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Gastric PEComa as a rare cause of upper gastrointestinal bleeding in pediatrics: Case report

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ABSTRACT

Perivascular epithelioid cell tumors, known as PEComas, are rare mesenchymal neoplasms with dual melanocytic and smooth muscle differentiation, whose pathophysiology and clinical behavior are still being studied. Their occurrence in the gastrointestinal tract is exceptional, particularly in children. We present the case of a 7-year-old girl who consulted for hematemesis and severe anemia. Imaging studies and digestive endoscopy revealed a gastric mass initially suspected to be a gastrointestinal stromal tumor (GIST). Surgical resection was performed, and the definitive diagnosis by pathological anatomy was gastric PEComa.

This paper aims to provide a clinical and diagnostic characterization of this extremely rare entity in pediatrics, raise awareness of its timely recognition, reflect on its differential diagnosis, and discuss its therapeutic approach.

Keywords: *perivascular epithelioid cell neoplasms; anemia; gastrointestinal bleeding; gastric neoplasms; gastrectomy.*

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INTRODUCTION

Perivascular epithelioid cell tumors (PEComas) are a heterogeneous family of rare mesenchymal neoplasms characterized by dual melanocytic and smooth muscle differentiation, with marked morphological, immunohistochemical, and genetic variability, which explains the complexity of their classification and biological behavior.[1]

Gastrointestinal tract involvement is uncommon and accounts for a minority of reported PEComas. In this location, the stomach, small intestine, and colon have been described as possible sites of origin. However, their clinical presentation is often nonspecific and frequently confused with other mesenchymal neoplasms, especially gastrointestinal stromal tumors (GISTs).[2]

Recent studies have highlighted the wide clinical variability of PEComas, which can present as incidental findings in asymptomatic patients or with severe clinical manifestations, including bleeding, severe anemia, or compressive symptoms.[3] This clinical heterogeneity contributes to diagnostic difficulty and reinforces the need to consider PEComas within the spectrum of mesenchymal tumors, even when the clinical presentation is not suggestive of malignancy.

PEComas of the gastrointestinal tract remain poorly recognized entities, with a limited number of published cases and no standardized guidelines for their diagnosis and therapy.[4]

CLINICAL CASE

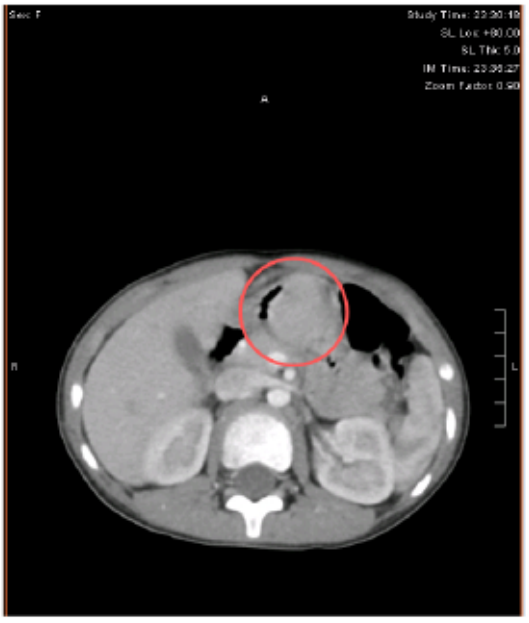
A 7-year-old girl, previously healthy, with no relevant personal or family history, consulted for colicky abdominal pain of 48 hours' evolution associated with projectile vomiting and melena. Admission to the pediatric intensive care unit was decided for diagnostic and therapeutic management. Initial laboratory tests showed severe anemia (hemoglobin: 5.1 g/dL), warranting a red blood cell transfusion.

An abdominal ultrasound revealed a solid, highly vascularized nodular image in the gastric antrum or pancreatic region. A triphasic abdominal CT scan showed a well-defined, 32 × 28 mm intraluminal polypoid mass in the gastric antrum, with a capsule and homogeneous enhancement, associated with a 10 mm adenopathy (*Figure 1*).

Upper digestive endoscopy revealed a 30 mm exophytic subepithelial lesion on the posterior wall of the antrum, with a central ulcer measuring 5-7 mm and a fibrin base (*Figure 2*).

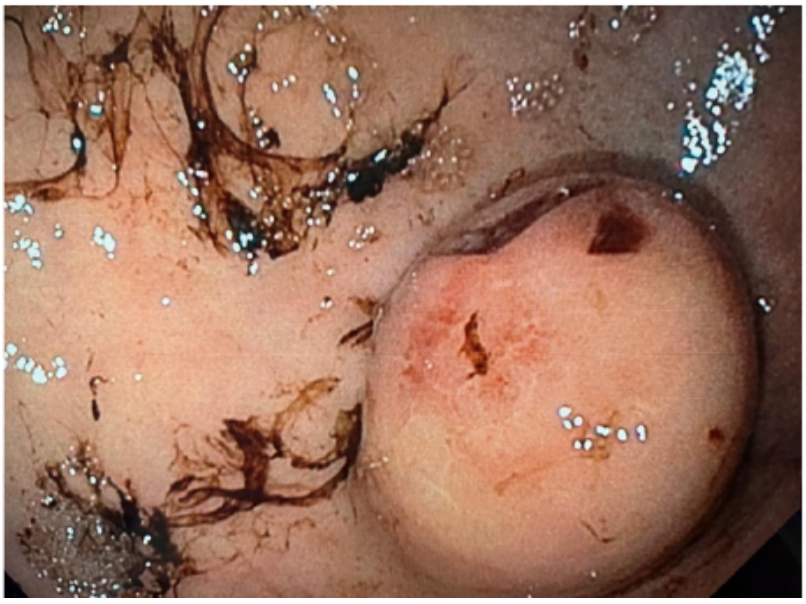
Due to refractory anemia caused by persistent bleeding, a decision was made, in conjunction with the Pediatric Surgery Department, to perform surgical resection. A partial gastrectomy was performed with resection of the tumor with free margins and a lymph node. The material was sent for pathological examination (*Figures 3* and *4*).

Figure 1. Contrast-enhanced computed tomography of the abdomen



A tumor mass is observed.

Figure 2. Endoscopic image of the stomach



An exophytic lesion with a central ulceration area is observed.

Based on the histological diagnosis and in correlation with the clinical, imaging, and macroscopic findings, a diagnosis of gastric PEComa-type myomesenchymal tumor with lymph node involvement was made.

The patient progressed favorably postoperatively and was discharged on the tenth day of hospitalization, with outpatient follow-up with the Oncology, Pediatric Surgery, and Pediatric Gastroenterology services.

Figure 3. Macroscopic image of the gastric tumor during surgery

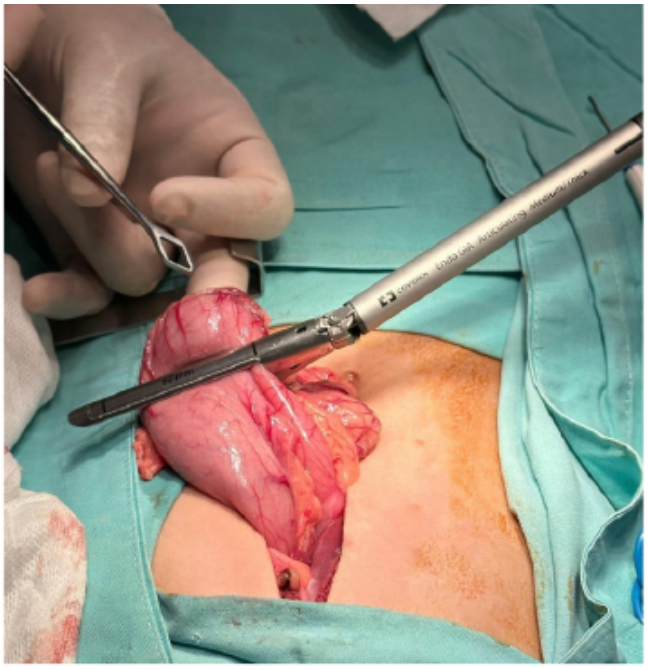
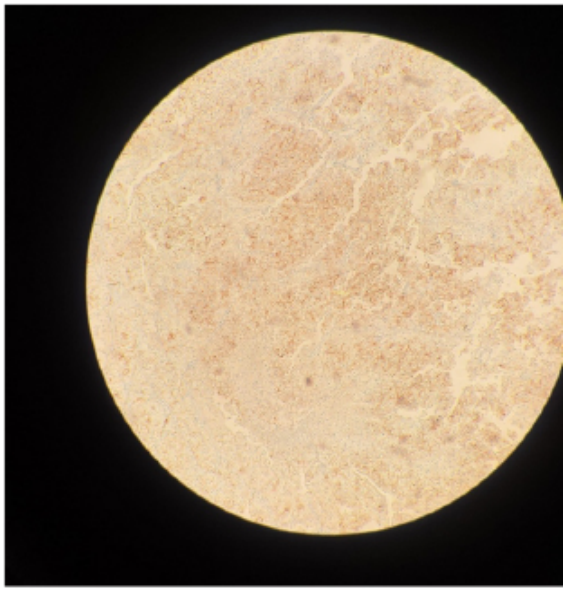


Figure 4. Immunohistochemistry performed with the Roche Benchmark automated window system, HMB-45 positive



Brown granular cytoplasmic staining in much of the tissue. Melanocytic or perivascular epithelioid differentiation, suggestive of PEComa.

DISCUSSION

PEComas are a rare group of mesenchymal neoplasms with dual melanocytic and smooth muscle differentiation, whose pathophysiology and clinical behavior remain under investigation.[1] This group includes entities such as angiomyolipoma, lymphangioleiomyomatosis, and clear cell lung tumor, but also encompasses rare tumors of various locations, including the gastrointestinal tract, liver, uterus, and soft tissues.

Likewise, PEComas diagnosed incidentally in extragastrointestinal locations, such as the lung, have been reported, reinforcing the clinical heterogeneity and broad spectrum of presentation of these rare tumors.[5]

In the largest clinical-pathological series published to date, Hammer et al. analyzed 70 cases of PEComa family tumors in children, adolescents, and young adults, demonstrating marked clinical heterogeneity, with variable anatomical locations and diverse biological behavior patterns.[6]

In pediatrics, PEComas are extremely rare, and only a third of documented cases involve the gastrointestinal tract, with gastric localization even rarer.[2] These tumors often pose a significant diagnostic challenge due to their morphological and immunohistochemical variability, overlapping with other pediatric mesenchymal neoplasms such as GISTs, clear cell sarcomas, or malignant gastrointestinal neuroectodermal tumors.[1] This confusion is common, as the clinical presentation and initial findings may be indistinguishable from other, more common mesenchymal neoplasms in this location. In fact, other reports also document the initial suspicion of GIST in cases of PEComas in the gastrointestinal tract, particularly in rectal and gastric presentations, where clinical, endoscopic, and imaging similarities with gastrointestinal stromal tumors can make preoperative diagnosis difficult.[3]

Despite being an unusual location, the stomach is the fourth most common site for gastrointestinal PEComas. In this context, complementary techniques such as immunohistochemistry and molecular studies are essential for reaching an accurate diagnosis.

From a histological point of view, PEComas are characterized by a population of epithelioid or spindle cells arranged in fascicles or nests around vessels, with positivity for melanocytic markers such as HMB-45, Melan-A, and MiTF, and occasional smooth muscle actin expression.[1] In childhood, unlike in adults, there is no clear predilection for females, and some cases have been associated with other pediatric malignancies

such as neuroblastoma and leukemia.[2] In addition, a recent study in the young population has identified a high prevalence of tuberous sclerosis (TS) and an association of unspecified PEComas (PEComa-NOS) with *TFE3* rearrangements in pediatric and young adult patients without TS. These molecular findings highlight the need for comprehensive follow-up and characterization.

The therapeutic management of localized PEComa is complete surgical resection with negative margins. Our case required a partial gastrectomy due to refractory anemia caused by persistent bleeding; tumor resection with clear margins and lymph node removal were achieved. The prognosis is closely linked to the possibility of complete resection. The lymph node involvement observed in the pathological anatomy should be carefully considered in oncological follow-up. The current literature continues to explore treatment guidelines for malignant forms and the role of adjuvant therapies.

In this case, the clinical picture and initial findings were indistinguishable from those observed in a gastrointestinal stromal tumor (GIST), highlighting the diagnostic difficulty and the potential for confusion with other, more common neoplasms. The fundamental importance of histopathological analysis and immunohistochemical studies as key tools for establishing a definitive diagnosis and appropriately guiding therapeutic management is highlighted. ■

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