Unilateral Renal Haemangiopericytoma: Report of a Rare Tumour

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 metastasise 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.
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 Haematoxilin-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 ultrasonography 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. Haematoxilin-eosin, X1000.

Fig. 3. Haematoxilin-eosin showed a cellular proliferation (arrow) with significant variability in cellularity, V= vessel, X1000.
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. Ultrasoundography and CT scan provide evidence of a (usually) large tumour, which may grow to a diameter of > 25 cm. Angiography may be available 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.


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. En la ultrasonografía se 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.

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