



## Case report

# Perirenal cystic lymphangioma mimicking a renal cyst in an elderly patient: Case report



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## ABSTRACT

Cystic lymphangioma is a congenital benign lymphatic tumor commonly diagnosed in childhood. Cervical and axillary locations are the most usual, while retroperitoneal involvement is exceptional. Perirenal cystic lymphangioma may be revealed by urological signs or by incidental findings. Imaging plays an essential role in characterizing the tumor, however differential diagnosis with benign or malignant renal tumors is not always possible, thus requiring histological evidence. We report the case of a 50-year-old man who presented with a cystic lymphangioma of the left kidney revealed by lumbar pain with imaging features mimicking an exocortical developing renal cyst. The work has been reported in line with the SCARE criteria.

## 1. Introduction

Cystic lymphangioma is a congenital benign lymphatic tumor most often discovered during the first two decades of life. It occurs preferentially in the cervical and axillary region, but all other regions of the body may be involved except the brain. Abdominal cystic lymphangiomas are rare and those of retroperitoneal location are exceptional and represent less than 1 % of abdominal cystic lymphangiomas [1]. Although benign, cystic lymphangioma can lead to severe complications depending on its location. We report a case of perirenal cystic lymphangioma, revealed by a urinary symptomatology. The work has been reported in line with the SCARE criteria [9].

## 2. Case presentation

A 50-year-old patient was experiencing recurrent episodes of low back pain. He had no particular family, medical or surgical history. His symptomatology dated back to more than a year ago, with the appearance of left lumbar pain, which were neglected thanks to self-medication with a level I analgesic of the paracetamol type, which initially relieved his discomfort. The evolution was marked by the recrudescence of the painful episodes which became more and more severe as well in intensity as in duration and yielding less and less by the self-medication. This motivated a consultation in general medicine during which, the clinical examination as well as the urinary strip were without anomaly. In front of this clinical symptomatology, the diagnosis of a renal lithiasis

disease generating episodes of renal colic was evoked, and a biological assessment evaluating the renal function, as well as a renal and bladder ultrasound were prescribed in first intention. The biological workup came back normal with a preserved renal function. The renal ultrasound suggested a renal cyst of superior polar location with *exo*-renal development to be better characterized by a CT urogram. The uroscanner showed secreting and excreting kidneys at physiological times without any obstruction on the excretory tracts with however, an isolate unilocular cystic mass developing in the left perirenal space (Fig. 1). It was well limited with bumpy contours, not enhanced by iodinated contrast, measuring 110x50x30mm or a volume of 82 ml, and developed in contact with the posterior and superior part of the left kidney which was pushed forward and which however kept an intact capsule allowing to evoke the diagnosis of perirenal cystic lymphangioma. No associated lesions were found either in the contralateral kidney or intraperitoneal. This mass, because of the symptomatology it generated, was resected surgically despite its radiological signs of benignity. This consisted of a perikystectomy during open surgery. Histological examination revealed a benign tumor proliferation made of multiple cystic cavities of variable size, bordered by endothelial cells without atypia, which confirmed the diagnosis of a perirenal cystic lymphangioma. The postoperative course was favorable.

## 3. Discussion

Cystic lymphangioma is a benign lymphatic tumor of malformative

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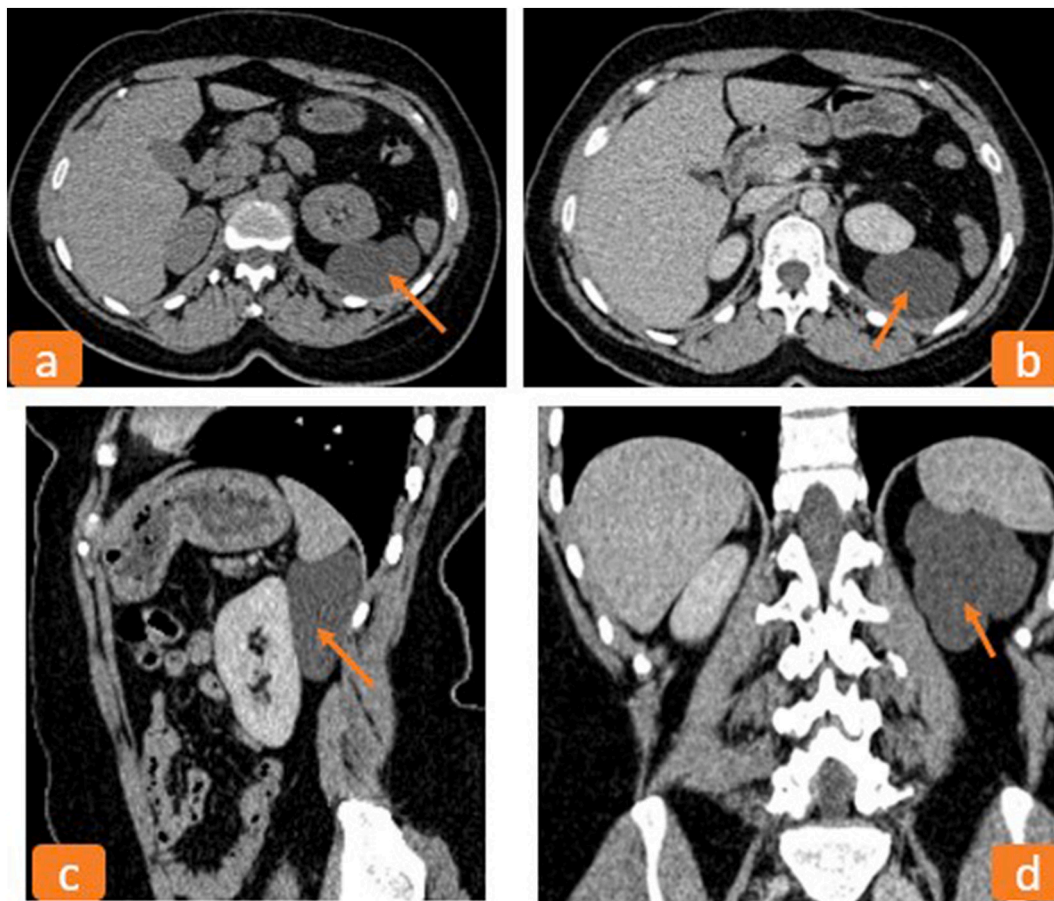
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origin. It is secondary to abnormal proliferation followed by sequestration of lymphatic vessels during embryonic life. It is therefore a congenital malformation in which the pathological or sequestered vessels fail to establish normal communication with the rest of the lymphatic system. This results in a failure of lymphatic drainage and an accumulation of fluid, leading to formation of uni or multilocular cysts [2,3]. Most of cystic lymphangioma are cervical or axillary in location. Abdominal localization is rare and it represents only 7 % of cystic masses of the abdomen [4]. Retroperitoneal location is exceptional [5]. The diagnosis of cystic lymphangioma is most often made in childhood, but slow-growing lymphangiomas such as those of retroperitoneal topography are often discovered in adulthood. There is no gender predilection [6]. In perirenal cystic lymphangioma, the cystic mass develops in the perirenal space. It is either discovered by incidental finding during an imaging examination or following the occurrence of lumbar pain as in our patient. It can also be manifested by hypertension, hematuria or proteinuria. The clinical examination may be normal in adults or may show flank tenderness. In children, nephromegaly is more frequently found. Ultrasound and CT scans are used to approach the diagnosis. They reveal a cystic formation with a thin and regular wall without a solid component, uni-loculated or multi-loculated with thin septa, most often regular [7]. Parietal calcifications are sometimes present, and the appearance of the cyst contents varies according to the purely liquid nature, or the presence of intracystic hemorrhage or infection. In case of complications or atypical presentation on ultrasound and CT, MRI provide an additional interest [3]. It confirms the cystic nature of the mass in hyposignal in T1 weighted and hypersignal in T2 weighted; allows identification of possible hemorrhagic areas, and especially on enhanced

sequences, there is a weak enhancement of the wall and septa and an absence of enhanced tissular portion in the intracystic [8]. It confirms the cystic nature of the mass with T1-weighted hyposignal and T2-weighted hypersignal; it allows identification of possible hemorrhagic areas, and especially on enhanced sequences, there is weak enhancement of the wall and septa and an absence of enhancement of the tissue portion in the intracystic [7]. However, many conditions may have similar features. The differential diagnosis, especially with benign or malignant tumors of the kidney, renal lymphoma, polycystic kidney disease, urinoma or renal abscess, is not always easy. Therefore, the diagnostic approach must be based on clinical, biological and imaging data and in ambiguous forms, only histology can provide a definite diagnosis. Thus, the presence of an infectious syndrome points to an abscess or superinfection of the cyst, and hematuria or intracystic enhancement should suggest a tumoral cause [7]. Complications found in pericystic lymphangiomas are intracystic hemorrhage, infection of cyst, compression of the renal hilum or rupture of the cyst. The choice of treatment depends on several factors. Asymptomatic cystic lymphangiomas do not require specific treatment and should be followed up on imaging. Percutaneous treatment consists of aspiration of the cystic lymphangioma followed or not by injection of a sclerosing agent. It is a minimally invasive technique with however a high recurrence rate. It is reserved for symptomatic cystic lymphangioma with a typical presentation on imaging. In case of diagnostic doubt, especially with a renal tumor, surgical treatment is the therapy of choice and the excision must be complete in order to reduce the risk of recurrence while preserving the surrounding structures [4].



**Fig. 1.** CT urogram in a 50-year-old man with lumbar pain in unenhanced axial section (a); in enhanced axial section (b) and sagittal (c) and coronal (d) reconstruction, showing a well-limited, lobulated, fluid-dense mass (arrow) with a thin wall slightly enhanced by iodinated contrast, developing in the left perirenal space in contact with the posterior-superior part of the kidney suggestive a retroperitoneal cystic lymphangioma.

#### 4. Conclusion

Cystic lymphangioma is a benign tumor with a varied clinical and radiological presentation. Perirenal involvement is very rare and can lead to complications. The differential diagnosis with a benign or malignant renal tumor can sometimes be laborious and therefore requires histological evidence to make a definite diagnosis.

#### Declaration of competing interest

The authors declare that they have no links of interest.

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