

ARE WE MISSING PORTAL VEIN THROMBOSIS?

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ABSTRACT

We report a case of portal vein thrombosis which was misdiagnosed as tropical splenomegaly syndrome. This is the first documented case confirmed radiologically at Universiti Hospital, Kuala Lumpur. A discussion on the management of portal vein thrombosis is also described.

Key Words: Portal Vein Thrombosis, Tropical Splenomegaly Syndrome.

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INTRODUCTION

Portal vein thrombosis (PVT) is a frequent cause of portal hypertension in children and in 50% of adults presenting with non-cirrhotic extra-hepatic portal venous obstruction (1). The two most common modes of presentation are variceal bleeding or splenomegaly (2). Infection remains the commonest aetiological factor with umbilical sepsis during neonatal period accounting for the majority of them. This infection spreads along the umbilical vein via the left portal vein to the main portal vein. However there remains about 50% where aetiology of the blocked portal vein is unknown (1). Prognosis is relatively good and variceal bleeding is responsible for most of the death (1, 2).

CASE REPORT

A 27-year old Indian lady presented with eighteen years history of abdominal swelling. Splenomegaly was noted at the age of nine when she was treated for malarial infection. A year prior to admission she was investigated elsewhere with a diagnosis of tropical splenomegaly syndrome. She was referred to Universiti Hospital for a cataract operation and was noted to have gross splenomegaly associated with pancytopenia. There was no stigmata for chronic liver disease.

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Investigations Showed:

Hb 6.9 g/100 ml, platelets 38,000, white blood count 1200, sedimentation rate 22 ml/hr, proteins 85 g/l, albumin 43 g/l, bilirubin 25 umol/l, alkaline phosphatase 69 U/l (normal 34-135), AST 22 U/l (normal 7-40), ALT 27 U/l (normal 4-54), BUSE normal, malarial antibody negative, HBsAg not detected, IgG 3950 mg/dl (normal 639-1349), IgA 271 mg/dl (normal 70-312), IgM 239 mg/dl (normal 56-352). Bone marrow trephine showed a cellular marrow with normal haemopoiesis. Ultrasound showed grossly enlarged spleen with a lot of surrounding collaterals. Liver was not enlarged. Portal vein was not visualized. Liver-spleen scan showed homogeneous colloid accumulation in the markedly enlarged spleen which extends below the umbilicus toward the right iliac fossa. Uptake in the liver is homogeneous (Figure 1). Endoscopy showed oesophageal varices. Liver biopsy was performed without any complication. Histology was normal with preservation of the liver architecture and no evidence of cirrhosis was seen.

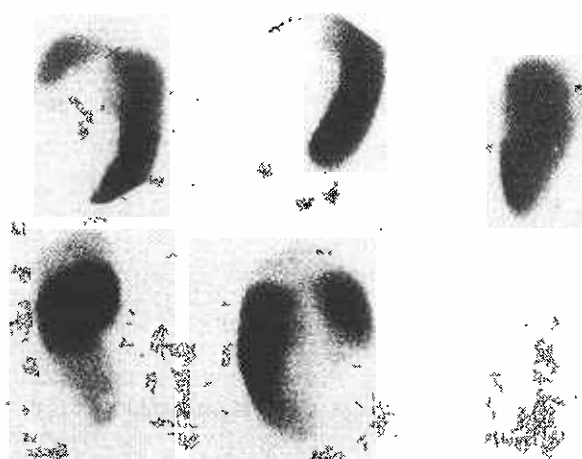


Figure 1: Liver Spleen Scan.

A coeliac axis arteriogram showed no viewing of the portal vein on the venous phase indirectly suggesting portal vein thrombosis (Figure 2). A direct spleno-porto venogram was contraindicated because of the thrombocytopenia. Since the portal vein was not visualized on ultrasound and with a normal liver scan and biopsy, a diagnosis of portal vein thrombosis was made.

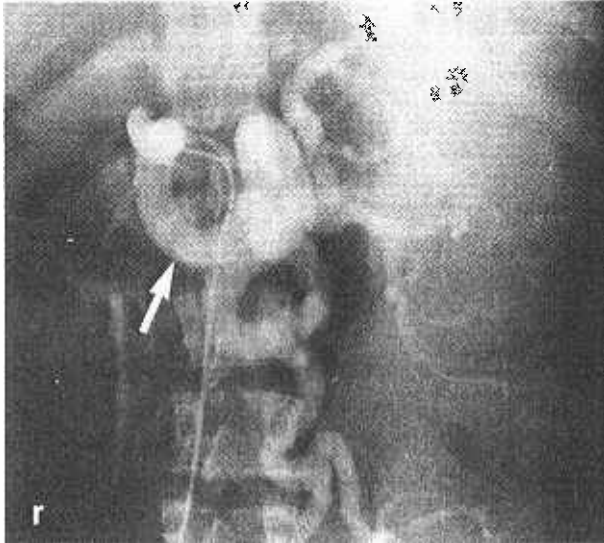


Figure 2a:
Coeliac axis angiogram:- Arterial phase showing enlarged splenic artery (↑) and branches. The catheter is in the coeliac axis which is displaced to the right.

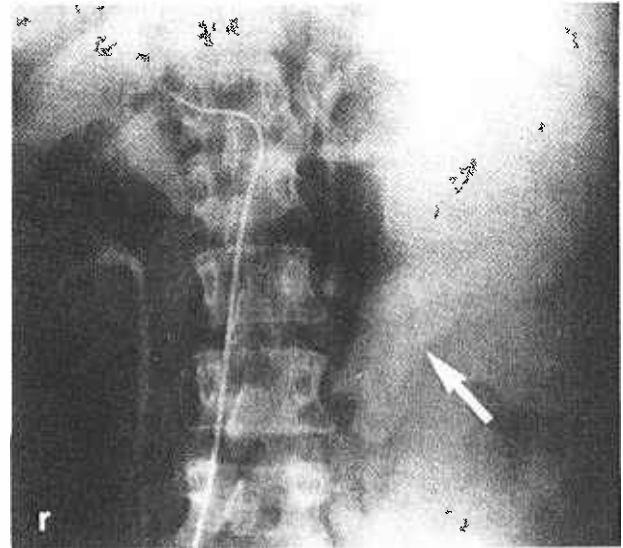


Figure 2b:
Venous phase of selective coeliac axis angiogram showing an enlarged splenic vein (↑) with a diameter of 27 mm. The portal vein is not visualised.

DISCUSSION

This patient was originally diagnosed as tropical splenomegaly syndrome in view of the finding of gross splenomegaly, normal liver function test and previous malarial infection. Investigation here showed otherwise with portal vein thrombosis (PVT). This is the first case of PVT confirmed at Universiti Hospital, Kuala Lumpur.

The management of PVT is not well-defined. Surgery on the whole remains unsatisfactory for control of variceal bleeding. In one large series of 97 patients with PVT (1), splenectomy was the least successful operation associated with the highest complication rate and re-bleeding. Eighteen of the nineteen patients who had splenectomy re-bled an average of 1.9 years later. The authors suggested if surgery was contemplated, the operation of choice is meso-caval shunt (1, 3). Even then, the authors commented on the difficulty of finding suitable veins. Variceal bleeding is the commonest cause of death but despite this, on the whole this is easily controlled with Sengstaken-Blakemore tube, vasopressin and sclerotherapy. This is not surprising in view of the relatively good liver function and blood coagulability.

Liver failure is seldom precipitated by a bleed unlike others with liver cirrhosis. Obliterative endoscopic sclerotherapy has been generally accepted now as the treatment of choice for acute variceal bleeding and prevention of further recurrent bleeding.

The term idiopathic tropical splenomegaly was coined to describe cases of pancytopenia associated with massive splenomegaly in tropical areas where aetiological factors were not understood in the past due to lack of investigational facilities. However a distinct entity called "Tropical Splenomegaly Syndrome" or "Hyperactive Malarial Splenomegaly" is now recognised. The criteria are residence in malarious areas, chronic splenomegaly, elevated IgM, positive malarial antibody levels, hepatic sinusoidal lymphocytosis and a clinical and immunological response to anti-malarial drugs (4).

It is interesting to speculate that we may be missing cases of PVT which were diagnosed as tropical splenomegaly in the past even though they do not satisfy the criteria as defined above. PVT should be considered in any patient with unexplained splenomegaly associated with portal hypertension. However one should also be aware that tropical splenomegaly is a recognised cause of portal hypertension (5).

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