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Hyperbilirubinemia Following Retroperitoneal Mass Resection in a Patient with Sickle Cell Anemia

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CASE

A 46-year-old man with sickle cell anemia was taken to the operating room for resection of a 19 cm right-sided retroperitoneal mass identified on CT performed for flank pain (Fig. 1A). The mass was removed en bloc with the adrenal and kidney (Fig. 1B). Hepatic mobilization with ligation of the venous branches draining the caudate lobe and the right hepatic artery was performed along with a small, non-anatomical partial hepatectomy at a location where the mass was densely adherent to the liver. On post-operative day#2 the patient was jaundiced. Total bilirubin was 24.4 mg/dL (direct component 20.0 mg/dL) with corresponding hemoglobin of 6.8 g/dL and mild transaminitis (Table 1). CT scan only revealed a 6 cm simple fluid collection within the resection fossa, but the bilirubin continued to rise. Magnetic resonance cholangiopancreatography on post-operative day#4 revealed a persistent fluid collection but no biliary ductal dilation. Subsequent cholescintigraphy revealed no extravasation of the ^{99m}Tc-mebrofenin radiotracer.

What Is Your Diagnosis?

- A. Common bile duct injury
- B. Biliary duct obstruction
- C. Hepatic sequestration
- D. Stauffer's syndrome

Diagnosis

- C. Hepatic sequestration

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DISCUSSION

Ligating the right hepatic artery placed the patient at risk for a bile duct injury however the negative Magnetic resonance cholangiopancreatography and cholescintigraphy excluded both bile duct leak and obstruction from the differential. Furthermore, an isolated right hepatic artery injury rarely results in clinical symptoms as blood flow should be maintained via collateral vessels originating from the remaining hepatic and gastroduodenal arteries.¹ Stauffer's syndrome with jaundice was unlikely as its manifestations should improve after tumor resection and in 90% of cases alkaline phosphatase is elevated.² Sickle cell hepatopathies (acute sickle cell hepatic crisis, acute intrahepatic cholestasis, and hepatic sequestration) are a spectrum of hepatic complications caused by intrahepatic sickling and sinusoidal obstruction, most often seen in patients with sickle cell anemia.³ Identifying the predominant sickle cell hepatopathy rely on the degree of conjugated bilirubin elevation and transaminitis.³ This patient likely had hepatic sequestration as these patients typically present with abdominal pain, jaundice, an acute drop in hemoglobin, a rapid rise in conjugated bilirubin, and hepatomegaly resulting from trapped sickled red blood cells in the liver sinusoids.³ Management is supportive with blood transfusion and/or exchange transfusion, and our patient received both.⁴ After the 3 to 4-day acute phase of hepatic sequestration hemoglobin levels increase and occasionally phlebotomy is needed to prevent blood hyperviscosity.⁵ Early recognition of acute sickle cell hepatopathies and prompt involvement of hematology is important as they can be fatal.⁶ Ultimately the patient recovered well and surgical pathology revealed an adrenal myelolipoma.

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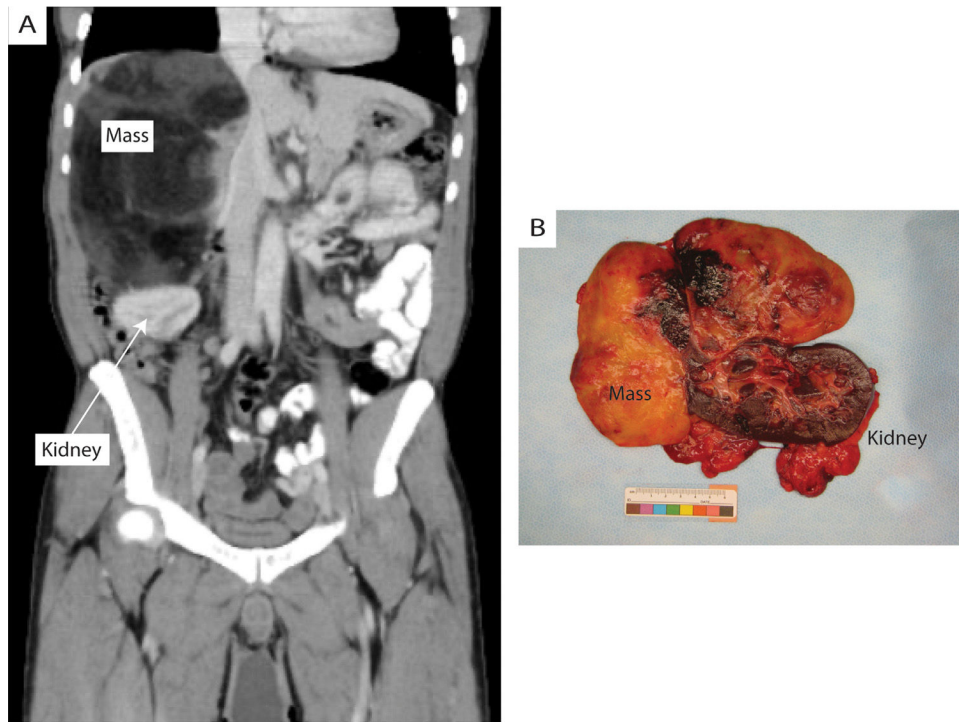


Figure 1. (A) Pre-operative contrast-enhanced CT scan of the abdomen and pelvis demonstrating large right-sided retroperitoneal mass, shown in coronal view. (B) Gross pathology image of resected specimen in cross section.

Table 1.

Relevant laboratory data prior to and following the operation

	Pre-op*	POD#0 (intra-op) [†]	POD#1	POD#2*	POD#3	POD#4	POD#5	POD#6	POD#11	POD#34	POD#61	Reference Values
Hemoglobin	8.7	9.0	9.2	6.8	9.7	11.2	11.0	10.7	11.6	9.9	10.3	13.9–16.3 g/dL
Bilirubin, total	2.4	3.8	9.9	24.4	34.1	34.8	37.6	35.0	13.8	4.3	2.9	0–1.2 mg/dL
Bilirubin, direct	-	-	5.5	20.0	33.0	34.0	36.0	28.4	8.7	2.44	-	0–0.4 mg/dL
ALT	46	63	68	65	63	54	55	58	70	46	42	0–40 U/L
ALP	199	135	136	105	120	112	136	158	310	277	210	30–120 U/L
AST	56	87	133	149	122	80	71	67	68	46	44	0–37 U/L

ALT, alanine amino transferase; ALP, alkaline phosphatase; AST, aspartate amino transferase; POD, post-operative day.

* The patient was transfused 2 units of packed red blood cells (pRBC) prior to the operation, 2 units pRBC on POD#2.

[†] Estimated blood loss from the surgery was 250 mL.