H. parainfluenzae, a finding that should not be surprising based on recent research on the gastrointestinal habitat.

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Conflicts of interest

The authors declare that there is no conflict of interest with regard to the article.

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Our experience in the surgical treatment of liver PEComa☆

Nuestra experiencia en el tratamiento del PEComa hepático

Liver PEComas are extremely rare and differential diagnosis is complex. The treatment of choice is surgical removal. We present our experience (4 cases in 6 years) and a literature review.

Case 1

A 46-year-old female with a history of right hepatectomy in 1995 due to a hepatocellular carcinoma (HCC).

After 10 years, damage to segment II was found. The biopsy showed non-tumour hepatocytes. Four years later, the magnetic resonance imaging (MRI) scan showed a growth in size, hypervascular behaviour and a biopsy consistent with HCC.

There was 2.4 cm of damage in segment II on the intraoperative ultrasound, and a limited resection was performed.

The final outcome was PEComa.

The histological preparations from the prior resection were requested and they were confirmed as a match with a liver PEComa (company unknown in 1995).

Case 2

A 45-year-old female with a malignant melanoma previously resected in her lower left limb, Clark IV. After 8 years, the follow-up CT scan found a solid 25 mm lesion on the MRI described as: hypervascular, hyperintense in T1 and hypointense in T2.

Biopsy consistent with PEComa.

On a morphologically normal liver, the intraoperative ultrasound identified a 2.5 cm hyperechoic lesion, which was well defined in segment VIII. A limited resection was performed.

Case 3

A 48-year-old female diagnosed with ocular choroidal melanoma. The extension study identified a 7 cm mass in the liver hilum with a liver PEComa biopsy.

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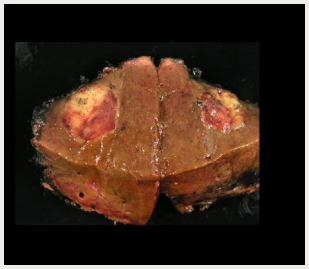
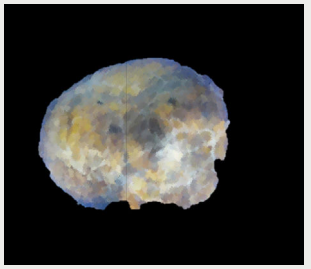
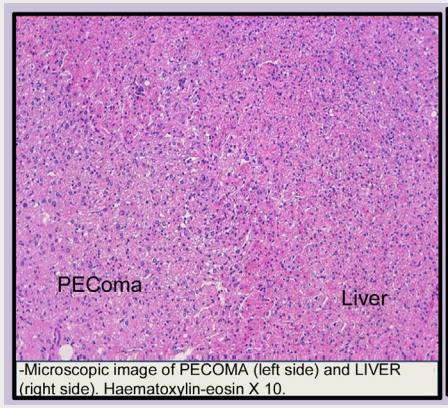
Table 1 Liver PEComas described in the literature review.

Author, year	Age and sex		Location	Size (cm)	Metastatic disease	Follow-up
Tsui, 1999	56	M	RLL	6.5	No	2 years
Tsui, 1999	37	F	RLL	3	–	–
Tsui, 1999	41	F	RLL	9	No	4 years
Tsui, 1999	46	F	RLL	12.5	No	12 years
Tsui, 1999	41	M	–	6	–	–
Yamasaki, 2000	30	F	RLL	3	No	1 year
Dalle, 2000	70	F	RLL	26	Hepatic	5 years
Yukichi, 2000	13	F	Lig. Teres	9	No	22 months
Xu, 2001	35	F	RLL	2 × 2	–	
Lin, 2002	30	F	RLL	3.6 × 3	–	
Parfitt, 2006	60	F	RLL	14	Multiple	9 years
Svajdler, 2007	55	F	LLL	3.5	–	–
Rouquie, 2007	67	F	LLL	9	No	2 years
Song Hua, 2007	67	F	Caudate	6	No	1 year
Song Hua, 2007	56	F	LLL	5	No	1 year
Labcharoensub, 2007	31	F	RLL	1.8	No	6 months
Li, 2007	56	F	LLL	5 × 4	–	–
Wang, 2007	46	F	RLL	4 × 4	–	–
Fang, 2007	56	F	LLL	5.1	No	24 months
Fang, 2007	56	F	LLL	5.1	No	24 months
Paiva, 2008	51	F	LLL	0.8	No	2 years
Sánchez, 2008	32	F	RLL	4	No	6 months
Qiu, 2008	67	F	RLL	15 × 12	–	–
Liu, 2008	31	F	RLL	8 × 6	–	–
Han, 2008	44	M	RLL	2 × 1.6	–	–
Zimmermann, 2008	53	M	–	8	No	17 months
Chen, 2009	36	F	RLL	7 × 5	–	–
Chen, 2009	45	F	RLL	5.5 × 4	–	–
Chen, Li, 2009	37	F	RLL	5 × 4	–	–
Strzelczyk, 2009	57	F	RLL	20	No	–
Akitake, 2009	36	F	LLL	3.5	No	18 months
Priola, 2009	36	F	LLL	11	No	34 months
Liu, 2010	32	F	RLL	5.5 × 5.5	–	–
Zhu, 2010	26	F	RLL	5 × 3	–	–
Shi, 2010	41	F	LLL	5.5 × 4	–	–
Shi, 2010	48	F	RLL	8 × 6	–	–
Deng, 2011	66	M	LLL	3 × 3.5	–	–
He, 2011	35	F	RLL	3.5 × 3	–	–
Zou, 2011	54	M	RLL	6 × 5	–	–
Selvaggi, 2011	42	M	RLL	7	Yes	1 month
Ahn, Hur, 2011	36	F	LLL	6.5	No	3 months
Zhang, Wang, 2012	55	F	RLL	3 × 3	–	–
Gao, 2012	59	F	RLL	6 × 5	–	–
Yan, Tan, 2012	38	F	RLL	4	No	48
Yan, Tan, 2012	34	F	RLL	4.3	Yes	14
Yan, Tan, 2012	49	F	LLL	2.5	No	32
Yan, Tan, 2012	75	F	RLL	8	No	36
Yan, Tan, 2012	33	F	RLL	2.5	No	27
Yan, Tan, 2012	71	F	RLL	–	No	29
Yan, Tan, 2012	41	F	LLL	Multiple	Yes	12
Liu, 2014	25	F	LLL	2.5 × 1	–	–
Otegi, 2015	46	F	LLL	2.4	No	6 years
Otegi, 2015	45	F	RLL	2.9	No	4 years
Otegi, 2015	48	F	Caudate	8	No	18 months
Otegi, 2015	53	M	LLL	2.5	No	11 months

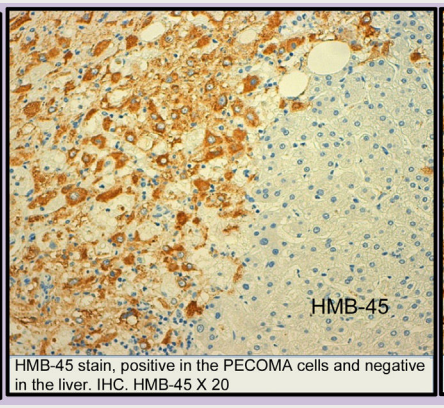
RLL, right lobe of liver; LLL, left lobe of liver; F, female; M, male; –, not found.

Table 2 Images and anatomopathological characteristics and immunohistochemistry.

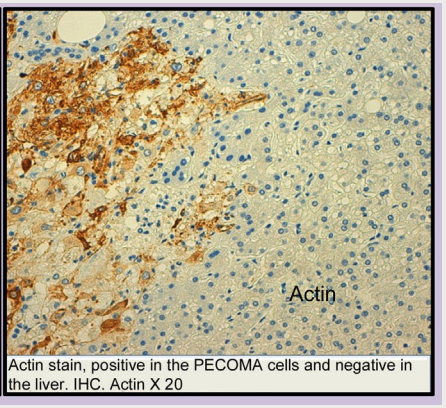
	Case 1	Case 2	Case 3	Case 4
Pieces				
Macroscopic description	White nodule that has a maximum diameter of 1.6 × 1.7 cm	Well-defined nodule, 2.9 cm in diameter with yellowish and dark brown coloured areas	Subserosal nodule 9 cm in diameter	Nodular lesion 2.5 cm in diameter with a liver-like colour and granular consistency
Microscopic description	Proliferation of epithelioid and spindle cells with eosinophil cytoplasm. Thick vascular structures were noticed, which break up both spindle and epithelioid cells. In one of the slices, adipose tissue was found interspersed with the cellular proliferation	Lumps that show different sized mature adipocytes, interspersed with epithelioid-like cells with large, clear, eosinophilic cytoplasm in groups and around veins with uneven walls	Proliferation of eosinophilic polygonal cells associated with veins which are dilated in appearance and showing a proliferation of elongated, myoid-like cells, epithelioid and adipocyte cells	Proliferation of polygonal cells with a cytoplasm that oscillates between clear and eosinophilic and a diffuse or trabecular growth with abundant medium-sized veins with surrounding epithelial cells. Approximately 30% of fat globules were observed interspersed with this proliferation
IHC positive	Melan-A HMB-45 Actin MITF, c-Kit	Melan-A HMB-45 Actin Calponin	Melan-A HMB-45 Actin	Melan-A HMB-45 Actin
IHC negative	Hepatocyte S-100	S-100	S-100	S-100
Ki-67 index	1%	<1%	1%	1%
Follow-up	6 years	4 years	18 months	11 months



-Microscopic image of PECOMA (left side) and LIVER (right side). Haematoxylin-eosin X 10.



HMB-45 stain, positive in the PECOMA cells and negative in the liver. IHC. HMB-45 X 20



Actin stain, positive in the PECOMA cells and negative in the liver. IHC. Actin X 20

During surgery, a lump in the caudate lobe was observed which compressed the vena cava and the left and middle suprahepatic veins (SHV). The caudate lobe was resected.

The eye lesion was subsequently treated using brachytherapy.

Case 4

A 53-year-old male with a history of frequent alcohol consumption examined due to general symptoms.

In the CT scan a 3 cm nodule was identified, adjacent to the left SHV, hypervascular with uniform uptake in the arterial phase.

Biopsy: angiomyolipoma.

In the intraoperative ultrasound, a 3 cm hyperechoic lesion was seen, which reached the left and middle SHV. A left hepatectomy was performed.

Images, pathological descriptions, immunohistochemical markers and the follow-up time are shown in Table 1.

There were no deaths and no post-operative complications during the post-operative follow-up.

In 1991, Pea et al.[1] described the presence of a cell type with specific cytoplasmic characteristics and strong positivity for HMB-45 in the angiomyolipomas (AML). One year later, these cells were given the name of *perivascular epithelioid cell* (PEC).[2]

In 2001, the World Health Organisation (WHO) defined PEComas as a group of rare mesenchymal tumours comprised of perivascular epithelioid cells with well-defined histologic and immunohistochemical characteristics.[3]

Currently the following are included in the PEComas group: angiomyolipoma (AML), pulmonary lymphangioleiomyomatosis, clear cell ''sugar'' tumours and clear cell myomelanocytic tumours of the falciform ligament.[4]

In a systematic review in 2012, Bleeker et al.[5] compiled 234 cases. The uterus is the most common location, and it is rare in the liver. In our literature search, we found 51 described cases of liver PEComas (Table 2).

The clinical presentation of the liver PEComa is variable and nonspecific, usually diagnosed by chance.[6–8]

The differential diagnosis of liver PEComa includes HCC, haemangioma, focal nodular hyperplasia and liver adenoma.[8]

In 2012, Tan and Xiao[6] published 7 cases of liver PEComas. Radiologically they were well-defined masses with uniform density. In both the CT scan and the intravenous contrast-enhanced MRI, there was varied and premature enhancement in the arterial phase with uniform clearance in venous and late phases. The radiological characteristics are similar to the HCCs. We must consider the PEComas in light of radiological patterns described in patients without elevated alpha-fetoprotein (AFP) and no hepatitis or cirrhosis.

The PEComas are made up of epithelioid cells, linked with vascular structures, eosinophil cytoplasm, abundant glycogen and the characteristic presence in the ultrastructure of premelanosomes. They show immunoreactivity against melanocytic markers HMB-45, microphthalmia transcription factor (MITF), Melan-A and *PNL2* melanoma-associated antigen, which indicates melanocytic differentiation between the tumours. They are generally negative for the S-100 which makes the differential diagnosis with melanoma less difficult.[4,8]

Two of the cases have a history of a melanoma, which makes the differential diagnosis with metastases of this tumour more difficult. But in both cases, in addition to the negativity for the S-100, vascular and adipose proliferation were recognised, which is typical of AML.

The co-expression of actin and HMB45/Melan-A is indispensable for diagnosing PEComa and, together with the negativity for the S-100, it allows for a differential diagnosis with a metastatic melanoma.

They can show variable expression for other melanocytic markers, including the S-100 protein, tyrosinase and the microphthalmia transcription factor (MTF) and for other muscle markers such as smooth muscle myosin (SMMS) and calponin.

c-Kit expression in PEComa cases is also described.

The majority of published PEComa cases seem to have a benign behaviour, although cases of tumour recurrence or distant metastasis have been described.[5] The tumour size (greater than 7 cm) and the pathological characteristics (high mitotic index and cellular necrosis) are considered a threat for recurrence.[6]

The most effective currently accepted treatment is surgery.[5,9] Adjuvant therapy is advised along with chemotherapy in PEComas with risk indicators.[5,10]

Despite PEComas being extremely rare, they are part of an emerging group of tumours.

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2444-3824/

Clinical features and outcome of acute ischemic proctocolitis

Características clínicas y pronóstico de la proctocolitis isqémica aguda

Ischemic injury to the rectum is rare owing to its rich vascular supply, occurring in <6% of the cases of ischemic colitis.[1,2] As in ischemic colitis, a spectrum of severity exists and ranges from superficial mucosal ischemia to full-thickness necrosis with perforation.[3] Early recognition of this clinical entity is of vital importance in order to avoid complications.[1] The authors report a series of 5 cases of acute ischemic colitis with rectum involvement and present review of the literature.

In this report, 5 patients were included, 4 man and 1 woman, with a median age of 70 year-old. The prevalence of cardiovascular risk factors was high (80%). All cases were admitted for lower gastrointestinal bleeding. None of the patients were taking nonsteroidal anti-inflammatory drugs in the days before presentation. In all patients who presented with bloody diarrhea, stool cultures (including *Escherichia coli* O157:H7), stool examination for ova (including *Entamoeba histolytica*) and parasites and *Clostridium difficile* toxin assay were obtained, and were negative. In 3 cases, a conservative approach was assumed due to their clinical stability and resolved without complications. The remaining 2 cases required a surgical approach and were admitted to an intensive care unit following surgery. One of these cases required multiple surgical procedures for complications. The median duration of hospitalization was 8 (IQR: 62) days, and all patients survived. The clinical features of each case are summarized in Table 1.

A report of each case is described below.

The first case concerns to a 70-year-old man, with a medical history of insulin dependent diabetes mellitus, left hemicolectomy for sigmoid adenocarcinoma (15 years before) and peripheral vascular disease requiring ileofemoral bypass 4 months before. The patient was admitted for bloody diarrhea, nausea and vomiting. Proctosigmoidoscopy was performed and revealed a purplish black proximal rectal mucosa and edema (Fig. 1) consistent with ischemic proctitis that was confirmed by histological examination. The mucosa of the distal rectum was spared. A computed tomography (CT) scan was performed and showed signs of acute ischemia from the rectum to the transverse colon, with an abnormal opacification of the inferior mesenteric artery. Due to the clinical stable condition, a conservative approach was assumed and the patient started on broad-spectrum antibiotics. He was discharged 8 days after, asymptomatic.

The second case was an 87-year-old woman with arterial hypertension. She recurred to the emergency department for constipation and sudden crampy abdominal pain. She was treated on antibiotics for a urinary tract infection until 3 days before being admitted. The patient presented rectal bleeding in the emergency department. CT scan showed thickening of the sigmoid colon and rectal wall. She underwent a proctosigmoidoscopy which revealed erythema, edema and ulceration of the rectum, from 5 cm proximal to the dentate line. Mild ischemic changes extended up to the splenic flexure. Biopsies were performed and the histopathological exam confirmed the endoscopic suspicion

Table 1 Clinical features of the patients with acute ischemic proctocolitis.

Case	Case 1	Case 2	Case 3	Case 4	Case 5
Gender	Male	Female	Male	Male	Male
Age	70	87	65	78	65
Diabetes mellitus	Yes	No	Yes	Yes	No
Dyslipidemia	No	No	Yes	No	No
Arterial hypertension	No	Yes	Yes	No	No
Presenting symptom	Bloody diarrhea	Rectal bleeding	Hematochezia	Rectal bleeding	Rectal bleeding
Treatment	Fluids Antibiotics	Fluids Antibiotics Hartmann's resection	Fluids Antibiotics	Fluids Antibiotics Hartmann's resection	FluidsAntibiotics
ICU admission	No	Yes	No	Yes	No
Hospitalization days	8	13	5	123	13

ICU: intensive care unit.