Unilateral Renal Haemangiopericytoma: Report of a Rare Tumour

Hemangiopericitoma Renal Unilateral : Reporte de un Raro Tumor

*J. G. A. Ribeiro; *C. A. B. Klojda ; *F. A. Machado; *L. A. Favorito; *A. G. L. Cavalcante & *,**M. A. Babinski

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.*, 24(2):165-170, 2006.

SUMMARY: The pericytes are modified smooth muscle cells (SMCs) that surrounding capillaries and to change the diameter of vascular lumen. In this article we report a case of renal haemangiopericytoma. A 78-year-old woman was admitted with a complaint of abdominal pain. Ultrasonography revealed a solid injury in the third portion of the left kidney. Abdominal CT scan revealed a tumoral mass. Due the high index of suspicion of malignant neoplasia the patient was submitted to left radical nephrectomy. The histopathological evaluation confirmed haemangiopericytoma by immunohistochemistry. The patient fared well and was alive 5 years after radical nephrectomy.

KEY WORDS: Kidney; Tumour; Haemangiopericytoma; Pathology; Immunohistochemistry.

INTRODUCTION

Haemangiopericytoma is a rare, malignant neoplasm presumably arising from pericytes, the smooth muscle cells that are external to the walls of capillaries and arterioles (Rubin & Farber, 1988; Argyropoulos *et al.*, 2005). These tumors are small and consist of capillary-like channels surrounded by (and frequently enclosed within) nests and masses of round to spindle-shaped cells. The tumor cell type is identified by a characteristic investment of basement membrane, similar to that of its normal counterpart (Rubin & Farber; Fornaro *et al.*, 1999).

Regarding the anatomical site, this tumour may occur anywhere but are most frequently encountered in the retroperitoneum (McCormack & Gallivan, 1954; Black & Heinemann, 1955) and lower extremities. They are highly malignant and typically metastasize to lungs, bone, liver, and lymph nodes (Rubin & Farber).

In a wide spectrum of renal tumours, haemangiopericytoma is one of the scarcer entities (Rubin & Farber and Argyropoulos *et al.*). Because these tumours are rare, questions remain about the preoperative diagnosis, the ideal therapeutic strategies and the prognosis of the disease. This tumour is generally rare in the urogenital

system, and except for the kidney, other sites include the bladder, the prostate and the spermatic cord (Argyropoulos *et al.*; Baumgartner *et al.*, 1976; Chen, 1987; Siemens *et al.*, 1998).

Since the first report of renal haemangiopericytoma by Black & Heinemann only 25 cases of this rare vascular tumour have been described in the literature. The term haemangiopericytoma was for the first time used by Stout & Murray (1942) to designate a peculiar type of vascular tumour involving capillary pericytes. Zimmermann (1923), pointed out that pericytes are modified SMCs that surround capillaries and modify the diameter of vascular lumen. Therefore, we report a case of a patient with left renal haemangiopericytoma submitted to radical nephrectomy.

REPORT OF CASE

A 78-year-old woman was admitted with a complaint of abdominal pain. Ultrasonography revealed a solid injury in the third portion of the left kidney (Fig.1) and the abdominal CT scan revealed a tumoral mass.

* Urogenital Research Unit, www.urogenitalresearch.org, Department of Anatomy, Biomedical Center, State of Rio de Janeiro University, Rio de Janeiro - Brazil.

** Department of Morphology, Biomedical Institute, Fluminense Federal University, Rio de Janeiro - Brazil.

Due the high index of suspicion of malignant neoplasia the patient was submitted to left radical nephrectomy. The tumour was solid and hard and, the cut surface of the tumour was homogenously colored pale pink. Microscopically it was haemangiopericytoma arising in the renal sinus, composed of well-developed pericytes and capillaries with so-called staghorn configuration.

The histopathological performed with Haematoxylin-eosin showed a monotonous cellular proliferation with no significant variability in cellularity, a 'staghorn' vascular pattern with pericytes packed around endothelium vascular channels (Figs. 2 and 3). Additionally, the immunohistochemistry (Avidin-Biotin Complex = ABC) revealed neoplastic cells immunopositive for anti-vimentin antibody (Figs. 4A and 4B) and anti-CD34 antibody (Fig. 5) associated to anti-keratin which was negative, confirming the differential diagnosis of renal haemangiopericytoma.

DISCUSSION

Haemangiopericytoma is an unusual perivascular

tumour, known to pathologists for >60 years, and classified as a soft-tissue vascular tumour featuring the uncontrolled proliferation of pericytes (Fornaro *et al.*), which are cells spiralling around capillaries, described by Zimmermann.

These cells are modified SMCs that surround capillaries and modify the diameter of capillary lumen, are thought to possess contractile powers, and appear to be associated with the control of caliber of the vessels (Argyropoulos *et al.*). These pericytes are in all capillary nets, haemangiopericytoma can appear in any part of the body. From 1942, when Stout & Murray described the first case of haemangiopericytoma, many things have changed, and even its very existence as a separate tumour type has been doubted (Fletcher, 1994). Because these tumours are rare, questions remain about the preoperative diagnosis, the ideal therapeutic strategies and the prognosis of the disease (Argyropoulos *et al.*). This tumour is generally rare in the urogenital system, and except for the kidney, other sites include the bladder, the prostate and the spermatic cord (Baumgartner *et al.*; Chen, 1987; Siemens *et al.*).

Fig. 1. Abdominal ultras-sonography showing a large mass; Haemangiopericytoma of the left kidney.

Fig. 2. Malignant renal haemangiopericytoma in a 79 year-old-patient, showing a 'staghorn' vascular pattern with pericytes (SMCs) packed around endothelium vessel (V) channels. Haematoxylin-eosin, X1000.

Fig. 3. Haematoxylin-eosin showed a cellular proliferation (arrow) with significant variability in cellularity, V= vessel, X1000.

Fig. 4A. Neoplastic cells immunopositive by ABC immunostaining anti-vimentin (arrows). V= vessel. X400.

Fig. 4B. Immunostaining anti-vimentin (arrows) on longitudinal vessel endothelium (V) with increased smooth muscle cells (SMCs). X400.

Fig. 5. Immunostaining to show the vascular (asterisk) endothelial cells immunopositive by ABC immunostaining anti-CD34 (arrow). X400.

For Argyropoulos *et al.* most patients (67%) were aged 16–50 years, and 39% were < 30 years old (data from 33 patients). This age distribution suggests that the kidney is affected by haemangiopericytoma in slightly younger patients than other types of RCC. There was no difference in mean age between sexes. There was a slight difference in the incidence in male and female patients, with a predominance of women (17 vs 16), in accordance with the reported data from haemangiopericytomas of various anatomical sites (McCormack & Gallivan). Most patients were aged <50 years and the most common symptom was the presence of a usually painless mass, as found in the case we report. Ultra-sonography and CT scan provide evidence of a (usually) large tumour, which may grow to a diameter of > 25 cm. Angiography may be a valuable preoperative examination and embolization before surgery may offer a better surgical field.

The pathological features of the tumour, together with the immunohistochemical results, confirm the differential diagnosis (Espat *et al.*, 2002). The differentiation between benign and malignant tumours is difficult, the presence of mitoses, increased cellularity, necrosis and haemorrhage suggests a malignant potential (McMaster *et al.*, 1975; Fletcher; Fornaro *et al.*; Espat *et al.*), and although it did not occur in the case we report. The prognosis seems to be related to size, age at diagnosis, histological patterns and recurrence of the tumour (Stout & Murray; Espat *et al.* and Argyropoulos *et al.*).

The histological pattern and the immunohistochemical results provide information for the differential diagnosis from other vascular neoplasms, and from mesenchymal tumours (Espat *et al.* and Argyropoulos *et al.*). Antibodies, e.g. those against CD31, CD34 and CD68, vimentin and cytokeratins, are used. The antibodies against CD34 and vimentin were used successfully in our study, since, these antibodies are characteristic of mesenchymal origin, and can identify neoplastic progenitor cells surrounding vascular spaces (Espat *et al.*).

According to Argyropoulos *et al.*, the prognosis seems to be related to size, age at diagnosis, histological patterns and recurrence of the tumour. As a rule, the most common metastatic site of renal haemangiopericytoma is the lungs, and these patients had the worst prognosis; both died, at 3 and 9 months after the initial diagnosis (Ordonez *et al.*, 1982; Espat *et al.*). The findings of this report and literature (Black & Heinemann; Ordonez *et al.*; Fornaro *et al.*; Espat *et*

al.; Argyropoulos *et al.*) of all previously published cases of renal haemangiopericytoma confirm the rarity of the tumour, and therefore the difficulties in obtaining valid information about its clinical course.

Our patient fared well and was alive 5 years after radical nephrectomy and to our knowledge, it was the 1st case in the Brazilian women with incidentally detected renal haemangiopericytoma.

RIBEIRO, J. G. A.; KLOJDA, C. A. B.; MACHADO, F. A.; FAVORITO, L. A.; CAVALCANTE, A. G. L. & BABINSKI, M. A. Hemangiopericitoma renal unilateral: reporte de un raro tumor. *Int. J. Morphol.*, 24(2):165-170, 2006.

RESUMEN: Los pericitos son células musculares lisas modificadas de los tubos capilares circundantes los cuales cambian el diámetro del lumen vascular. En este artículo relatamos un caso de hemangiopericitoma renal presente en una mujer de 78 años, quien manifestaba dolor abdominal. La ultrasonografía determinó una lesión sólida en la tercera porción del riñón izquierdo. La exploración abdominal de CT reveló una masa tumoral. Debido a la alta sospecha de neoplasia maligna, la paciente fue sometida a nefrectomía radical izquierda. La evaluación histopatológica confirmó el hemangiopericitoma, a través de inmunohistoquímica. La paciente sobrevive cinco años después de la nefrectomía radical.

PALABRAS CLAVE: Riñón; Tumor; Hemangiopericitoma; Patología; Inmunohistoquímica.

REFERENCES

- Argyropoulos, A.; Liakatas, I. & Lykourinas, M. Renal haemangiopericytoma: the characteristics of a rare tumour. *BJU Int.*, 95:943-7, 2005.
- Baumgartner, G.; Gaeta, J.; Wajzman, Z. & Merrin, C. Hemangiopericytoma of the urinary bladder: a case report and review of the literature. *J. Surg.Oncol.*, 8: 281-6, 1976.
- Black, H. R. & Heinemann, S. Hemangiopericytoma: report of a case involving the kidney. *Urol.*, 74:42, 1955.
- Chen, K. T. Hemangiopericytoma of the prostate. *J. Surg. Oncol.*, 35:42-3, 1987.
- Espat, N. J. ; Lewis, J. J.; Leung, D.; Woodruff, J. M.; Antonescu, C.R.; Shia, J.; Brennan, M.F. Conventional hemangiopericytoma. Modern analysis of outcome. *Cancer*, 95:1746-51, 2002.
- Fletcher, C. D. Hemangiopericytoma: a dying breed? Reappraisal of an entity and its variants. *Curr. Diagn. Pathol.*, 1:19-23, 1994.
- Fornaro, R.; Terrizzi, A.; Secco, G.B.; Canaletti, M.; Baldi, E.; Bonfante, P.; Sticchi, C.; Baccini, P.; Cittadini, G. Jr.; Fiorini, G.; Ferraris, R. Renal hemangiopericytoma. Anatomopathologic and clinico-therapeutic considerations. A case report. *G. Chir.*, 20:20-4, 1999.
- McCormack, L. J. & Gallivan, W. F. Hemangiopericytoma. *Cancer*, 7:595-601. 1954.
- McMaster, M. J.; Soule, E. H. & Ivins, J. C. Hemangiopericytoma. A clinicopathologic study and long-term followup of 60 patients. *Cancer*, 36: 2232-44, 1975.
- Ordonez, N.G.; Bracken, R. B. & Stroehlein, K.B. Hemangiopericytoma of kidney. *Urol.*, 20:191-5, 1982.
- Rubin, E. & Farber, J. L. *Pathology*. J. B. Lippincott Company Philadelphia, 1988. p. 494.
- Siemens, D. R.; Nickel, J. C. & Young, I. D. Hemangiopericytoma of the spermatic cord. *Br. J. Urol.*, 81: 329-30, 1998.
- Stout, A. P. & Murray, M. R. Hemangiopericytoma: a vascular tumor featuring Zimmermann's pericytes. *Ann. Surg.*, 116:26, 1942.

Zimmermann K. W. Der feinere Bau der Blutcapillaren.
Ztschr. Ant. Entwcklngsgesch, 68:29, 1923.

Correspondence to:
Prof. Dr. Márcio A. Babinski
Department of Morphology,
Biomedical Institute,
Fluminense Federal University,
Rio de Janeiro - BRASIL

Fax: + 55 21 2587-6121

E-mail: *mababinski@gmail.com*
babinski@vm.uff.br

Received : 13-12-2005

Accepted: 02-02-2006