



Scientific letters

Primary Liver PEComa[☆]

PEComa hepático

Perivascular epithelioid cell tumours (PEComa) are rare mesenchymal neoplasms that express myogenic and melanocytic markers.

We present the case of a 45-year-old woman who was being studied for dyspepsia and elevated Ca 19.9. CT detected a 4-cm lesion in liver segments V/VI, with heterogenous enhancement in arterial phase and almost complete washout in portal phase; the uterus was myomatous, with a left adnexal cyst. MRI revealed a hyperintense lesion in T2 that was hypointense in T1, with uptake in the arterial phase and washout in portal phase, except in the periphery, which presented delayed enhancement (Fig. 1). A follow-up ultrasound demonstrated growth of the mass, and therefore we decided on resection; the patient had an uneventful recovery. The pathology study determined the tumour was a PEComa measuring 3.5 cm in diameter, with free surgical margins, marked nuclear pleomorphism, and no presence of mitosis. The proliferation rate

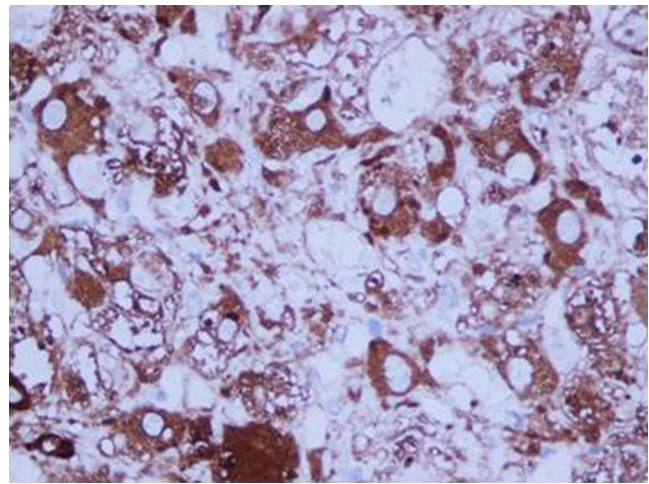


Fig. 2 – Expression of Melan-A melanocyte marker.

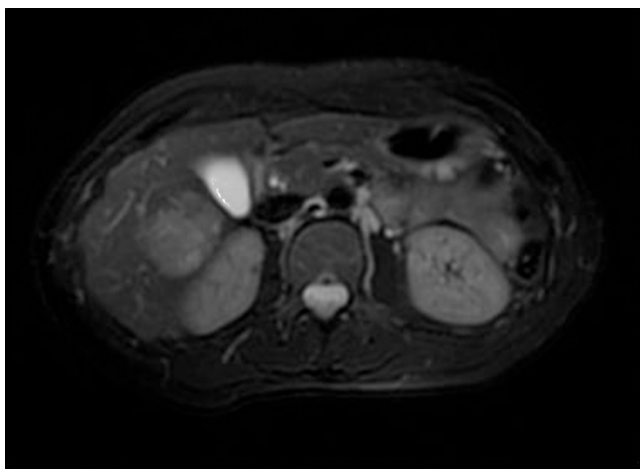


Fig. 1 – MRI demonstrating hyperintense SOL in T2.

(Ki67) was 3%–5% and the tumour expressed HMB-45, Melan-A (Fig. 2), 1A4 and HHF35 with uncertain behaviour. Afterwards, and given the suspicion that the hepatic mass was probably metastasis of gynaecological origin, hysterectomy and double adnexectomy were carried out. The histopathologic study showed a myomatous uterus and 5-cm leiomyoma that expressed HHF35 and was negative for melanocytic markers (HMB-32 and Melan-A), with no other adnexal pathological findings.

PEComas are rare mesenchymal neoplasms where the epithelioid cells express muscle (actin, desmin) and melanocytic (HMB-45; Melan-A, MiTF) markers.¹ Less than 50 cases of hepatic PEComa have been described.⁵ They predominantly affect women with a mean age of 46.³ The most frequent location is genital (especially the uterus), followed by the genitourinary apparatus, gastrointestinal tract and retroperitoneum; it is also associated with tuberous sclerosis.^{2–4}

[☆] Please cite this article as: Arribas Jurado M, Revollo I, Rubio Fernández A, Galeano Díaz F, Blanco Fernández G. PEComa hepático. Cir Esp. 2015;93:600–601.

Symptoms are variable: from no symptoms to discomfort in the right hypochondrium or acute abdomen.

The degree of diagnostic error by imaging techniques is higher than 90%,⁴ so it is therefore difficult to make a correct preoperative diagnosis. The differential diagnosis should include: focal nodular hyperplasia, hemangioma, hepatocarcinoma, adenoma, metastasis of endocrine or renal tumours, etc.

Standard treatment is radical surgery, both for the primary tumour as well as local recurrence or distant metastasis. The role of chemotherapy and radiotherapy is uncertain; these therapies are often considered in cases of locally advanced disease, local recurrence or distant metastasis, although their results are arguable.^{1,2,4}

The prognosis is difficult to establish because of the lack of understanding about the biological behaviour of these tumours and the variety of results in the few studies published. According to some authors, size >5 cm, mitotic rate >1/50 HPF, necrosis, cells with nuclear pleomorphism, infiltrating growth or high nuclear grade are considered factors for poor prognosis and associated with more aggressive tumour behaviour, recurrence and/or distant metastasis.² Folpe^{1,6} proposed a classification based on these criteria: (a) benign, if there are none of the former parameters; (b) uncertain biological potential, if only size >5 cm or giant cells with nuclear pleomorphism are found; and (c) malignant, with more than 2 criteria for poor prognosis.

In our patient, the lesion was classified as having an uncertain diagnosis, and the case was presented at a multidisciplinary committee meeting for gastrointestinal tumours, at which time close follow-up of the patient was decided upon.

As stated by Khaja,⁵ we believe that, in cases of space-occupying lesions of the liver with uncertain radiological diagnosis, the treatment of choice is resection with free margins.

Conflict of Interests

The authors declare no conflict of interests.

REFERENCES

1. Cuevas Herrero Ó, Escobar Lezcano L, Rodríguez Blaco M, Artigas Raventós V. PEComa, un raro tumor de células epitelioides. *Cir Esp*. 2015;93:e65–7.
2. Navarro-Pelayo Lainez MM, Ramos Font C, Rebollo Aguirre AC, Rodríguez Fernández A, Llamas Elvira JM. Tumor epiteloide perivascular uterino. Utilidad de la tomografía por emisión de positrones con ¹⁸F-fluorodesoxiglucosa en su estadificación y seguimiento. *Rev Esp Med Nucl*. 2010;29:258–62.
3. Domínguez-González M, Coronel Villarán MJ, Rubio Garrido J, Holgado Fernández A, Herreros López JA, Martín Gutiérrez A. Tumor de células epitelioides perivasculares en cuerpo uterino asociado a complejo de esclerosis tuberosa en mujer de 18 años. *Prog Obstet Ginecol*. 2013;56:147–50.
4. Sánchez Pérez B, Suárez Muñoz MA, Aranda Narváez JM, Fernández Aguilar JL, Santoyo Santoyo J. Tumor de células epitelioides perivasculares (PEComa) hepático. *Cir Esp*. 2009;85:184–6.
5. Khaja F, Carilli A, Baidas S, Sriharan A, Nordford S. PEComa: a perivascular epithelioid cell tumor in the liver – a case report and review of literatura. *Case Rep Med*. 2013;2013:1–4.
6. Folpe AL, Mentzel T, Lehr H-A, Fisher C, Balzer B, Weiss SW. Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin. *Am J Surg Pathol*. 2005;29:1558–75.

Martina Arribas Jurado^{a,*}, Ivía Revollo^a,
Alejandro Rubio Fernández^b, Francisco Galeano Díaz^a,
Gerardo Blanco Fernández^a

^aServicio de Cirugía Hepatobiliopancreática y Trasplante Hepático, Hospital Infanta Cristina, Badajoz, Spain

^bServicio de Anatomía Patológica, Hospital Infanta Cristina, Badajoz, Spain

*Corresponding author.

E-mail address: marribasj85@gmail.com (M. Arribas Jurado).

2173-5077/

© 2014 AEC. Published by Elsevier España, S.L.U. All rights reserved.

Migration and Perforation of a Gastric Band in the Small Bowel[☆]

Migración y perforación de una banda gástrica en el intestino delgado

Laparoscopic adjustable gastric banding (LAGB) is a safe and reversible procedure that entails low mortality and morbidity (approximately 0.05% and 11.3%, respectively).¹ Even so, it has

a high readmission and revision surgery rate of 52%.¹ Late-onset postoperative complications are frequent and are probably not completely documented. Erosion and migration

[☆] Please cite this article as: Oppliger F, Ríos H, Manríquez L. Migración y perforación de una banda gástrica en el intestino delgado. *Cir Esp*. 2015;93:601–603.