CASE REPORT

Pararenal sclerosing PEComa

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SUMMARY

Small renal or pararenal masses and retroperitoneum lesions are extremely difficult to diagnose. Imaging technology is a precious diagnostic tool; however, it places physicians in a difficult position since many lesions are not precisely diagnosed. Clinical and radiological findings can guide suspicion towards the diagnosis; however, in our current practice most diagnoses are based on histological findings. We aim to present a pararenal sclerosing perivascular epithelioid cell tumour (PEComa), a rare entity, whose diagnosis is only possible through invasive approaches and histological analysis. This rare lesion not only is difficult to diagnose but also has an uncertain behaviour, which is of major importance concerning its follow-up and prognosis. This case report is an attempt to add more data that will help establish criteria for diagnosis and follow-up of this rare disease.

BACKGROUND

Nowadays, there are enormous difficulties to diagnose accurately renal or pararenal masses. Despite some benign or malignant imagiological aspects, many lesions have unclear diagnosis and behaviour. Most of these lesions are rare, have a brief follow-up and ambiguous classifications which obscure their diagnoses. Perivascular epithelioid cell tumours (PEComas) are one of the challenging diagnoses. PEComas are a heterogeneous group of mesenchymal neoplasms, characterised by epithelioid cells with immunoreactivity to human melanoma black-45 (HMB-45), however in sclerosing cases HMB-45 (seen predominantly in smooth muscle), is confined to sclerotic areas, not in epithelioid cells. The sclerosing variants have characteristically a sclerotic stroma and HMB-45 negative epithelioid cells have incomplete melanocytic differentiation based on the positive reaction to MART-1.¹ This sclerosing variant was recently described (2008) and represents 19% of all PEComas.² It is a rare entity with ubiquitous distribution (pararenal and retroperitoneum preference) with uncertain behaviour (but predominantly benign) and more frequently seen in middle-age women.²⁻⁴ This is a case of a sclerosing pararenal PEComa that has had a benign course (4 years of follow-up). This clinical evolution is in agreement with the known published cases; however, this case report adds important information since it has a long follow-up period. It would be important in the near future to establish rigorous criteria to identify malignant features to precisely recognise these lesions and their comportment.

CASE PRESENTATION

A 51-year-old woman was referred to our outpatient urology clinic complaining of a continuous left back (lumbar) pain. Persistent (2 months) pain

symptoms, without relief with position or analgesics. She had no known pathology including renal (lithiasis), vascular disease either obstructive uropathology or tuberous sclerosis, nor osteoarticular disease. She had no previous injury or trauma. No urinary symptoms or haematuria. The patient did not have any significant familial disease (namely cancer) or any constitutive symptoms.

Physical examination was normal. Abdominal CT performed (2008) showed a rounded-to-oval mass on her left flank with approximately 5 cm. Renal AngioTC (figure 1) showed cleavage plane within the lower pole of the left kidney and psoas muscle with a slight enhancement after contrast. Such findings were not possible to distinguish from an angio-myolipoma (AML) or a renal cell carcinoma. Further tests were performed, namely an abdominal magnetic resonance image (MRI) that showed a lesion with 5.9×4.3×4.9 cm hypointense on T1 (figure 2A) and T2 (figure 2B), with no signs of invasion, apparent cleavage plane with the psoas muscle. Biochemical analysis was performed to exclude hypothetical paraganglioma.

Transperitoneal laparoscopic excision was performed (figures 3 and 4). Pathology revealed a pararenal sclerosing PEComa completely excised.

INVESTIGATIONS

After surgery, diagnosis was made through histopathologic evaluation.

In this particular and rare case it was important to exclude its malignancy and purpose a strict follow-up.

Macroscopically tumour was well defined and solid (figure 5).

Microscopic histological features and immunohistochemical phenotype were characteristic for sclerosing PEComa. We found cells with an epithelioid appearance, a clear pale eosinophilic

Figure 1 Renal AngioTC—5 cm lesion attached to left kidney.

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Figure 2 (A and B) Renal RM—5.9×4.3×4.9 cm pararenal lesion hipodense without cortex invasion ((A) T1-weighted images; (B) T2-weighted images).

Figure 3 Transperitoneal laparoscopic approach.

Figure 4 Macroscopic appearance.

cytoplasm and perivascular distribution with hyalinised fibrous cord, without tumoral necrosis and normal mitotic index ($<1/50$ HPF; figure 6). HMB-45-immunoreactive cells are scattered in sclerotic areas and epithelioid cells are diffusely positive for muscle-specific actin and desmin. It was possible to observe hyalinised blood vessels alternating with hypocellular collagenous-hyalinised areas, sometimes calcified. Cellular areas are less than hyalinised ones. MNF116, protein S100, CD117 and CD34 were negative.

DIFFERENTIAL DIAGNOSIS

PEComas comprise a numerous family of different lesions as angiomyolipoma, lymphangiomyomatosis, clear cell 'sugar' tumour of the lung and other visceral lesions.³

Paraganglioma is also an important diagnosis to be taken into account.

These tumours may also manifest as sarcomas, with undifferentiated appearance that should be considered in differential diagnosis.⁵

OUTCOME AND FOLLOW-UP

Currently the patient is clinically well (follow-up 4 years). Abdominal and pelvic CT (3 months) abdominal ultrasound (6 months and 1 year) and abdominal CT (4th year) showed no

Figure 5 Histology—H&E stain (×400), epithelioid cells with perivascular distribution, hyalinised stroma. Normal mitosis index. No tumoral necrosis observed.

Figure 6 Immunohistochemistry—human melanoma black-45 stain.

signs of local or distant disease. Besides the common benign evolution some PEComas had unfavourable course with metastatic disseminations (lung, liver, bone and abdominal wall)^{2 4 6}; however, in vast majority of published data sclerosing PEComas have benign course, which is in agreement with the presented case.

DISCUSSION

According to WHO PEComa lesions, encompasses angiomyolipoma, including epithelioid variants, clear cell sugar tumours of the lung, lymphagioleiomyomatosis, clear cell myomelanocytic tumour, unusual clear-cell tumours and a group of uncommon lesions that arise in soft tissue, visceral organs or skin.^{2 7}

PEComas are now widely recognised histologically, and have been reported to occur in different anatomical sites. However, a distinctive variant of PEComa ‘sclerosing PEComa’ has only recently been described in the literature.²

There are few published cases of pararenal sclerosing PEComas^{4 8} and known data confirm the difficulty for diagnosis and follow-up for this entity.

Physicians still have an insufficient knowledge about its imagiological appearance and which characteristics determine a malignant behaviour.

Although sclerosing PEComas seem to have a benign course, some authors affirm that unfavourable outcome is linked with lesions larger than 7 cm in size, high mitotic activity, necrosis, high nuclear grade or infiltrative growth.³

Learning points

- ▶ PEComas constitutes a heterogeneous group of mesenchymal neoplasms.
- ▶ Features that can guide the diagnosis are: middle-age women, pararenal retroperitoneum lesion, human melanoma black-45 immunoreactivity and epithelioid cells appearance.
- ▶ Sclerosing PEComa is a distinctive variant for of benign renal tumour with an indolent clinical course in most cases; however, there are published cases of metastatisation.
- ▶ Important prognostic factors to predict aggressiveness are: size, high mitotic activity, necrosis, high nuclear grade or infiltrative growth.

Competing interests None.

Patient consent Obtained.

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