

BILATERAL NEPHROMEGALY, HEPATOMEGALY AND RETROAORTIC LEFT RENAL VEIN IN A PATIENT WITH APLASTIC ANAEMIA: AN UNUSUAL PRESENTATION

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ABSTRACT

This paper reports on a rare picture of hepatomegaly and bilateral nephromegaly along with retroaortic left renal vein which were noticed in a seventeen-year old male cadaver during routine dissection. The patient had been diagnosed with very severe aplastic anemia and treated with thymogam. He had received multiple transfusions and eventually died of septicaemic shock. Histological study of tissue samples of liver revealed portal canal fibrosis, bridging fibrosis, venous congestion, sinusoidal dilatation, hypertrophied Kupffer cells loaded with pigments, while kidney sections showed venous congestion without any major structural abnormality. From these observations we conclude that the patient had developed cardiac failure during the course of his ailment. We also concluded that the bilateral nephromegaly noted on routine dissection could be due to an underlying leukemic or myelodysplastic disorder whose presence is masked both clinically and diagnostically by the severe aplastic anaemia. The retroaortic course of the left renal vein was considered to be an incidental finding as no changes were noted solely on the left kidney though it has a possibility of causing retroaortic renal vein hypertension as co-morbidity.

Key words: aplastic anaemia, nephromegaly, hepatomegaly, retroaortic left renal vein, Myelofibrosis, leukemia

INTRODUCTION

Aplastic anemia is a condition where the body, due to some insult to the cytotoxic cells, mounts an autoimmune response against the hematopoietic stem cells, thus affecting all cell lines producing a pancytopenia¹. The presence of bilateral nephromegaly, hepatomegaly and retroaortic left renal vein are confounders, as they are not usually found in a person with aplastic anaemia.

The left renal vein develops as a remnant of the anastomotic venous network between sub and supracardinal veins in embryo. Abnormal persistence or deletion of any segment of this venous network leads to anomalous course of left renal vein with reference to its relation to aorta such as circumaortic venous ring or retroaortic renal vein^{2,3}. The retroaortic renal vein is frequently noticed incidentally in donor nephrectomy cases^{4,7}. Isolated retroaortic renal vein generally remains asymptomatic without causing discomfort to the individual^{3,6,8}. However, if compressed by the related aorta, it can lead to retrograde venous congestion in the kidney^{3,8}.

In this study we discuss a case of aplastic anaemia

with bilateral nephromegaly, hepatomegaly and a retroaortic left renal vein. We reviewed literature to look for associations between these findings in aplastic anemia.

OBSERVATION

Abnormalities of few abdominal viscera were noticed in a seventeen-year old male cadaver during routine dissection demonstration to undergraduate medical students in our department. The liver was enlarged and extended seven centimeters below the costal margin; the surface was smooth and no visible nodule was noticed. The vasculature and extrahepatic biliary drainage apparatus were typical in formation and course. The kidneys were enlarged and displayed ill-defined lobes; however there was no scarring. The right kidney measured 14 X 9 X 5 cm while the left measured 14 X 9 X 6 cm. The arteries to both the kidneys took a normal course. The right renal vein typically drained into the inferior vena cava, while the left renal vein passed behind the abdominal aorta to drain into the IVC at an acute angle at L3 vertebral level. The left renal vein received the left gonadal and the left suprarenal veins (Fig 1 A); the suprarenal vein was found to be running down as a continuation of left inferior phrenic vein. We also noticed reduction in the thoracic volume due to the enlarged abdominal organs pushing the diaphragm upwards (Fig.1 B). The heart and the lungs however showed normal gross anatomy picture.

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Samples of liver and kidney tissues were taken and processed for routine hematoxylin and eosin staining to evaluate the microscopic morphology of these organs. Liver showed presence of venous congestion, portal area fibrosis and bridging fibrosis (Fig. 2 A), and pigment laden Kupffer cells in the wall of dilated sinusoids (Fig. 2 B). We could not make out hepatic venule dilatation in sections. Venous congestion was seen in kidney tissue with no structural anomaly (Fig. 3).

Examination of his medical history revealed that he had expired due to complications of aplastic anaemia (diagnosed by bone marrow biopsy) within two months of onset of initial symptoms (symptomatic anemia and melena). He had suffered from pancytopenia and lower gastrointestinal bleeding, undergoing gastrointestinal tract scopies and angiogram for the same. Ultrasound abdomen was a normal study except for the enlarged kidneys. Liver function test was normal except for hypoalbuminemia. Immunophenotyping for PNH was negative. He was treated with thymogam for his aplastic anaemia and had multiple transfusions with blood products. He eventually developed a fungal pneumonia and died of septicaemic shock.

DISCUSSION

The present case revealed the following abnormal conditions viz. hepatomegaly, bilateral nephromegaly and a retroaortic course taken by the left renal vein to reach the inferior vena cava. The patient had expired due to complications of severe aplastic anemia. Hepatomegaly, nephromegaly and retroaortic renal vein are not features usually seen in aplastic anemia. The histological features of the liver viz. sinusoidal dilatation, fibrosis of portal canal are features of aplastic anemia of which the patient suffered. The hepatomegaly can be attributed to the multiple transfusions given over a short span of time causing congestive cardiac failure. However, we could not see hepatic venule dilatation, nor was there perisinusoidal fibrosis in this case. Since nephromegaly is not usually a finding in a person in cardiac failure, literatures were reviewed to look for other possible causes for the organ enlargement. It was found that kidney can act as a focus of extramedullary haematopoiesis in conditions where there is depletion of bone marrow space, like myelofibrosis. This can present as a bilateral nephromegaly⁹. It was also found that bilateral nephromegaly can arise due to lymphoblastic infiltration in leukemic disorders¹⁰. No literature was found linking the nephromegaly to aplastic anaemia.

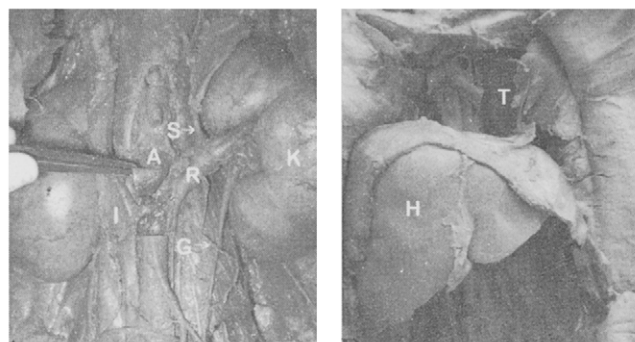


Figure 1-
A: Shows the enlarged kidneys (K) with a left renal vein (R) passing posterior to the abdominal aorta (A) and draining into the inferior venacava (I). The left gonadal vein (G) and the left suprarenal vein (S) can be seen draining into it.
B: Shows hepatomegaly (H) and the reduced thoracic cavity (T).

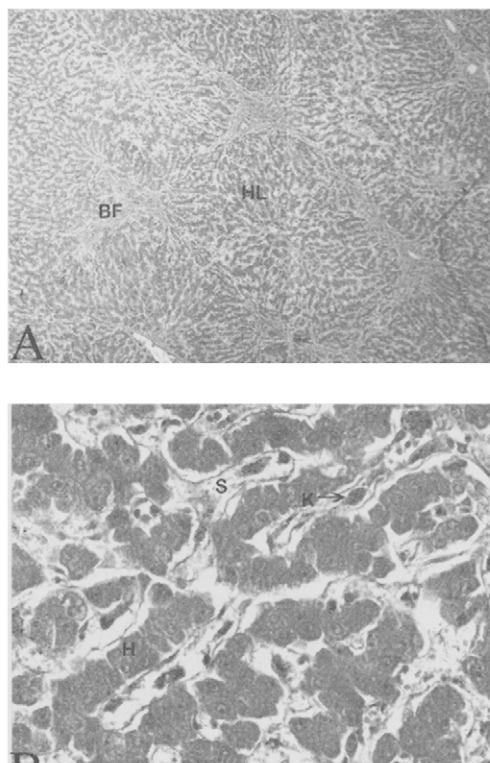


Figure 2-
A: Shows the hepatic lobule (HL) at 40x magnification. Bridging fibrosis (BF) can be seen extending between the hepatic lobules.
B: Shows the hepatocytes (H) and the sinusoids (S) at 400x magnifications. Pigment laden Kupffer cells (K) can be seen in the dilated sinusoids.

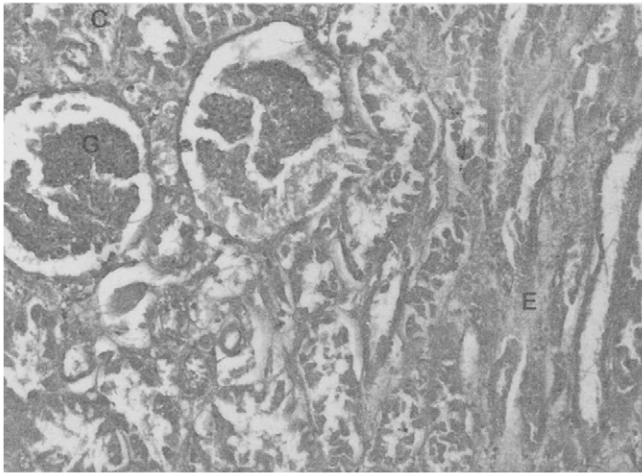


Figure 3 - Shows the cortex of the left kidney at 200x magnification. There is presence of venous congestion(C) and extracellular extravasation of fluid (E). The glomeruli (G) can be seen.

It has been shown that some patients with severe aplastic anaemia undergoing immunosuppressive therapy with antilymphocyte/ antithymocyte globulin tend to develop acute leukemic or myelodysplastic disorders post treatment. This happens by a process known as “field leukemogenic effect”, where there is an unmasking of secondary clonal disorders in the bone marrow cells when the primary disorder i.e. aplastic anaemia is treated¹¹. We suggest this as a possible reason for the nephromegaly seen in this case. This conclusion presented us with a new question as to the normal renal parameters of this patient on investigation. Further literature review showed that in children with leukemic disorders, asymptomatic bilateral nephromegaly was sometimes seen with no abnormal renal symptoms or abnormal biochemical parameters¹². Aplastic anaemia, which happens to be the dominant presenting feature, seems to have suppressed the diagnostic features of the secondary haematological disorder causing the nephromegaly.

The retroaortic course of the left renal vein seems to be an incidental finding as no change was noted solely on the left kidney macroscopically or microscopically.

We conclude that liver presented features of aplastic anemia and the features of kidney in this case could be a treatment related abnormality.

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