|| ISSN(online): 2589-8698 || ISSN(print): 2589-868X ||
International Journal of Medical and Biomedical Studies
Available Online at www.ijmbs.info
NLM (National Library of Medicine ID: 101738825)
Index Copernicus Value 2019: 79.34
Case Report Volume 5, Issue 5; May: 2021; Page No. 12-15

RENAL HEMANGIOPERICYTOMA: A POSSIBLE CAUSE OF ACUTE ABDOMEN

Victor Bastos Frade [1], José Genilson Alves Ribeiro [1], Rodrigo Barros [1], Maria Clara Machado Breves [1], Bruno de Souza Bianchi Reis [2], Márcio Babinski [3], Angelo Maurilio Fosse Junior [1]

[1] *Urology, Fluminense Federal University, Niterói, Rio de Janeiro, Brazil*
[2] *Pathology, Fluminense Federal University, Niterói, Rio de Janeiro, Brazil*
[3] *Morphology, Fluminense Federal University, Niterói, Rio de Janeiro, Brazil*

Article Info: Received 28 March 2021; Accepted 09 May 2021
DOI: https://doi.org/10.32553/ijmbs.v5i5.1884
Corresponding author: Rodrigo Barros
Conflict of interest: No conflict of interest.

Abstract

Introduction: Hemangiopericytoma (HPC) is a type of mesenchymal fibroblast tumor with anomalous perivascular presentation and malignant potential. We report a case of renal HPC presented with acute abdomen.

Case presentation: A 59-year-old male patient presented fever and refractory abdominal pain in the right hypochondrium region. The abdominal computed tomography (CT) showed solid mass in the anteroinferior region of the right kidney, with slightly lobulated contour, heterogeneous enhancement after contrast, with hypodense areas compatible with necrosis, measuring 14.2 x 12.6 x 10 cm, compressing and displacing the inferior vena cava and ascending colon. The patient underwent right laparoscopic radical nephrectomy, without complication. The anatomopathological study showed mesenchymal neoplasia with moderate mitotic index, with foci of necrosis and moderate cellular pleomorphism. The immunohistochemical study confirming the diagnosis of HPC.

Conclusion: Renal HPC is an extremely rare neoplasm. Clinical manifestations are variable, being a possible cause of acute abdomen.

Key words: renal cancer, hemangiopericytoma, acute abdomen

Introduction

Hemangiopericytoma (HPC) or solitary fibrous tumor (SFT) is a type of mesenchymal fibroblast tumor with anomalous perivascular presentation and malignant potential. The first description of STF occurred in 1931 and referred to a pleural tumor (1). In 1942 the first case report of HPC describing a perivascular muscle cell-related tumor called pericytes was published (2). The phenotypic, behavioral and genetic pattern of HPC was the same as that of SFT, so these are currently grouped as the same neoplasm according to the World Health Organization (WHO) classification of soft tissue tumors (3). This tumor can occur in any organ, since the pericytes are widely distributed throughout the body, although more frequently found in the pleura, retroperitoneum, extremities, pelvis and meninges. Renal HPC is especially uncommon and very difficult to differentiate from other common renal neoplasms.

Case Report

A previously healthy white 59-year-old male patient presented fever and abdominal pain in the right hypochondrium region. Due to the refractoriness of pain to the use of analgesics and non-steroidal anti-inflammatory drugs even after fever resolution, he came to our emergency department with worsening abdominal pain associated with palpable mass in the right hypochondrium. Laboratory tests revealed anemia with leukocytes and normal kidney function. The abdominal computed tomography (CT) showed solid mass in the anteroinferior region of the right kidney, with slightly lobulated contour, heterogeneous enhancement after contrast, with hypodense areas compatible with necrosis, measuring 14.2 x 12.6 x 10 cm, compressing and displacing the inferior vena cava and ascending colon. Perirenal and peri-lesional fat infiltration was seen, with no signs of invasion of the renal vessels, lymphadenopathy or metastasis (Figures 1 and 2).

The patient underwent right laparoscopic radical nephrectomy, without complication, need for blood transfusion or intensive postoperative care. The evolution was favorable, but the patient was discharged on the sixth postoperative day due to ileum. The anatomopathological study showed mesenchymal neoplasia with moderate mitotic index (4 mitoses in 10 fields), with foci of necrosis and moderate cellular pleomorphism. No vascular invasion was identified (Figure 3). The immunohistochemical profile was positive for CD34, CD99, Ki-67, Bcl-2 and STAT-6 and negative for RE, Wt-1, CD10, CKAP, EMA, AE1 /

AE3, CK8 / 18, AML, HMB45 and melan-A (Figures 4). The patient remains in outpatient follow-up every six months with oncology and urology analysis, without metastatic foci or any other clinical-radiological manifestation.

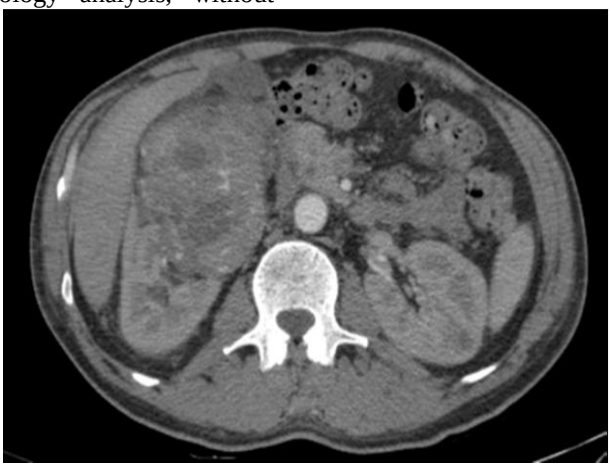

Figure 1: Contrast tomography showing solid mass in the inferior antero region of the right kidney with heterogeneous enhancement and hypodense areas compatible with necrosis.

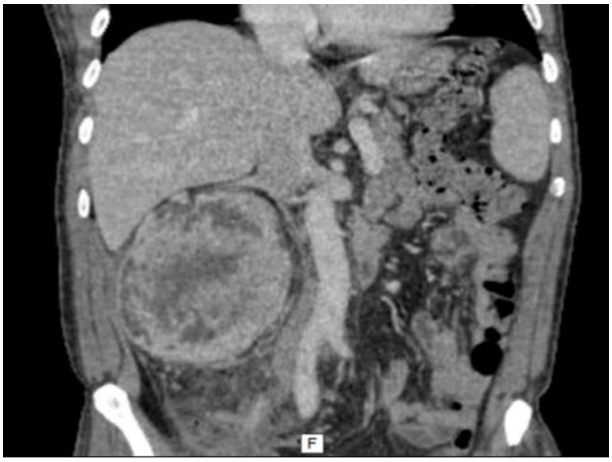

Figure 2: Coronal section contrast-enhanced tomography showing heterogeneous mass of 14.2x12.6x10 cm in the right kidney, demonstrating displacing of the inferior vena cava.

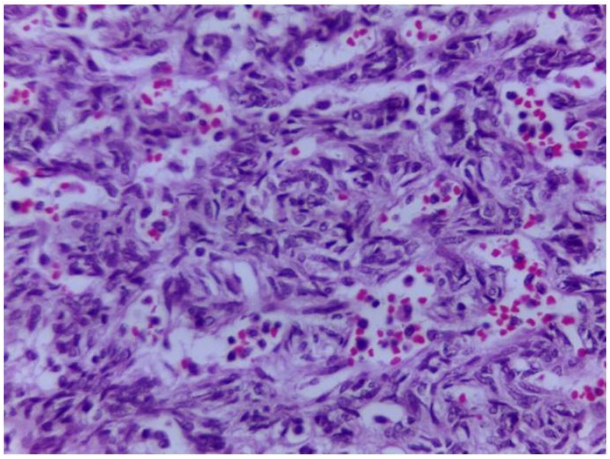

Figure 3: Neoplasia composed of spindle cells, of medium size, forming anastomosed vessels, in "hemangiopericytoma" pattern. (400X)

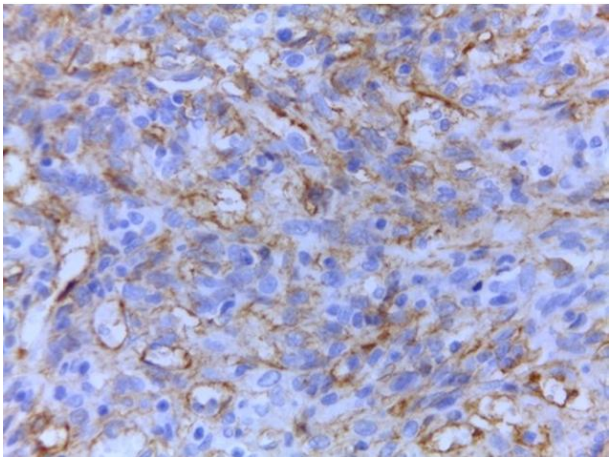

Figure 4: Immunohistochemical analysis with anti-CD34 positive marker in tumor cell cytoplasm.

Discussion

HPC is a neoplasm characterized by uncontrolled proliferation of pericytes, modified smooth muscle cells that are located on the outer surface of capillaries and venules. These cells regulate blood flow and vessel permeability (2). This tumor is mainly found in the thoracic cavity and pelvis, and only rarely affects the kidney. There is no significant difference in involvement between sexes, and although it may affect patients of any age, the main victims are older adults, with rare descriptions of the involvement of children and young adults. Clinical presentation is variable, with common complaints such as abdominal mass, hematuria, abdominal pain on the flanks or lower back. Moreover, most of these patients are asymptomatic (5). The patient described here reported progressive and acute abdominal pain, probably due to the large proportions of the tumor, without other manifestations.

The tumor is most often located in the renal hilum and can be visualized by imaging tests such as ultrasonography (US), which shows nonspecific tumor imaging, as well as CT and magnetic resonance imaging (MRI), which provide greater detail (4). Through CT, a well-defined, slightly hyperdense lesion is usually observed on non-contrast examination and enhanced after contrast, with a homogeneous or heterogeneous pattern of varying in size surrounded by a pseudocapsule, with a large network of vessels and sometimes small satellite nodules (3). MRI may show a hyperintense signal at T1 and hypo or isointense at T2, with the hypointense lesions being more related to hypercellularity and abundant collagenous stroma, while hyperintense lesions suggest necrotic, myxoid or cystic alterations (3). In the present case, we used CT, which showed a tumor close to the hilum, as in the majority of the cases described in the literature, with size of 14.2 x 12.6 x 10 cm.

Macroscopic histological evaluation usually shows a solid, firm, well-circumscribed tumor with gray, white, or yellowish-brown surface, suggesting partial bleeding or necrosis. The most accurate analysis is made from the microscopic characteristics using techniques such as hematoxylin-eosin and silver staining to facilitate visualization. In microscopy, the predominant pattern is irregular cells in the form of spindle or vortex beams, with some areas of alternation between dense and sparse cells interspersed with collagen fibers and others rich in visible hemangiopericytoma blood vessels. Cellular atypia is usually mild and clear bleeding and necrosis are more difficult to see (3). In immunohistochemical analysis, antibodies against CD31, CD34, CD68, CD-99, S-100, vimentin, cytokeratins and epithelial membrane antigen are used. In this neoplastic type, it is expected to find positivity for CD99, vimentin, Bcl-2, CD34 and negativity for CD31, keratin, actin and S100 (6). However, CD34 historically stands out as the main antibody for STF diagnosis, while the expression of NAB2-STAT6 has also been shown to be a surrogate marker, with good sensitivity and specificity, so it can be used as a prognostic factor. Markers such as PAX8, PAX2, SMA, GRIA2 are also described in the literature, as well as BCOR expression, which could be related to a worse prognosis and risk of malignancy (3,7). The tumor described in this report tested positive for CD34, CD99, Ki-67, Bcl-2 and STAT6.

Treatment consists of tumor removal and radical surgery is recommended to avoid incomplete resection and recurrence. There is little evidence to demonstrate the efficacy of other therapeutic modalities such as radiotherapy, chemotherapy and immunotherapy (8,9). The main site of metastasis is the lung, followed by bones, liver, lymph nodes, pancreas, peritoneum and the renal vein (8,9,10). In addition, there are reports of local malignancy and contralateral kidney recurrence over a period of 8 to 20 years after the first treatment, highlighting the importance of long-term follow-up of these patients (9).

Most reports include tumor-related benignity with local and metastatic recurrence rate of about 12 to 16% of cases, so these tumors are classified as having intermediate biological potential and low risk of metastasis according to the 2002 WHO classification (3,11). However, there are

reports of aggressive behavior, even in histologically benign lesions (12). Our patient underwent radical surgery two years ago and is being followed at the same health institution, with no sign of recurrence or metastatic foci so far. Follow-up is important to detect any metastatic manifestation, as it is possible for lesions to arise several years after the initial therapeutic approach.

Conclusions

Renal HPC is a rare neoplasm and presents variable clinical manifestations. However, is a possible cause of acute abdomen. There are nonspecific findings of imaging tests, and the diagnosis is usually made postoperatively with the combination of histological and immunochemical analysis. The treatment advocated so far is radical surgery, while other therapies such as radiotherapy and chemotherapy are being tested, but with effectiveness yet to be confirmed. Long-term follow up is mandatory and patients should be informed about prognosis and the possibility of recurrence and tumor metastasis.

References

1. Klemperer P, Coleman BR. Primary neoplasms of the pleura. A report of five cases. Am J Ind Med. 1992;22(1):1-31.
2. Stout AP, Murray MR. Hemangiopericytoma: A vascular tumor featuring Zimmermann's pericytes. Ann Surg. 1942 Jul;116(1):26-33.
3. Fu W, Bing Q, JingJing Z, Lijian L, Zhibin X, Qijian D, Jiwen C. Clinicopathological characteristics of renal solitary fibrous tumor: A single institution experience. Medicine (Baltimore). 2019 Feb;98(8):e14618.
4. Yi X, Wang J, Zhang Y, Wang Z, Zhang Z, Gong G, Liu L, Xiang W, Liao W, Zee C, Chen BT. Renal solitary fibrous tumor/hemangiopericytoma: computed tomography findings and clinicopathologic features. Abdom Radiol (NY). 2019 Feb;44(2):642-651.
5. Ribeiro J. G. A., Klojda C. A. B., Machado F. A., Favorito L. A., Cavalcante A. G. L., Babinski M. A. Unilateral Renal Haemangiopericytoma: Report of a Rare Tumour. Int. J. Morphol. 2006 June; 24(2): 165-170.
6. Rodríguez Cruz MI, Hernández Sánchez JE, Blázquez BS, Prieto Nogal SB, Gómez Tejeda LM. Malignant solitary fibrous kidney tumor with peritoneal disease: a case report. Case Rep Nephrol Urol. 2014 Apr 11;4(1):70-4.
7. Argani P, Kao Y. C, Zhang L, Sung Y.S, Alaggio R, Swanson D, Matoso A, Antonescu C.R. BCOR Overexpression in Renal Malignant Solitary Fibrous Tumors: A Close Mimic of Clear Cell Sarcoma of Kidney. The American Journal of Surgical Pathology, 2019 mar; 43 (6) , pp. 773-782.
8. Cuello, J.; Brugés, R. Malignant solitary fibrous tumor of the kidney: report of the first case managed with interferon. Case Rep Oncol Med. 2013; 2013: 564980.
9. Sammoud S, Ferjani S, Hamdani M, Toumi A. Malignant Renal Solitary Fibrous Tumor With Two Local Recurrences and Distant Pulmonary Metastasis. Urology. 2019 May;127:9-12.
10. Sasaki H, Kurihara T, Katsuoka Y, Nakano T, Yoshioka M, Miyano S, Sato Y, Uejima I, Hoshikawa M, Takagi M, Chikaraishi T. Distant metastasis from benign solitary fibrous tumor of the kidney. Case Rep Nephrol Urol 2013;3:1–8.
11. Gold JS, Antonescu CR, Hajdu C, Ferrone CR, Hussain M, Lewis JJ, Brennan MF, Coit DG. Clinicopathologic correlates of solitary fibrous tumors. Cancer. 2002 Feb 15;94(4):1057-68
12. Bezerra ES, Andrighetto OP, Costa MVS, Chiarelli LO, Moleiro W, Schultz L. Bilateral renal involvement by solitary fibrous tumor - Report of a case in the post-WHO/2016 era. Urol Case Rep. 2017 Dec 9;17:7-9.