Renal lymphangiomatosis during pregnancy: management with percutaneous drainage

Abstract We report a unique case of exacerbation of renal lymphangiomatosis during pregnancy which was managed percutaneously until delivery. Renal lymphangiomatosis is a very rare benign disorder that might cause abdominal pain and rarely hypertension and hematuria. Surgical treatment options may result in nephrectomy. Percutaneous drainage of symptomatic renal lymphangiomas should be viewed as an efficient therapeutic option particularly when surgery is contraindicated.

Key words Renal lymphangiomatosis · US · CT

Introduction
Renal lymphangiomatosis is a very rare disorder that may be confused with other renal cystic diseases. There are few cases reported in the literature and only one case report with an exacerbation during pregnancy [1, 2, 3, 4, 5]; however, this is the first case of a pregnant woman with bilateral renal lymphangiomatosis, hypertension, and severe dyspnea who was successfully managed percutaneously until delivery.

Case report
A 35-year-old pregnant woman (gravida 1, para 0) was admitted to another institution with complaints of right flank pain, hematuria, and hypertension. Ultrasonography revealed a 26-week pregnancy and bilateral multiple renal cysts. There was also a right perirenal retroperitoneal collection which was catheterized with a presumptive diagnosis of urinoma in the other health facility. Daily drain-age of perirenal fluid was approximately 1500 cc. The patient was referred to our hospital with the diagnoses of hypertension, cystic renal disease, and urinoma.

On the initial ultrasonographic examination, both kidneys were enlarged with the parapelvic located multicystic lesions (Fig. 1). Renal parenchymal thickness was within normal limits (15 mm). No cystic lesion was identified within the parenchyma and there was no loss of corticomedullary differentiation. On the right side there was a huge perirenal retroperitoneal multiseptated collection (30 × 18 × 15 cm) despite the catheterization. Much smaller perirenal cystic lesions were also seen on the left side. These ultrasonographic findings prompted the diagnosis of renal lymphangiomatosis.

Renal function tests were normal. The biochemical examination of the drained fluid revealed that it was compatible with lymphatic fluid but not with urine, supporting the diagnosis of renal lymphangiomatosis (protein:0.4 g/dl; urea: 9 mg/dl; creatinin: 0.4 mg/dl). Total daily drainage was approximately 2000–2500 ml. Total blood count showed hypochromic microcytic anemia (Hb: 8.5 g/dl) and leukocytosis (13,600/µL). Microorganisms isolated from drain fluid and fever were the evidence of drain infection. In addition to antihypertensive medication, appropriate antibiotics regimen was initiated.
Fig. 1 Transverse sonogram of the right kidney reveals medial replacement of the kidney with huge amount of perirenal fluid collection. There are also multiple parapelvic cystic lesions.

Fig. 2 Transverse sonogram of the right kidney obtained 3 weeks after the initial scan. Note that besides the reduction of the perirenal fluid volume, there is an increase in number of septations in the cystic area.

Fig. 3 Postpartum contrast-enhanced CT demonstrates cystic lesions in parapelvic and perirenal areas of both kidneys.

Fig. 4 Sagittal sonograms of the right kidney obtained 4 months later. Note that there is considerable diminution of the perirenal collection on the right as compared with Fig. 1.

On the seventh day of hospitalization, since there was a considerable loss of fluid and electrolytes and no decrease in the amount of daily drainage, it was decided to withdraw the catheter. Since the patient was pregnant, surgery was not considered, but conservative management was preferred.

One week later, in addition to fever and leukocytosis, severe dyspnea, abdominal discomfort, and right flank pain occurred; hence, the right perirenal area was recatherized with an 8-F pigtail catheter under US guidance and the fluid and electrolyte loss was covered by ringer lactate and amino acid solutions.

Repeated US scans revealed that the volume of the perirenal fluid gradually decreased in correlation with the gradual decrease of the daily drainage. During this period, the number of septations in the cystic area increased (Fig. 2). Repeated microscopic examination of the drained fluid showed no evidence of infection and also the white blood count was normal.

On the 29th day of hospitalization the catheter was withdrawn since daily drainage was < 50 ml. Subsequently, symptoms such as dyspnea and abdominal pain did not recur.

After a follow-up of 3 weeks, a cesarean section was performed at the gestational age of 34 weeks, yielding a premature child of 1700 g and an Apgar score of 10.

In the first postpartum week abdominal CT was obtained. Computed tomography showed cystic lesions in parapelvic and perirenal areas of both kidneys (Fig. 3). Control abdominal US performed 4 months later revealed considerable diminution of the perirenal collections (Fig. 4). During the follow-up period of 13 months, no antihypertensive medication was needed and symptoms such as dyspnea, abdominal discomfort, pain, and hematuria did not occur.