UNCOMMON RENAL MASSES: PERIRENAL EXTRAMEDULLARY HEMATOPOIESIS AND MULTIPLE LYMPHANGIOMATOSIS WITH A PERIRENAL LYMPHANGIOMA


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Summary.- OBJECTIVE: To present two cases of infrequent renal masses, trying to achieve the diagnosis before surgery. METHODS: We describe a case referred from the Department of Hematology in which bilateral perirenal masses were described in the CT scan; after biopsy they where classified as extramedullary hematopoietic tissue. The other case was a patient presenting to the emergency room with dyspnea. CT Scan showed lungs with multiple cysts, chylothorax and a cystic-solid mass in the left perirenal space. In the lung biopsy they reported lung lymphangiomatosis, so we didn’t perform renal biopsy. RESULTS: Most renal masses are renal carcinomas (85%). The less common diagnosis are sarcomas, lymphomas, upper urinary tract transitional cell carcinomas, metastases of other primary tumours, the Erdheim-Chester disease, the Castleman disease and benign tumours. All these diseases might show similar images in the CT scan and MRI, being the biopsy and histological study necessary for the diagnosis. CONCLUSIONS: Perirenal extramedullary hematopoiesis and perirenal lymphangiomatosis are rare diseases that need a pathologic study for their diagnosis.

Keywords: Perirenal tumors. Extramedullary hematopoiesis. Disseminated lymphangiomatosis. Perirenal lymphangioma.

Resumen.- OBJETIVO: Presentar dos casos de masas renales infrecuentes, intentando llegar a su diagnóstico preoperatoriamente. MÉTODOS: Describimos un caso remitido por Hematología por hallazgos en TAC de masas perirrenales bilaterales, que al biopsiar referían hematopoyesis extramedular. El otro caso se estudió por disnea, apreciando en el TAC pulmones con múltiples quistes y en abdomen, masa quístico sólida perirenal izquierda. La biopsia pulmonar nos informó de Linfangiomatosis pulmonar, con lo que obviábamos la biopsia renal. RESULTADOS: La mayoría de las masas renales sólidas son hipernefromas (85%). El resto de las masas son sarcomas, linfomas, tumores de vías infiltrantes y tumores benignos. Para su diagnóstico disponemos de la clínica y las pruebas radiológicas (ecografía, TAC, RNM y PET-TAC), pero ante hallazgos inespecíficos el diagnóstico se basará en el estudio histológico. CONCLUSIONES: La hematopoyesis renal extramedular y el Linfangioma perirrenal son tumores raros y su diagnóstico preoperatorio es difícil.


INTRODUCTION

We discuss two cases of perirenal masses to review the literature. The first one is a case of perirenal extramedullary hematopoiesis in a patient with myelofibrosis due to polycythemia vera, that was referred to the urology consultation for perirenal masses. Also we report a second case of a perirenal lymphangioma...
mass in both perirenal spaces (Figure 2), and an increase of the density of the soft tissue in the presacral zone. The posterior mediastinum was occupied at the prevertebral level with a pathological soft tissue. The MRI describes a generalized darkness of the bone marrow, in T1 and also in T2, suggestive of soft tissue mass. We observe the same tissue at the perirenal space. In the presence of bilateral perirenal tumours, we decide to perform a fine-needle biopsy to get the diagnosis, generating a right retroperitoneal hemorrhage, which did not require aggressive treatment. The result of the biopsy reported: fine capillaries vessels, watching habit groups in all its forms erythroid maturation and not atypical megakaryocytes and granulocytic cells (Myeloperoxidase-). Blasts were not observed, infiltration of epithelial cells (cytokeratin-) or lymphoid proliferation phenomena. Diagnosis: extramedullary hematopoiesis.

Due to the pain and the paralysis of left leg, radiation treatment was decided on the intervertebral spaces of L4-L5, improving symptoms.

The patient was treated by the hematologist with hydroxyurea and allopurinol and died from multiple organ failure a few months after, diagnosed by a bone marrow biopsy of a last phase of acute leukemia.

**CASE REPORT 1**

The first case is a male of 75 years old, with history of myelofibrosis due to polycythemia vera, and a splenectomy performed 3 years ago caused by a massive painful splenomegaly. The patient arrives at the ER with pain and a paralysis of the left inferior limb.

Blood analysis: Thrombocytosis, at first normal leucocytes but after a leucocytosis of 58,000.

In the CT we observe an asymmetry among the intervertebral foramen on the L4-L5 spaces, with occupation of the right foramen (Figure 1), a soft tissue mass in both perirenal spaces (Figure 2), and an increase of the density of the soft tissue in the presacral zone. The posterior mediastinum was occupied at the prevertebral level with a pathological soft tissue. The MRI describes a generalized darkness of the bone marrow, in T1 and also in T2, suggestive of soft tissue mass. We observe the same tissue at the perirenal space. In the presence of bilateral perirenal tumours, we decide to perform a fine-needle biopsy to get the diagnosis, generating a right retroperitoneal hemorrhage, which did not require aggressive treatment. The result of the biopsy reported: fine capillaries vessels, watching habit groups in all its forms erythroid maturation and not atypical megakaryocytes and granulocytic cells (Myeloperoxidase-). Blasts were not observed, infiltration of epithelial cells (cytokeratin-) or lymphoid proliferation phenomena. Diagnosis: extramedullary hematopoiesis.

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**CASE REPORT 2**

The second case is a 35 yo woman with a history of dyslipidemia, irritable bowel and a surgery of a ruptured anterior cruciate ligament and meniscus in his right knee in 2005.

She arrived to the ER with dyspnea and left chest pain. Physical Examination: silent air in the left lung.

Rx: left pleural effusion diagnosis by thoracentesis of chylothorax. After, the Pneumologist requested a thoracic CT scan, confirming the existence of an almost massive left pleural effusion and the presence of small, diffuse, pulmonary cysts in both hemithorax (Figure 3).

Suspecting the possibility of lymphangioleiomyomatosis, the Thoracic Surgery Service performed VATS proving the presence of left chylothorax. Lung and pleural biopsy was performed, describing dilated pulmonary parenchyma and proliferation of lymphatic vessels surrounded by lymphocyte cells, vascular congestion. With special techniques D2 40 we’re able to confirm the proliferation of lymphatic vessels surrounded by muscle fibers positive for muscle-specific actin.

AP Diagnosis: Diffuse pulmonary Lymphangiomatosis.

Afterwards they decided to place a pleuro-peritoneal valve (Denver), pending on whether the patient was a subsidiary of lung transplantation, according to evolution of the disease.

Two and half years after the chylothorax bypass, the patient is stable, O2 saturation 98% and with a normal spirometry. A small pleural effusion persists.

The abdominal CT scan revealed a left perirenal lesion 5 x 3 x 5 cm, hypervascular arterial phase; in the late phase the contrast is quickly washed away, the lesion contains low density internal zones that aren’t enhanced on the image. The mass is infiltrating the renal capsule. The abdominal MRI, shows a left perirenal vascularized mass. The mass is hypointense on T1 and hyperintense on T2 (Figure 4 A-B).

In the context of a diffuse pulmonary lymphangiomatosis, we diagnosed the renal mass as a perirenal lymphangioma, deciding for a non-aggressive treatment.

The patient is asymptomatic and in 2 years of evolution, the renal mass remains the same size.

DISCUSSION

Extramedullary hematopoiesis (EH) occurs when the hematopoietic tissue develops outside of the bone marrow, for reasons such as congenital hemoglobinopathies, leukemias, lymphomas, primary or secondary myelofibrosis or bone metastasis (1).

In our case it was for myelofibrosis due to polycythemia vera, that developed multiple sites of extramedullary hematopoiesis, on the intervertebral space (L4, L5 and S1), the pre-sacral zone, the perirenal space and in the mediastinum.
In case of myelofibrosis, the most frequent locations of EH are: the liver, the spleen, thoracic paraspinal regions; and the less common are: the heart, the lungs, the lymph nodes, mediastinum, retroperitoneal space and kidneys (2).

EH perirenal involvement is very rare and is manifested by two distinct patterns: diffuse infiltrative process surrounding the kidneys or soft tissue masses mixed with fatty tissue.

Splenectomy facilitates the development of the EH in unusual organs, as in our case.

In the CT, the EH appears as a hypovascular soft tissue mass, with or without fat, as in our case.

In the MRI, (on T2) given that they are soft tissue masses, the images appear with low signal intensity because of the hemosiderin content (5).

Since there are bilateral renal masses in the clinical setting of EH, we think in this etiology, but the final diagnosis is established by a fine needle biopsy.

The prognosis of this disease is quite bad. In the presence of bilateral renal masses, we should perform a differential diagnosis with lymphoma (3), leukemias, plamatic cell tumours, renal metastases from other primary tumours (melanoma, prostate, lung, and breast), a bilateral sarcoid tumour (4), the Erdheim-Chester disease, a lipoid granulomatosis that can affect the perirenal space, Castleman’s disease is an idiopathic lymphoproliferative disorder with a rare affection of the perirenal space in few cases, make a differential diagnosis with and we must also think in a tuberous sclerosis in the presence of a bilateral renal angiomyolipomas.

All this bilateral perirenal tumours have a very low incidence, and make it difficult to diagnose between them.

In the CT scan and the MRI, we can see similar images in all this pathologies, the are solid hipovascular masses, because of that we have pay attention to the symptoms, and the final diagnosis will be with a biopsy. Then we can offer a specific treatment for this diseseases.

The renal and perirenal lymphangioma is a benign tumour, generated by an obstruction of the lymphatic vessels that drain the kidney. There are more frequent in the childhood, mostly seeing in armpit and neck. However we can find it also in the mediastinum, lungs, kidneys and bones. The frequency of the lymphangiomas in all the tumours are less than 0.05% (10). Primary retroperitoneal tumours are rare, 80% of those are malignant. Lymphangiomas are very rare, accounting for 1% retroperitoneal masses (10).

The renal and perirenal lymphangioma may be unilateral or bilateral, they may be presented as unilocular or multilocular cysts, focal or diffuse, and appear in images with or without reinforcement or enhancement of their septum and periphery (2,8).

The Lymphangiomatosis is characterized by multiple lymphangiomas in various organs, which are locally infiltrative. These lymphangiomas can be subdivided into simple, cavernous and cystic, this last type originates mainly in the retroperitoneum.

Multiple organ involvement is common, obviating the need of biopsy.

Thoracic involvement occurs in adults, causing pleural thickening, chylothorax, and bones present lytic lesions with sclerotic margins caused by lymphatic vessels. Clinically may be asymptomatic as it was in our case, but if they are big enough, they can cause hypertension, hematuria, proteinuria or intracystic hemorrhage. Renal lymphangiomas are exacerbated during pregnancy, acquiring a larger size and may cause ascites (6).

The diagnosis was based on the symptoms, in the radiological exams we found: on the ultrasound: renal or perirenal lesion with uni or multilocular cysts with septa; in the CT scan: lymphangiomas appear as big cystic masses with thin walls, which do not capture contrast (is most typical), but can also occur with enhancement or peripheral enhancement or septa (7).

In the MRI, renal or perirenal lymphangiomas appear hypointense on T1 images and hyperintense on T2, with a well-defined wall. Typically, there is an immediate peripheral enhancement after the contrast injection, and delayed reinforcement in the center, caused by a slow lymphatic flow within the cystic mass. But these images can be altered by the presence of chyle existing high signal intensity on T1 and intermediate in T2 (8).

The differential diagnosis must be performed with the cystic tumour masses, both benign and malignant, so polycystic kidneys will replace the entire renal parenchyma with cysts; the multilocular cystic nephroma affects the kidney, the renal hydatid disease (you can see in the CT scan baby cysts), and the cystic leiomyosarcoma, appreciate predominantly solid lesion and fibrosis.

We should differentiate it from non-tumoural cystic masses like: renal or perirenal abscesses (these are collections with infectious symptoms), kidney haematoma (history of traumatisim, anticoagulant therapy or after surgical procedures), and urinomas (extravasated urine by trauma, renal colic or iatrogenic maneuvers).

Regarding treatment, as it’s a benign tumour you should try not to be agressive (9), a few techniques have been described for treatment, such as, marsupialisation, percutaneous drainage with good results. (6,7) or sclerotherapy.

The problem is when the diagnosis isn’t suspected prior
to surgery, and it’s the nephrectomy what will solve our diagnostic doubts; the exeresis of the cysts and the masses is not always possible (when it’s an infiltrative lesion).

CT and MRI findings are useful to suspect of a lymphangioma. Puncture of the mass does not always show us chyle, and it’s not diagnostic.

In our case with by diagnosing a lung lymphangiomatosis, we didn’t proceed with an aggressive treatment and we performed periodical controls of these perirenal masses

**CONCLUSIONS**

The bilateral perirenal hematopoiesis is very rare. The symptoms and the diagnostic tests, such as CT and MRI can guide us through the diagnosis. Definitive diagnosis is made by biopsy and we think it’s a disease to keep in mind when bilateral renal masses are present.

The perirenal lymphangioma is a very rare tumour. In the CT and MRI, is typical to observe a big cystic mass with thin walls, always keeping trying to keep this in mind [5]; but in some cases, the cysts might be highlighted by the contrast, and along with the presence of chyle in the cysts, the diagnose will be more difficult, leading the surgeon to perform a nephrectomy.

In the context of a diffuse lymphangiomatosis, the diagnosis is easier and we must opt for a non aggressive treatment.

**REFERENCES AND RECOMMENDED READINGS**

(*of special interest, **of outstanding interest)