Bleeding Oesopageal Varices in the Absence of Intrahepatic or Extrahepatic Obstruction of the Portal System and without Portal Hypertension

By GEORGE W. JOHNSTON, M.B., F.R.C.S.
Department of Surgery, Queen's University, Belfast

OESOPHAGEAL VARICES are generally secondary to portal hypertension and are therefore usually associated with obstruction to the portal circulation within the liver as in cirrhosis or in the portal vein itself. Thus, according to the site of obstruction, portal hypertension is usually classified as intrahepatic or extrahepatic in type (Whipple, 1945). However, an increasing number of cases of portal hypertension with extensive collateral circulation in the absence of any demonstrable organic obstruction, are being reported (Osler 1900; Rousselot 1940; Pemberton and Kiernon 1945; Whipple 1945; Hallenbeck and Shocket 1957; Tisdale et al 1959; Leather 1961; Imanaga et al 1952; Polish et al 1962; Siderys and Vellios 1964; Turnberg 1964). Varices have also been noted in patients with liver disease but without demonstrable elevation of portal pressure (Morton et al 1954; Homer et al 1964). There have been few reports, however, of varices in the absence of either liver disease or portal hypertension. The following case is an example of this unusual and interesting condition.

CASE REPORT

E.T., a female aged 58, first presented at hospital in April 1963, with a history of three episodes of vomiting clots of blood during the previous three months. All investigations at that time including barium studies, liver function tests, and occult bloods were negative.

She was next seen in September 1964 complaining of vomiting clots of blood about once per month for the previous 1½ years. There was no history of indigestion, heartburn, jaundice or alcoholism. Repeat barium studies were negative but oesophagoscopy revealed the presence of varices and full investigation was initiated. Clinical examination revealed only obesity and mild anaemia. Liver and spleen were not palable and there were no signs of liver disease. Haemoglobin was 10.2 gm. per 100 ml. and the anaemia was of the microcytic hypochromic type. Bleeding time, clotting time and prothrombin time were all normal. Electrophoresis showed a marked reduction in albumin with a slight increase in alpha-2-globulin (total protein 5.9; albumin 2.84; alpha-2-globulin 0.86 gm. per 100 ml.).

Liver biopsy obtained using a Terry needle was normal. X-ray chest was normal apart from slight general cardiac enlargement. Splenic venogram showed a normal portal vein and normal intrahepatic pattern without evidence of abnormal collateral circulation (Fig. 1). The intrasplenic pulp pressure was 140 mm. of saline. Azygos venography revealed no obstruction of the azygos system (Fig. 2).

On repeat oesophagoscopy in November 1964 the presence of moderately large varices on the post oesophageal wall, extending from 30 to 34 cms., was confirmed. These were injected with "Varistab" by the method previously described (Johnston

and Rodgers, 1964). She has complained of vomiting a few clots on one occasion since discharge but occult bloods have remained negative.

Discussion

Palmer and Brick (1955) classified oesophageal varices as follows:

1. Those associated with portal hypertension for obvious intrahepatic or extrahepatic causes.
2. Those associated with portal hypertension but without any clinical or anatomical explanation for the hypertension.
3. Those associated with rise in the superior vena caval pressure.
4. True primary varices limited to uppermost part of the oesophagus.
5. Idiopathic varices without either portal or superior vena caval hypertension.

Of the 350 patients with oesophageal varices reviewed by Palmer and Brick (1955), 13 appeared to be of this last variety, but the portal pressure was measured in only 4 of these cases. Previous reports of idiopathic varices were based on anatomical and biochemical findings without measurement of portal pressure and thus can not be considered proven (Nochimowski 1932; Friedman 1934; Shafer and Kittle 1950; Garrett and Gall 1953; Mithoefer 1953). Schaefer and colleagues (1964) reported 2 patients with varices in the absence of portal hypertension or cirrhosis

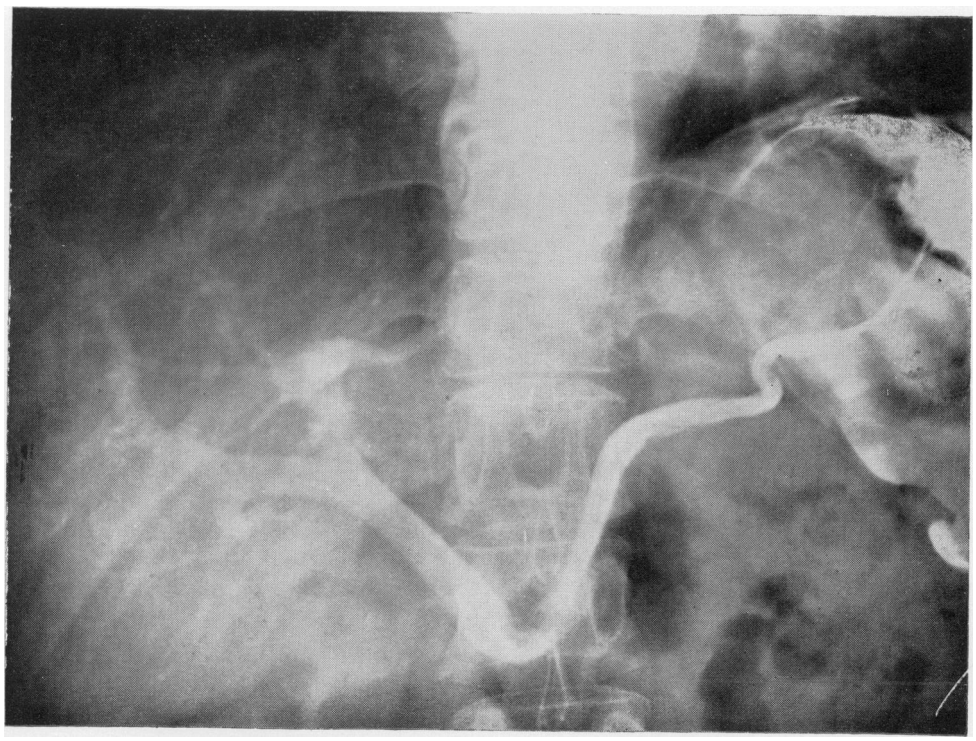

Fig. 1. Splenic venogram showing normal intrahepatic and extrahepatic portal vascular pattern.

of the liver. However, in both cases there was a history of alcoholism and the presence on biopsy of fatty metamorphosis of the liver. Rack and colleagues (1952) reported a similar case of bleeding oesophageal varices associated with slight fatty metamorphosis of the liver but without portal hypertension. In none of the cases previously reported was obstruction of the azygos system excluded. In the case reported there was no evidence of obstruction of the portal or azygos systems, the liver architecture was normal and there was no elevation of portal pressure.

Although she has had no catastrophic haemorrhages, the persistent blood loss has resulted in an iron deficiency anaemia. It is considered that injection of the varices with sclerosant, repeated if necessary, will control the condition without resort to more radical surgery.

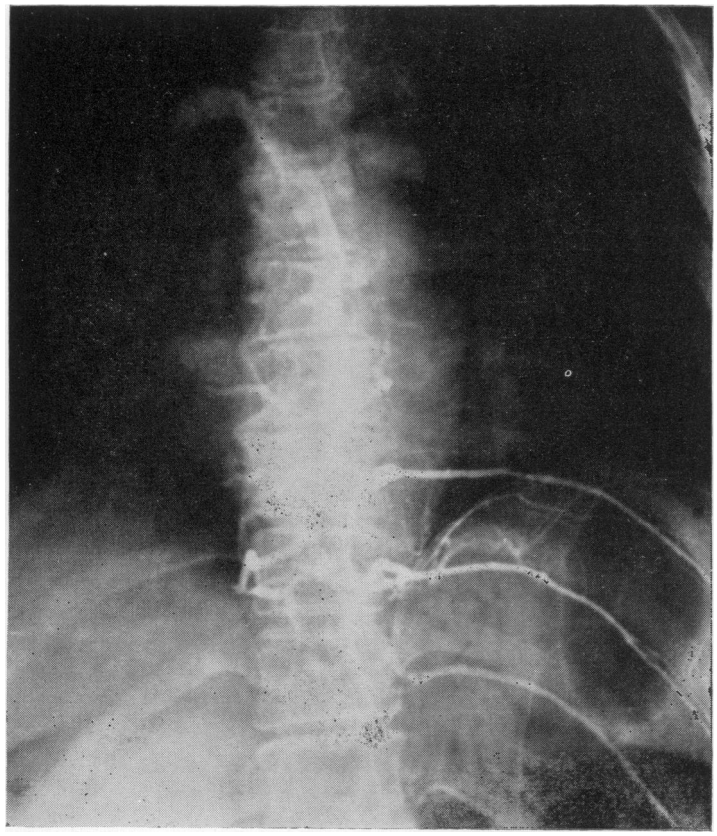

Fig. 2. Azygos venogram showing normal vascular pattern.

Summary

The finding of idiopathic oesophageal varices in a 58 year old female is reported. Liver function studies, liver biopsy, intrasplenic pressure, and the splenic and azygos venograms have all been normal. Injection of the varices is considered the treatment of choice in this case.

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