Renal lymphangiectasia: know it in order to diagnose it

Dear Editor,

Here, we report the case of a 9-year-old girl with hyperparathyroidism. Ultrasound showed renal cysts and increased echogenicity of the parenchyma in both kidneys. The diagnostic hypothesis was hyperparathyroidism secondary to chronic/polycystic kidney disease. The patient presented with gradually worsening kidney function and hypertension, and new imaging scans were requested. The ultrasound showed anechoic, multiloculated images in the pyelocaliceal region of both kidneys, and perirenal, subcapsular cysts. A computed tomography (CT) scan was acquired, although no contrast agent was used, which precluded an accurate characterization. Nevertheless, the CT scan revealed changes similar to those observed on ultrasound. We also performed magnetic resonance imaging (MRI), which showed pyelocaliceal, perirenal cysts, with altered intensity of the signal of the renal parenchyma and loss of corticomedullary differentiation (Figure 1A), confirming, in conjunction with the clinical and biochemical data, the diagnosis of renal lymphangiectasia (RL).

RL is a rare benign disease that occurs because of miscommunication between the renal lymphatic drainage system and the retroperitoneal lymphatic system[1]. As a result, there is accumulation of lymph in the renal lymph ducts, making them ectatic and forming simple or multiloculated, typically asymmetric and bilateral, collections in the pyelocaliceal, perinephric, or parenchymal regions, although, in some cases, only a part of one kidney is affected (Figure 1—B,C). There is no predilection for a given gender or age group. As of 2005, only 40 cases had been described[1,2].

In most cases, RL is an incidental finding, with or without signs and symptoms of pain, increased abdominal volume, hematuria, ascites, edema of the lower limbs, hypertension, erythrocytosis with renal vein thrombosis, and, rarely, chyluria[3]. Such manifestations can be explained by the distention of the renal fascia and compression of the renal parenchyma by cysts, fistulization to the pelvic cavity, and changes in the renin-angiotensin system[2-4]. In rare cases, chronic kidney disease has been reported[5]. To our knowledge, there have been no specific reports of clinical evolution to hyperparathyroidism, although a relationship with chronic kidney disease can be assumed.

A CT scan can reveal expansive perirenal formations with fluid attenuation, bounded by the renal fascia, that conform to

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http://dx.doi.org/10.1590/0100-3984.2015.0190

Figure 1. A: Coronal T2-weighted MR sequence showing a loss of corticomedullary differentiation in both kidneys and multiple cystic lesions, with thin walls, located in the cortex (arrows). B: Cystic formations in the subcapsular cortex (arrows). C: Axial T2-weighted MRI sequence showing cysts located in the renal sinususes (arrowheads) and perinephric spaces, simulating pelvic dilatation. D: The same images simulating cystic collections in the subcapsular cortex (arrow).
Primary undifferentiated sarcoma in the thorax: a rare diagnosis in young patients

Dear Editor,

A 30-year-old man was admitted to the thoracic surgery department of a tertiary hospital for investigation of a thoracic mass. Having previously received treatment for pneumonia, he presented with a two-week history of progressively increasing pain in the right hemithorax and right flank, between the anterior axillary line and midaxillary line. On clinical examination, there was an absence of breath sounds in the right hemithorax.

Computed tomography (CT) of the chest showed an extensive, heterogeneous, mostly solid mass in right thoracic region (Figure 1), with areas of inner content of low attenuation (21–26 Hounsfield units) and foci of bleeding, without intervening calcifications and without osteolysis of the rib. Laboratory tests produced results within the limits of normality. The patient underwent percutaneous biopsy, and the pathology examination revealed undifferentiated sarcoma (Figure 2).

Sarcomas represent a heterogeneous group of tumors derived from mesenchymal cells (1–3). They account for 1% of all neoplasms and occur mainly in the extremities (in 60% of cases), gastrointestinal tract (in 25%), retroperitoneal space (in 20%), and the head and neck region (in 4.1%). Primary sarcomas of the thorax are exceptionally rare, accounting for only 0.2% of lung cancers and only 5% of all the thoracic neoplasms. Such sarcomas can involve the lungs, mediastinum, pleura, and, mainly, the chest wall. The presence of sarcoma in any other part of the body must be ruled out, because metastasis to the chest is much more common than is primary sarcoma of the thorax (4–7).

The most common histological types of primary sarcomas are angiomysarcoma, leiomyosarcoma, rhabdomyosarcoma, and sarcomatoid mesothelioma (8). In the chest wall, the most common primary sarcomas are Ewing’s sarcoma, primitive neuroectodermal tumor, malignant fibrous histiocytoma, chondrosarcoma, osteosarcoma, synovial sarcoma, and fibrosarcoma (9). Radiologically, these tumors typically present as large, heterogeneous masses. However, their appearance can vary from an intrabronchial

Figure 1. CT scan showing a primary sarcoma in the right hemithorax. A: CT scout image showing opacification of the right hemithorax. B: Coronal CT reconstruction with heterogeneous enhancement (arrow). C: Axial CT slice showing contralateral mediastinal deviation.

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http://dx.doi.org/10.1590/0100-3984.2015.0025