recovered, they were submitted (except the previously diagnosed patients) to diagnostic procedures including Doppler sonography (SG-94 pts), CT-scan (CT-62 pts), celiac arteriography with venous phase (61 pts) and splenoportography (SPG-4 pts).

Subsequently the patients were treated either surgically (26 decompressive shunting procedures and 20 non-shunt operations) or by repeated endoscopic sclerotherapy. Patients with portal and spenic vein thrombosis due to chronic pancreatitis were treated conservatively. In 5 patients with liver tumors a partial hepatectomy was performe (in 4 cases the tumour were inoperable). Two patients with polycythaemia were referred to Hematology Department for chemotherapy.

All patients with recent PSVT admitted during 1975–1980 (3 with coexisting hepatic vein thrombosis and 2 with portal trunk thrombosis alone) were treated conservatively. Further 8 patients, hospitalised after 1980, were submitted to thrombolytic treatment with Streptokinase (3 cases) and recently with recombinant tissue plasminogen activator (rt-PA, Actilyse).

The analysis of the above presented group of patients with PSVT included the results and reliability of diagnostic imaging procedures and results of treatment regarding the etiology of

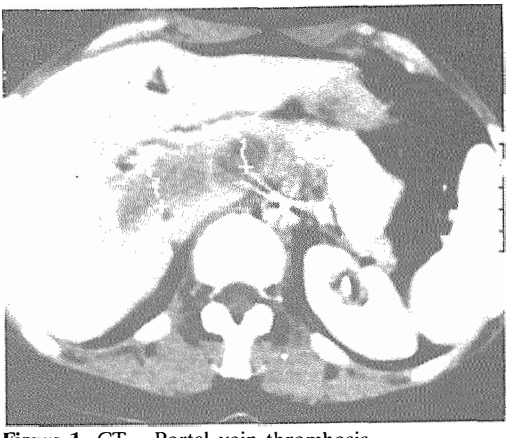

Figure 1. CT – Portal vein thrombosis.

PSVT or coexistence with predisposing diseases.

Results

SPG showed the presence of portal obstruction in each of 4 cases. The venous phase of arteriography was conclusive in only 38/61 cases (62%). Apart from the SPG, which is not performed now, the highest reliability was attributed to SG and CT (Figure 1), detecting PSVT in 90/94 patients (96%) and 59/62 cases (95%) respectively.

Results of various methods of treatment in particular groups of patients are presented in Table 1.

Discussion

The most numerous was the group of patients presenting clinical symptoms of portal hypertension due to pre-hepatic venous occlusion, in whom the diagnosis of PSVT was established in childhood and followed by long-lasting treatment including surgery and sclerotherapy. After having grown up they have been referred to us by pediatric centers for continuation of treatment. Most of them are young people in good general condition, often with partly recanalized portal system. They require, however, permanent medical attendance and (not infrequently) repeated sclerotherapy.[21,22] Regarding their past history, only few of them can be considered as candidates for surgical treatment. In our experience only in 5 cases we were able to perform a venous shunt; 2 patients with uncontrollable recurrent variceal bleeding required a non-shunt surgery (splenectomy with gastroesophageal devascularisation, esophageal stapling). Generally, the results of treatment in this group are satisfactory, fatal cases of uncontrollable hemorrhage bein rare.[21,22]

The coexistence of PSVT with liver cirrhosis ("double block") deteriorates the prognosis.[1,6,9,16] Gastroesophageal varices are usually very large and the frequency of recurrent massive bleedings, followed by hepatic insufficiency is remarkably higher compared to other cases

Table 1. Results of various methods of treatment in particular groups of patients.

Number of patients	Etiology or coexisting disease	Sex		Treatment					Hospital deaths
				Surgical		Non-surgical			
		F	M	Shunt	Non shunt	ES,S–B,V	Thrombolytic	Other	
75	Liver cirrhosis	23	52	21	8				16 (55%)
						46			9 (20%)
120	Unknown etiology	62	58	5	12				1 (5,8%)
	PSVT since childhood					103			4 (3,8%)
9	Liver tumors	3	6			5	2	4	2
8	Budd-Chiari	6	2					3	3 (100%)
	syndrome						3 SK, 3 rtPA		0
5	Oral contraceptives	5						2	2 (100%)
							3 rtPA		0
2	Polycythaemia		2					2	0
3	Intraoper. trauma	1	2			3			0
225	Total	100	125						

S-B = Balloon tamponade, V = Vasopressin infusion, SK = Streptokinase, rtPA = Actilyse, ES = Endocsopic sclerotherapy.

of portal hypertension.[1,9] It was proved that the percentage of rebleedings and postoperative deaths is three times higher compared with patients with portal hypertension caused by liver cirrhosis alone.[9] Intensive sclerotherapy improves the results of treatment of patients with "double block".[9]

In the group of patients with liver tumours there were 4 cases of hepatoma in the cirrhotic liver. Two patients required sclerotherapy. The resectability of the lesion depended on dimensions of the tumour itself, as well as the extent of intrahepatic portal thrombosis. The final intraoperative estimation has significantly improved since the intraoperative sonography is used. It enables the exact identification of the affected parts of the liver tissue and occluded branches of portal vein.[13,14]

Our patients with PSVT and chronic pancreatitis, as well as those observed by other authors, did not require any special treatment.[13] Splenomegaly observed in 2 cases and ascites in 1 case, were slowly decreasing during 2 years of follow-up, as the recanalization of occluded veins progressed.

Chemotherapy (Hydroxycarbamide) administered to patients with polycythaemia led to regression of thrombotic occlusions after 6 and 9 months with decrease of ascites. One patient with PSVT, caused by intraoperative lesion required sclerotherapy. In these cases thrombosis did not result in total portal occlusion and severe hemodynamic disorders, which explains a relatively mild course of the disease.

Much more dramatic course, despite the initially slight symptoms, was observed in patients PSVT, coexisting with trombosis of hepatic veins. All of 3 conservatively treated patients died of progressing liver failure and massive variceal bleeding. Better results were achieved by thrombolytic treatment – Streptokinase in 3 and rt-Pa in 2 cases.[7,18] Significant clinical improvement appeared in the first day of treatment with rt-PA and after 2–3 days of Streptokinase therapy. Although the full recanalization of the occluded veins was not recorded, the Doppler flowmetry allowed to document a significant increase of both portal and hepatic venous flow.

Thrombolytic therapy with rt-PA was also administered in 3 cases of isolated portal thrombosis caused by oral contraceptives. All these women had a short history (less than 1 month) of abdominal pain, splenomegaly and slight ascites.[7] Also in these cases the therapy was started at the moment of diagnosis and resulted in rapid clinical improvement. In no case an immediate full recanalization of the thrombosed veins was seen at SG, but progressing improvement in portal flow was recorded at repeated Doppler examinations. After two years all 3 patients are doing well, with proved repermeabilisation of the portal system and hepatopetal blood flow. It has to be stressed that in none of our patients with esophageal varices thrombolytic therapy caused variceal bleeding.

Conclusions

The etiology of PSVT is not uniform, in many cases it remains unknown. In cirrhotic patients the diagnosis of coexisting PSVT seriously deteriorates the prognosis. Clinical features of PSVT vary in dependence upon the extensiveness and dynamism of thrombotic process, but sooner or later portal hypertension develops with all its consequences.

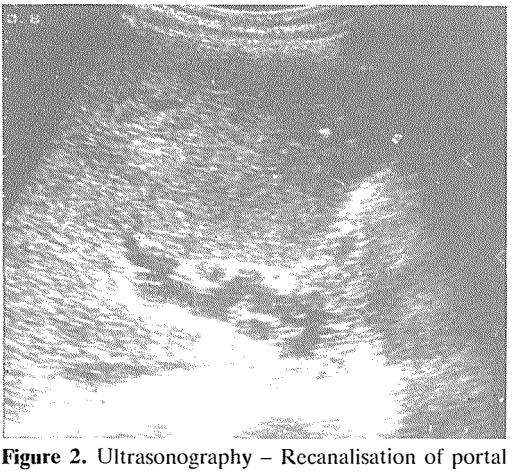

Figure 2. Ultrasonography – Recanalisation of portal vein.

The most effective diagnostic methods are SG and CT, with preference for SG (Figure 2) because of it availability and safety. Repeated SG allovs a systematic monitoring of the disease and treatment results. In the cases of recent thrombosis early diagnosis enables effective treatment.

In our opinion, every case of recent, progressing PSVT requires thrombolytic tgreatment, as the only means to achieve radical improvement, if not full recovery.

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