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Case Report

Right side splenorenal fusion with marked extramedullary hematopoiesis, a case report

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ABSTRACT

Developmental abnormalities of the spleen are common, much less common is splenic fusion abnormality. It may present as a renal mass sometimes with symptoms of hypersplenism (anemia) or as a renal mass only, mimicking primary or secondary renal neoplasms on imaging studies. Its documented that occurred on the right side which is significant that provides evidence for the possible migration of spleen cells in embryogenesis as an explanation for some other splenic fusion anomalies.

We report a case of splenorenal fusion in a 5-year old girl with only right functional kidney who initially presented with a large renal mass which mimicked a primary renal neoplasm with no history of splenic trauma or splenectomy in past medical history. Also, the child didn't present with symptoms of hypersplenism. The child underwent right nephrectomy for a renal mass that was subsequently confirmed as right splenorenal fusion pathologically.

1. Introduction

Heterotopic splenic tissue in the renal parenchyma can occur in patients with intact spleen (splenorenal fusion) or those who had history of splenic trauma or had been splenosis. Splenorenal fusion has been previously reported and our case is only the third young child case [1–8]. Splenorenal fusion is a developmental splenorenal fusion has been described in the literature. More rarely, splenorenal fusion may arise as a developmental anomaly secondary to the fusion of nephrogenic mesoderm and splenic anlage in the second month of gestation [9]. Etiologically, it may also be secondarily acquired as a result of splenosis after trauma or splenectomy. Also, the presence of a renal mass in such patients should raise the suspicion of splenosis. Based on previous reports, splenorenal fusion more reported in left kidney [3,4]. Patients usually present with an asymptomatic mass or less likely with hypersplenism symptom clinically.

2. Case report

A 5-year-old girl with history of chronic pyelonephritis due to primary left vesicoureteral reflux referred to our hospital with right upper quadrant. Although, her renal function depended to only right kidney which should be safe for living. On physical examination, a mass in size of about 6 cm in the largest diameter was palpated in right upper quadrant. The routine laboratory tests like urinalysis, complete blood count, and serum chemistries have been done. Evaluation revealed elevated BUN (23 mg/dl) and creatinine levels (2.9 mg/dl) (reference range BUN, 5 to 18 mg/dl, reference range creatinine, 0.3–1.2 mg/dl). She was also anemic (hemoglobin 9.6 g/dl; reference range, mean 13.5 g/dl and hematocrit, 27%; reference range, 36%–45%). Platelets were low at 120×10^3/μl (reference range, 150–400 $\times 10^3$/μl), white blood cell count was normal at 7980/μl (reference range, 4500–11 000/μl). A renal ultrasound revealed a large mass in the right kidney measuring $6 \times 5 \times 4.5$ cm. Abdominal and pelvic computed tomographic (CT) scan showed the $6 \times 4.5 \times 4$ cm mass located at middle and upper poles of right kidney near to renal sinus and due to deforming the kidney with probably intact perinephric fat, Gerota fascia and renal capsule (Fig. 1). The primary suspicion was Wilms tumor or clear cell carcinoma. She underwent a radical nephrectomy 7 days after admission and clinical work up.

3. Pathologic findings

At surgery, the upper and middle poles of the right kidney were markedly enlarged. The Gerota fascia and renal capsule seems to be intact (Fig. 2A). Macroscopic examination of the removed kidney tissue showed a large kidney with a bulged and brownish external surface with intact capsule (weighed 680 g and measured $11 \times 9 \times 5.5$ cm). On cut section, renal tissue has been replaced by a well-defined, congested and hemorrhagic mass measuring $6 \times 4.5 \times 4$ cm in size (Fig. 2A).

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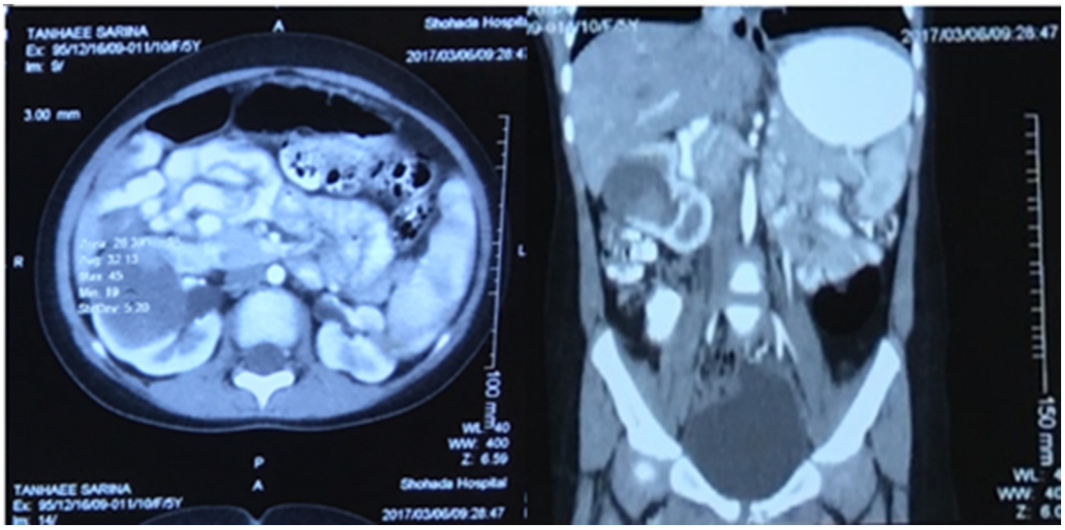

Fig. 1. Computed tomographic scan of the abdomen displyed a hypodence well defined soft tissue mass in kidney parenchyma.

Microscopic examination revealed an intraparenchymal splenic tissue with congestion that was separated of the unremarkable renal parenchyma with a definite pushing border. (Fig. 3B) Foci of extramedullary hematopoiesis with erytroid, myeloid and megakaryocytic precursors associated with few macrophages have been identified (Fig. 2C). There were also scattered lymphoid aggregates like white pulps (CD20 positive in matured lymphocytic cells) (Fig. 2D). Adrenal tissue was intact. There is no evidence of a primary renal malignancy. The patient was discharged after 4 days. During the postoperative follow-up, the patient was in a good condition. There were no complications from the procedure. About two months postoperatively, all values of hematological indices back to normal range.

4. Discussion

Developmental abnormalities of the spleen are common, most often consists of an accessory spleen in the area of the splenic hilum, which is estimated 10%–30% of patients in autopsy series and 16% of patients undergoing contrast enhanced abdominal CT [10]. Much less common is splenic fusion abnormality which include the well-described in gonads, liver, retroperitoneal but still rare in kidney [11]. Splenogonadal, splenopancreatic and splenorenal fusion anomalies may also be detected incidentally. This rare anomaly may result from disturbances in the embryogenesis because both organs arise from dorsal mesogastrium near each other may result in a fusion. A review of the literature revealed

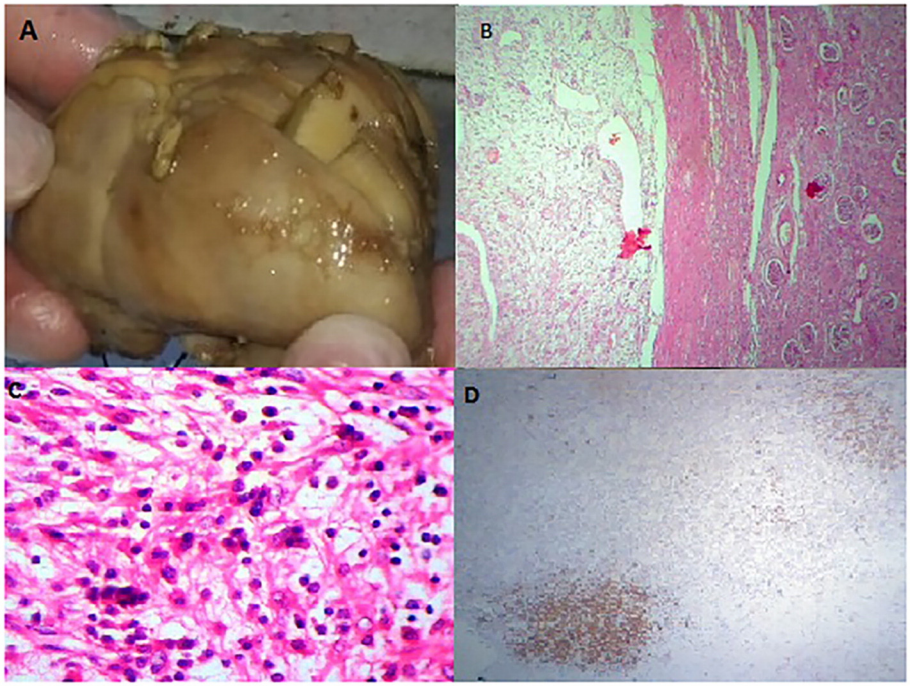

Fig. 2. (A,B,C,D). A, Surgical specimen showing the mass in the superior and middle poles in the right kidney with firm white areas of dense fibrous/scar renal tissue that surrounding dark red splenic tissue. B, Histologic sections revealed splenic tissue with red pulp and dilated sinuses. C, Histologic sections revealed marked extramedullary hematopoiesis with prominent megakaryocytes. (B, C; hematoxylin-eosin, original magnification ×400). D, Immunohistochemical stains highlight the positive population of CD20-positive B-lymphocytes in a focus of white pulp in a background of red pulp. (D; Immunostain CD 20, original magnification ×400).

six cases of left-sided renal splenosis and three on the right. Seven cases was adult and two cases were children [1–9]. Recognition of this anomaly is important because definite diagnosis can prevent an unnecessary invasive and costly procedures like nephrectomy. Even though, previous splenorenal fusion cases were mostly symptomatic and reported in left side and in adult. We report a case of right side splenorenal fusion in 5 years old girl with anemia and thrombocytopenia which was not definitely diagnosed by CT scan and MRI only. Typically, splenorenal fusion as a renal mass cannot be reliably distinguished from clear cell carcinoma, Wilms tumor or other renal neoplasms by sonography, CT, or angiography. However, if splenorenal fusion is suspected, the splenic tissue can be demonstrated with 99 m Tc-sulfur-colloid scan or 99 m Tc–labeled, heat-damaged red blood cell scan [12,13]. If a mass is small, fine-needle aspiration (FNA) can be next step to confirm the diagnosis but it is not a gold standard test. However, red and white blood cells in FNA smear can be detected in possible organizing hematoma or chronic inflammation. Therefore, the gold standard test for definite diagnosis of splenorenal fusion is histopathology assessment on renal mass biopsy.

Additionally, splenorenal fusion is a rare mass in kidney that should be considered for each patient who diagnosed for renal mass. Preoperative of diagnostic procedures such as CT scan, MRI could not be helpful for definite diagnosis because mimic a neoplasm on imaging. 99 m Tc-sulfur-colloid scan or 99 m Tc–labeled, heated red blood cell scan associated with fine needle aspiration of mass can confirm diagnosis. The gold standard test for definite diagnosis of splenorenal fusion is histopathology assessment on renal mass biopsy specimen to make safe renal parenchyma for the patient is very important to avoid unnecessary nephrectomy especially for patient who living with only one functional kidney.

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Conflicts of interest

No conflicts interest

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