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Case Report OMICS International

# PEComa of the Colon Presentation of a Case and Review of the Literature

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**Abstract**

**Introduction and objectives:** PEComas are very rare tumors even their location in the colon.

**Material and Methods:** We report a case of a 42 years old male patient with an acute abdomen and a PEComa tumor dependent of the cecum wall. He underwent emergency surgery for right hemicolectomy and mesenteric implant resection

**Discussion:** PEComas (Perivascular Epithelioid Cells tumors) are rare mesenchymal neoplasms. In the digestive tract there are few cases reported in the literature.

## Introduction

PEComas (Perivascular epithelioid cell tumors) are rare mesenchymal neoplasms [1]. There is a marked female preponderance and a tirad of cases occur in the pediatric age group [2]. They have been identified in multiple anatomical sites such as the liver, uterus, vulva, rectum, heart, lung, bladder, pancreas and abdominal wall. They are generally associated with few symptoms, and if they exist, are usually abdominal pains and bleeding [1,3-6].

Due to their low frequency, their clinical and biological behavior is not well known [1]. It has different histological and immunohistochemical characteristics defined by melanocytic (HMB-45) and smooth muscle (actin and desmin) markers co-expression [3], but the negativity to S-100 protein [4].

The main treatment is surgical excision. However, chemotherapy and radiotherapy have been used in locally advanced, metastatic and malignant histology tumors.

## Clinical Case

We report a case of a 42 years old male, with no clinical background, attended at emergency room for 48 hours of pain and bloating, associated with fever and feeling of rectal and urinary urgency.

Physical examination revealed a moderately distended and tenderness on the left upper quadrant and right flank, with defense associated.

The requested blood test had a leukocyte range of $14 \times 10^3/\mu L$ (reference 4-10.5 $\times$ $10^3/\mu L$) with associated neutrophilia, PCR 167 mg/dl (reference less than 5 mg/dl).

An urgent abdominal CT scan revealed a large abdominal mass about 21 cm (latero-lateral) $\times$ 13 cm (antero-posterior) $\times$ 23 cm (craneocaudal) (Figure 1), extending from left upper quadrant to pelvic.

It was cystic-solid lobed and associated trabeculation of the adjacent fat, occupying from cecum to the left colon, contacting with the sigma and the bladder within organ dependence established.

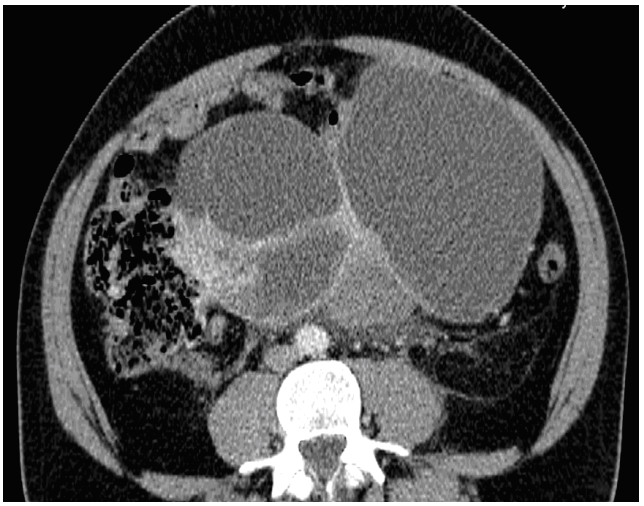

**Figure 1:** Abdominal CT scan with a mass dependent of cecum wall.

With these findings and exploration, the patient underwent an urgent exploratory laparotomy. A large double cystic-solid tumor of 10-12 cm depending of the anterolateral cecum wall was evidenced (Figure 2). There was also an implant similar to the mass at mesentery.

A right hemicolectomy and resection of the implant was performed. There were no incidences during the postoperative period and the patient was discharged on the fifth day.

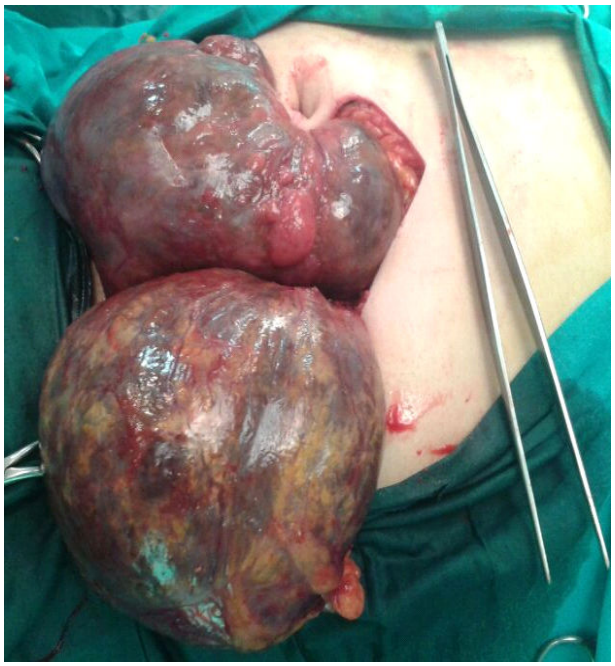

**Figure 2:** Large double cystic-solid tumor of 10-12 cm depending of the anterolateral cecum wall.

The pathological specimen study revealed a PEComa with at least three high-risk criteria (high cellularity and pleomorphism, size >5 cm and infiltrative growth pattern), which corresponds to a malignant PEComa. However, the mitotic index is very low (1 mitosis/50 high power fields) and although there is wide hemorrhage and cyst formation, really necrosis is not observed. No vascular invasion seen (Figure 3).

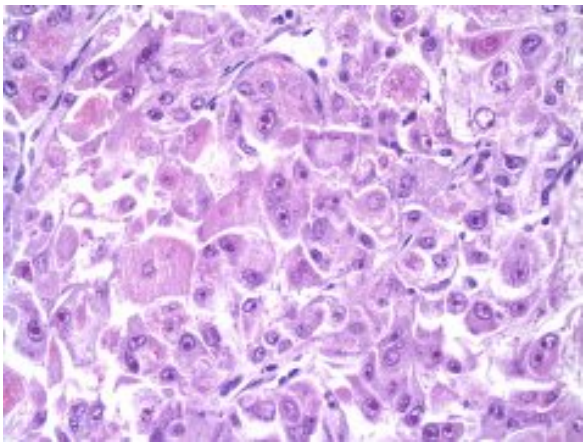

**Figure 3:** Microscopic detail, with epithelioid cells with large and eosinophilic cytoplasm, with large nuclei and prominent nucleoli. Among the cell nests are seen fine vessels (HE; 400×).

Currently the patient is untreated and free from disease two years after surgery.

## Discussion

PEComas are tumors characterized by melanocytic differentiation. In the colon very few cases have been described, with an estimated incidence <0.1% of colon cancers [7].

For this low frequency and because of its histologic features are variable, it is important differential diagnosis to exclude other tumors. It should include various types and mesenchymal epithelial tumors, including malignant melanoma, clear cell sarcoma, gastrointestinal stromal tumors (GIST) and epitheloid leiomyosarcoma [8].

Due to the expression of HMB45, malignant melanoma and clear cell sarcoma should be excluded. Furthermore, PEComas shows positivity for S100 protein and lacking expression of biogenic markers [9]. Moreover, the characteristic of genomic rearrangements (q13; q12) observed in almost 90% of clear cell sarcoma was not observed in these tumors [10].

It is also important to include GIST in the differential diagnosis because they are the most common mesenchymal tumors of the digestive tract. Immunohistochemistry to detect c-kit detector tyrosine kinase (CD117), CD34, y HMB45 is useful for diagnosis because, unlike GIST, the HMB45 expressing PEComas rarely express CD117, and are negative for CD34 [7]. Moreover, tumor cells positive for CD117 show membranous staining pattern mild to moderate, in contrast to the strong membranous cytoplasm pattern of GIST [11].

The biological behaviour of this kind of tumor is it not well defined [3]. Criterial histology for malignancy is unclear, partly, because of the long term data about natural history is unknown for intestinal lesions.

A size greater than 5 cm, areas with coagulative necrosis and high rate mitosis are considered as malignant behavior. Other important criteria for malignancy include: edge tumor resection infiltration, high cellularity and vascular invasion [7]. Clinical feature is nonspecific, so preoperative diagnosis it difficult to accomplish [8].

There is no definitive treatment of PEComa. Surgical resection of the tumor it is usually enough in benign behaviour cases. However, infiltrative or metastatic tumors usually have a poor prognosis, and treatment strategy include chemotherapy, radiotherapy, and immunotherapy [12]. Nevertheless, because of the small number of cases, there are controversies, especially in advanced cases. [13].

Regarding follow up and survival, patients with intestinal lesions have been short term follow up, so data is limited. It is necessary more documented cases to fully define its natural history and behaviour of this kind of tumor [7].

In conclusion, it's a rare tumor, especially when located at the colon, with difficult diagnosis, and nonspecific clinical features. Like in this case, with the particularity of had being surgically treated in the context of acute abdomen.

## References

1. Baek JH, Chung MG, Jung DH, Oh JH (2007) Perivascular Epithelioid Cell Tumor (PEComa) in the transverse colon of an adolescent: a case report. Tumori 93: 106-108.
2. Ryan P, Nguyen VH, Gholoum S, Carpineta L, Abish S, et al. (2009) Polypoid PEComa in the rectum of a 15-year-old girl: case report and review of PEComa in the gastrointestinal tract. Am J Surg Pathol 33: 475-82.

3. Doyle LA, Hornick JL, Fletcher CD (2013) Pecoma of the gastrointestinal tract: clinicopathologic study of 35 cases with evaluation of prognostic parameters. Am J Surg Pathol 37: 1769-82.
4. Martignoni G, Pea M, Reghellin D, Zamboni G, Bonetti F (2008) PEComas: the past, the present and the future. Virchows Arch 452: 119-132.
5. Priola AM, Priola SM, Cataldi A, Marci V, Fava C (2009) Acute abdomen as an unusual presentation of hepatic PEComa-A case report. Tumori 95: 123-128.
6. Martignoni G, Pea M, Reghellin D, Gobbo S, Zamboni G, et al. (2010) Molecular pathology of lymphangioleiomyomatosis and other perivascular epithelioid cell tumours. Arch Pathol Lab Med 134: 33-40.
7. Freeman HJ, Webber DL (2010) Perivascular epithelioid cell neoplasm of the colon. World J Gastrointest Oncol 2: 205-208.
8. Genevay M, Mc Kee T, Zimmer G, Cathomas G, Guillou L (2004) Digestive PEComas: a solution when the diagnosis fails to "fit". Ann Diagn Pathol 8: 367-372.
9. Folpe AL, Mentzel T, Lehr HA, Fisher C, Balzer BL, et al. (2005) Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: a clinicopathologic study of 26 cases and review of the literature. Am J Surg Pathol 29: 1558-75.
10. PL Lyle, CM Amato, JE Fitzpatrick, Robinson WA (2008) Gastrointestinal melanoma or clear cell sarcoma? Molecular evaluation of 7 cases previously diagnosed as malignant melanoma. Am J Surg Pathol 32: 858-866.
11. Torii I, Kondo T, Takuwa T, Matsumoto S, Okumura Y, et al. (2008) Perivascular epithelioid cell tumor of the rib. Virchows Arch 452: 697-702.
12. Park SJ, Han DK, Baek HJ, Chung SY, Nam JH, et al. (2010) Perivascular epithelioid cell tumor (PEComa) of the ascending colon: the implication of IFN-α2b treatment. Korean J Pediatr 53: 975-978.
13. Lee M, Cho KJ, Yu C, Park Y, Kim JC, et al. (2012) Perivascular epithelioid cell tumor of the sigmoid colon with transcription factor E3 expression. Ann Diagn Pathol 16 : 306-311.