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SURGICAL EXPERIENCE WITH PORTAL HYPERTENSION DUE TO HEPATOPORTAL SCLEROSIS (NONCIRRHOTIC PORTAL FIBROSIS)

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ABSTRACT

Hepatoportal sclerosis or idiopathic portal hypertension has a worldwide distribution with prevalence in developing and temperate countries. Of 64 patients with this disease seen during a twelve year period, 36 underwent splenectomy or a shunt procedure. The indications for surgical intervention were severe hypersplenism and persistent left upper abdominal pain and/or history of frequent episodes of esophageal bleeding. There were two operative deaths from uncontrollable bleeding. Of 34 patients, none developed esophageal bleeding or encephalopathy on follow-ups ranging from one to 12 years. Splenectomy in patients without bleeding was effective. Proximal splenorenal shunt was also satisfactory in all cases. However, in the last few cases, Warren's shunt has produced even more encouraging results.

In addition to a general overview, features of this disease not previously discussed or emphasized in the literature, operative findings and surgical methods used, along with long-term results, are presented.

INTRODUCTION

Portal hypertension in the absence of cirrhosis or other apparent mechanical obstruction has been a perplexing syndrome since the time of Banti and others. Although this syndrome had been reported and discussed sporadically prior to 1965,[1,2] it became a clinically distinct entity after the classic report of Mikkelsen, et al.[3]

Since then, several investigators in india,[4,5,6] Europe[7] and Japan[8] have done much to further elucidate the clinicopathological features of this disease, and have clearly separated it from different forms of liver cirrhosis.[9] The cardinal features of this disease consist of moderate-to-huge splenomegaly with secondary hypersplenism and anemia with portal hypertension, usually presenting with esophageal varices and occasional well-tolerated bleeding episodes. The liverfunction tests are normal or only minimally abnormal. On gross inspection and microscopic examination of the liver, there should be no evidence of cirrhosis. This disease has been referred to in the literature by a variety of names such as hepatoportal sclerosis,[3,10] noncirrhotic portal fibrosis,[11] and idiopathic portal hypertension.[12,13] Evidently, by definition, other known causes of portal hypertension such as partasites, tumor, blood disease, gross thrombosis or obliteration of the extrahepatic portal system or obvious arteriovenous fistula must not be present.

PATIENTS AND METHODS

Between 1971 and 1984, 36 patients who fulfilled the criteria for idiopathic portal hypertension (IPH) underwent operation. In order to compare the clinical and laboratory features of this disease with that of cirrhosis and to evaluate the prevalence of IPH in our patient pouplation, a study was undertaken. For this purpose, the records of all cases of portal hypertension with and without cirrhosis admitted to the Tehran University Hospitals during the five year period from 1979 to 1983 were reviewed.

The criteria utilized for designating the diagnosis of IPH to patients were in accord with those described by Nayak, et al,[5] Anthony, et al,[9] and Okuda, et al.[14] However, we excluded all patients with positive HB_sAg from our series as well.

All patients had a detailed history and physical examination, with special attention to their past history and previous illnesses. A history of pica was especially sought. All patients underwent routine blood tests and liver function tests.

All patients has a barium meal esophagogram, and underwent endosopic esophagoscopy. Many patients also has a bone marrow aspiration examination. All patients underwent at lest one liver needle biopsy. In addition to routine hematoxylin and eosin, specimens were stained with trichrome for collagen and silver impregnation for reticulin fibers.

The indications for surgical intervention in these patients were severe secondary hypersplenism and persistent lefr upper quadrant pain and/or a history of frequent episodes of upper GI bleeding.

All patients underwent a preplanned intraoperative investigation. This included examination of the liver and spleen and performing a generous wedge biopsy from the right lobe of the liver. Portal pressure was measured routinely via a cannula passed through a jejunal vein. The splenic artery was ligated temporarily to determine the effect of its ligation on portal pressure. Portal pressure was again measured at the conclusion of the operation.

RESULTS

66 patients with IPH were evaluated in our series. 36 patients underwent surgery and thus had open liver biopsy and intraoperative portal pressure measurements. The remaining cases were selected based upon liver needle biopsies and clinical and laboratory findings. The frequency of IPH (as compared to cirrhosis) in our patient population was 12 percent.

The main symptoms in these patients were left upper quadrant pain, discomfort, easy fatiguability, inability to do heavy work, and hematemesis.

In the past histories of the patients, two findings deserve emphasis. Firstly, many patients gave a history of a "febrile" illness for some fime in their past, and some also had diarrhea. None had any idea as to the nature of this febrile illness. Secondly, of 26 patients questioned regarding a history of pica, 20 patients (76%) admitted to this habit as a child or in their early teen years. Some admitted to have continued doing so until their late teen years and one patient kept the habit even up to admission at age 25. No detailed information regarding the amount and frequency of ingesting clay or plaster could be obtained, as patients were usually unwilling or embarrassed to discuss the matter fully. 90% of the patients were from small towns or villages.

Table 1. Age and sex distribution of patients with IPH

Age group (yrs)	Male	Female
0-10	0	0
11-20	22	7
21-30	8	10
31-40	4	3
41-50	5	5
over 50	1	1
total	40	26

Table II. Clinical features

Clinical findings	percentage of patients
Splenomegaly	100%
left-sided abdominal pain	70%
esophageal varices on barium meal	53%
history of hematemesis	39%
palpable liver	29%
ascites	22%

On physical examination; none of the patients were apparently malnourished, but most were anemic and seemed weak. Six young patients (between 15 and 20 years old) showed evidence of delayed growth and retarded secondary sexual development. Not a single patient in the entire group had stigmata of cirrhosis or significant ascites. Subcutaneous abdominothoracic venous collaterals were absent to minimal.

There were 40 males and 26 females. The age distribution of the patients is presented in Table I. The prevalence of the salient clinical findings are shown in Table II.

Laboratory findings revealed anemia and pancytopenia, with little abnormality of liver function tests, except for marked and persistently abnormal prothrombin time. Hemoglobin values ranged from 7 g/dl to 13.5 g/dl (mean 9.6 g/dl). The WBC count was decreased in 50% of the cases, but never below 2500. The platelet count ranged between 60,000 to 300,000, with an average of 120,000. Bilirubin values were within normal range in 75% of the patients and in cases in which the value was elevated, it was never higher than 2.5 mg/100 ml. Serum electrophoresis revealed an increase in $alpha_2$ globulin (60%), $beta_1$ globulin (50%), and gammaglobulins (80%). There was a mild-to-moderate decrease of serum albumin in many patients, but this value never dropped below 2.8 g/100 ml. Among liver function tests, increase in the prothrombin time was the most consistent finding. It was 12 seconds (control: 12 seconds) in only three patients.

Table III. Types of operations performed in 36 patients

Type of Operation	Number of patients
splenectomy	19
proximalsplenorenalshunt	10
distalsplenorenal(Warren's)	3
mesocaval(H-graft)	3
portocaval	1

Table IV. Portal pressure in 36 patients

Portal pressure in cm normal saline	No. of patients
below 19	0
20-29	14
30-39	16
over40	6

All liver needle and open biopsies were consistent with the diagnosis of noncirrhotic portal fibrosis. Two specimens were kindly reviewed by Professor D. G. Ishak of the United States Armed Forces Institute of Pathology, Washington, D. C. Pathological features consisted of portal fibrosis of varying degrees in all biopsies. The fibrosis in some cases was concentric in portal areas and in others, fibrosis extended into the parenchyma but not connecting with the adjacent portal areas to form nodules. In many cases there was mild-to-moderate infiltration of mononuclear cells in the portal areas.

Surgical Procedure

36 patients underwent operation. Based upon intraoperative portal pressure readings and its decrease after temporary interruption of splenic artery flow and presence or absence of history of hematemesis, either a splenectomy alone or a shunt procedure was performed. Patients who did not have a history of upper GI bleeding or whose portal pressure decreased to below 20 cm normal saline after ligating the splenic artery underwent splenectomy alone, otherwise a shunt procedure was carried out. The types of operations performed are presented in Table III. Initial intraoperative portal pressure readings are presented in Table IV.

The average decrease in portal pressure after ligating the splenic artery was 10.7 cm normal saline, and the average decrease in portal pressure after shunt procedures was 16.3 cm normal saline.

Explanation of intraabdominal findings is noteworthy. The liver was always smooth and without nodularity, but firmer than usual with a tinge of gray-white color due to increased fibrosis. The spleen was large-to-huge with a firm consistency and a thickened capsule. In many patients, the spleen was markedly adherent to the diaphragm and adjacent viscera by firm adhesions (perisplenitis). Through these adhesions and in all cases, through the lienorenal and gastrosplenic ligaments, large, dilated and tortuous veins coursed toward the diaphragm and upper stomach. In many cases, the findings in the left upper abdomen suggested that an inflammatory process had been present in the spleen sometime in the past.

The splenic vein usually revealed focal areas of thickening and thinning. In recent cases, we have conducted routine biopsies of the splenic vein and pathology has confirmed mural and intimal thickening (phlebosclerosis or endophlebitis). The splenic artery was very large, dilated and tortuous in the majority of cases.

Splenectomy Group

In 16 cases, the intended splenectomy was performed. In three patients, the planned shunt operation was abandoned because of difficult splenectomy due to total adherence of the spleen to the diaphram and continuous oozing of blood which took considerable time and effort to control. One of these patients died of hematemesis three days later.

After discharge, all 18 patients who underwent splenectomy were well. Neither of them developed encephalopathy or recurrent bleeding. The signs and symptoms of anemia disappeared after operation, and the patients' symptoms were alleviated.

Shunt Group

17 patients underwent portasystemic shunts (Table III). In 10 patients a proximal splenorenal shunt was performed, and in three patients, a distal splenorenal shunt (Warren's shunt) was carried out. The splenic artery was ligated in the latter cases. In these patients, the size of the spleen decreased and hematological abnormalities improved.

In four patients, other shunts were performed due to unavailability of suitable splenic vein. In three patients, an H-graft mesocaval shunt was performed, and the mesenteric vein was not ideal in any of these cases. Except for one operative death, all patients did very well postoperatively with improvement of anemia and subsidence of LUQ pain. One patient in the H-graft group developed hematemesis postoperatively due to graft thrombosis. Reoperation in this patient was followed by rebleeding and the patient died in the hospital. It is noteworthy that none of the patients developed encephalopathy, which is probably due to the relatively good hepatic function in this disease.

DISCUSSION

Although the etiology and pathogenesis of IPH remains unclear, much is known about its pathological stigmata.[3,4,8,12] From the pathological point of view, our patients are quite similar to those reported from the U.S.,[3] England,[7] and India.[10] Clinically however, they resemble more closely those reported from northern India.[6,11]

Although the pathological sequelae seem to be identical worldwide, the offending agents may be different. The basic pathological change is hepatoportal fibrosis and sclerosis. However, the morphology of the spleen in several of our patients did not seem to reflect simple passive congestion.

Based on their experience with injection-corrosion techniques, Boyer, et al[12] believe that the basic pathology lies in thrombotic obstruction of terminal intrahepatic branches of the portal vein. But at present, the clinicopathological criteria for designating this disease consist of portal hypertension and splenomegaly with hepatic portal fibrosis without cirrhosis or other known causes.[14] There seems to be as yet some uncertainty and overlap between IPH and the tropical splenomegaly syndrome.[15,16,17]

Regarding the etiology of this disease, different hypotheses have been proposed but no single view has yet been proved or unanimously accepted. Infection, probably as a non-specific offending agent, has been implicated. Some workers have proposed heavy metal or inorganic substances such as arsenicals as a cause. Thomas, et al[19] have found vinyl chloride poisoning as a cause of idiopathic portal hypertension. Considering the worldwide distribution of this disease and prevalence in temperate developing countries and its decline in Europe, the US and Japan, it is quite likely that an environmental offensive agent or agents initiate this disease. The offending agent may be different in various parts of the world. The high percentage of pica in our patient group may have some relation to this disease. The anemia in these patients was usually severe, with depleted bone marrow iron stores. The stools of these patients, tested several times for occult blood in the absence of hematemesis and melena, were always negative.

With regard to the appropriate treatment and natural history of this disease, much less is written in the literature. We examined three patients after eight, nine and 12 years respectively. Their physical conditions were good and there was no change in their liver function tests and repeat needle liver biopsy again showed periportal fibrosis with no evidence of cirrhosis. Many patients examined one to six years postoperatively were in good health and had no complaints. It is interesting that none of our operated patients developed postoperative esophageal bleeding or encephalopathy since our first operation on these patients over 13 years ago. Zeegan, et al[7] obtained similar results. We do not have follow-ups on non-operated patients. Splenectomy has been an effective procedure in those patients with no history of frequent and severe hematemesis and long-term follow-ups have validated this conclusion. However, a shunt procedure is generally preferred. Warren's shunt seems to be a particularly effective procedure. No postoperative pain or fever developed in these patients, the spleen decreased in size markedly and the hematologic profile improved subsequently.

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