Hygroma Renalis: An Extremely Rare Renal Lesion

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INTRODUCTION
Lymphangiomas are benign tumors or malformations of lymphatic vessels. They are most commonly seen in the neck, axillary region, and mediastinum. Hygroma renalis is a type of lymphangioma (lymphangiectasia) located in the pericystec area and is therefore named as pericystic lymphangiectasia.\(^{1,2}\) Hygroma renalis is usually asymptomatic and easily detectable by ultrasonography or computed tomography (CT). We present a patient with vague right lumbar pain and fullness. Radiologic examination revealed a multicystic pericystic and perirenal lesion. Histological examination and immunohistochemistry analysis were consistent with the diagnosis of hygroma renalis.

CASE REPORT
A 30-year-old woman presented to our center complaining of vague right lumbar pain and fullness. She had undergone partial nephrectomy about 4 years before due to a simple renal cyst associated with chronic nonspecific pyelonephritis. Physical examination, kidney function tests, and hemagglutination test were negative for hydatid cyst. Abdominal ultrasonography and CT scan revealed a huge multiloculated cystic mass that was attached to the atrophic right kidney. The mass was extended up to the umbilicus via the retroperitoneum with sharp lobulated contours (Figure 1). The left kidney, liver, pancreas, spleen, and bladder were normal. No intraperitoneal fluid accumulation was detected.

The patient underwent right nephrectomy. A 700-g mass sized \(16 \times 10 \times 8\) cm was excised. On pathologic examination, remnants of the kidney surrounded by a...
A multiloculated cyst were identified, extending to
the pericalyceal and hilar area covering the ureter.
The cyst contained a creamy fluid and its wall was
extremely thin; in most areas, the thickness was about
1 mm (Figures 2 and 3). Microscopic examination
revealed a multicystic lesion with flat endothelial cells.

The cystic spaces were filled with a proteinous fluid
and thin fibrous walls. The renal tissue had features
of nonspecific chronic pyelonephritis without any
intraparanchymal cyst (Figures 3 to 5). The most possible histopathological diagnosis
was cystic lymphangiomia; however, in order
to rule out other probable diagnoses such as
mesothelial, epithelial, and mullerian serous cysts,
immunohistochemical staining was performed
using cytokeratin and calretinin and both were
reported to be negative. Thus, the final histologic
diagnosis was hygroma renalis. The patient made a
complete recovery and was disease-free 20 months
postoperatively.

DISCUSSION

Lymphangiomas are benign malformations or
postulate, benign, cystic tumors of the lymphatic
vessels that are most frequently discovered in
childhood with few cases reported in adults.(1) They
are mostly located in the neck, axillary region,
mediastinum, abdominal cavity, and in extremely
rare cases, in the retroperitoneum (less than 1% of
all lymphangiomas). (2) Four histologic subtypes of
lymphangiomia have been described that include
cystic, papillary, cavernous, and vascuolymphatic
malformations. A combination of these types may be
seen in the same lesion. The presence of endothelial-
lined lymphatic channels separated by the connective
tissue is the main histologic feature of the disease.(3)
In the renal region, these lesions are often located adjacent or attached to the kidney, especially around the renal pelvis and are, therefore, called pericalyceal lymphangiectasis (lymphangioma) or hygroma renalis. The lesion may consist of a single or multiple cysts and may be either unilateral or bilateral. (3) It has variously been considered to be hamartomatic malformation analogous to cystic hygromas of the head, neck, and mediastinum, as an acquired lesion resulting from lymphatic obstruction, or as a true vascular neoplasm. Occasionally, the lymphatics of the renal capsule are also involved and therefore, the whole kidney is covered by cysts that give a superficial appearance similar to polycystic disease. Although preoperative diagnosis is usually possible by CT or magnetic resonance imaging, confirmatory diagnosis of hygroma lesions requires laparotomy and complete excision in most cases, followed by histopathological examination. (4,5) Confusion with other cystic lesions of the kidney such as calyceal diverticula, renal cysts, and cystic tumors of the liver, pancreas, and retroperitoneum may occur. (6)

To our best knowledge, only 24 cases of hygroma renalis have been previously reported in the world literature. (2) It is usually asymptomatic, but it may be associated with urinary tract obstruction that can be treated by conservative management. Operation should be considered only in patients with complications. (2) In the case of surgery, simple resection of the hygroma is adequate and radical nephrectomy is contraindicated in the management of uncomplicated patients or patients with normal kidneys. (2) Although very uncommon, potential aggressive behavior of lymphangiomas has been reported. (7,8)

REFERENCES