

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 pericalyceal area and is therefore named as *pericalyceal 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 pericalyceal and perirenal lesion. Histological examination and immunohistochemistry analysis were consistent with the diagnosis of hygroma renalis.

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 × 10 × 8 cm was excised. On pathologic examination, remnants of the kidney surrounded by a

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CASE REPORT

A 30-year-old woman presented to

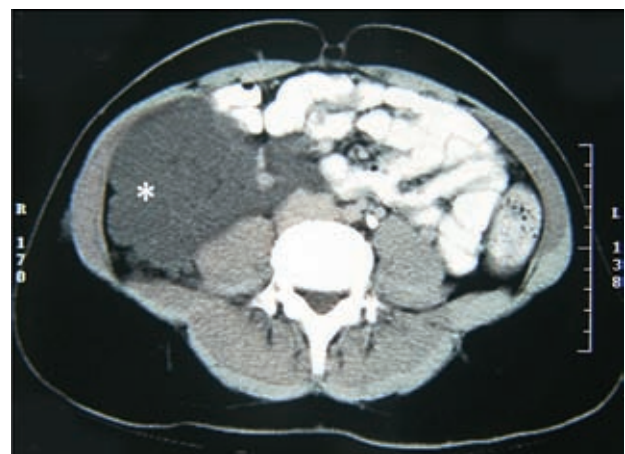
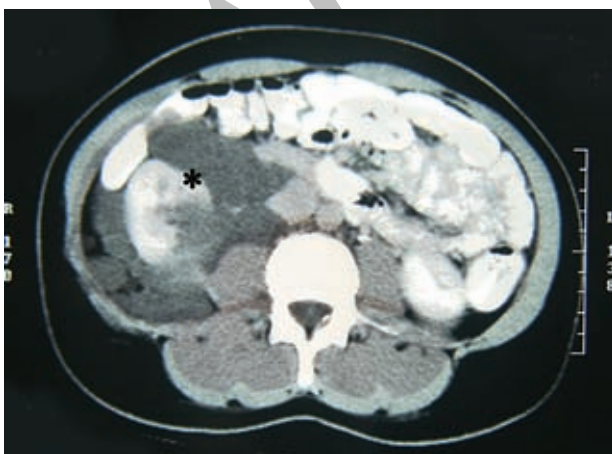


Figure 1. CT Scans. Left, a cystic mass surrounding the right kidney (black asterisk). Right, the obvious cystic mass with sharp and lobulated border extending near the midline (white asterisk).

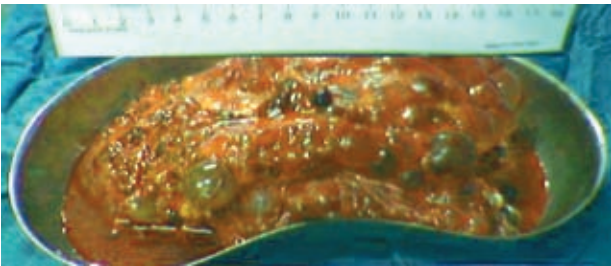


Figure 2. Gross appearance of the lesion diagnosed as hygroma renalis.

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.

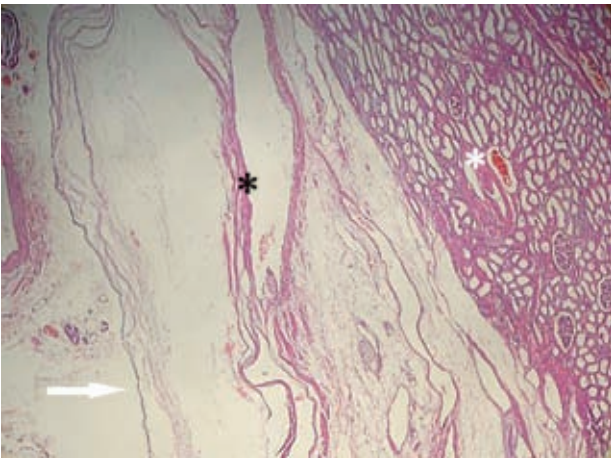


Figure 3. Microscopic view shows the thin wall of the cyst (arrow) adjacent to the renal tissue (white asterisk) with ectatic capsular vessels (black asterisk; hematoxylin-eosin, $\times 100$).

The cystic spaces were filled with a proteinous fluid and thin fibrous walls. The renal tissue had features of nonspecific chronic pyelonephritis without any intraparenchymal cyst (Figures 3 to 5).

The most possible histopathological diagnosis was cystic lymphangioma; 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.

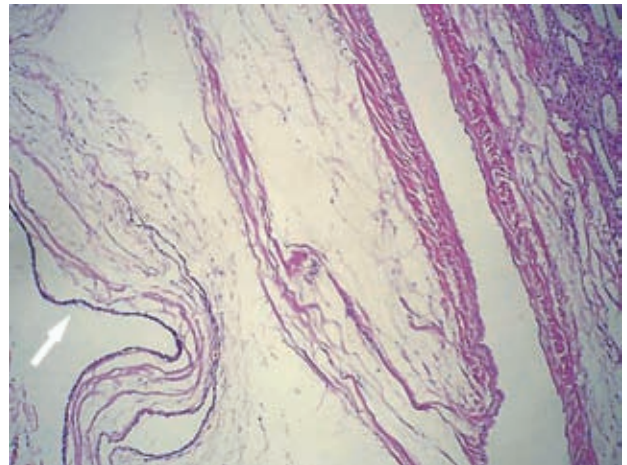


Figure 4. A close-up view of Figure 4 demonstrating the cyst wall (arrow) lined by a layer of the flat cells (hematoxylin-eosin, $\times 200$).

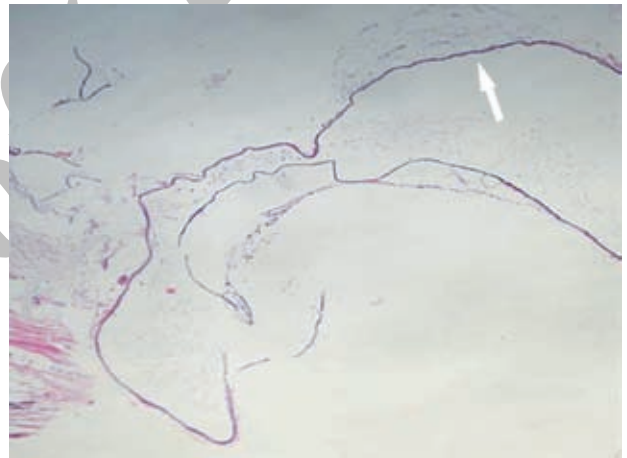


Figure 5. Thin wall of the cyst (arrow) lined by one layer of flat cells containing proteinous material (hematoxylin-eosin, $\times 100$).

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 lymphangioma have been described that include cystic, papillary, cavernous, and vasculolymphatic 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.⁽²⁾

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 hamartomatous 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)

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