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Perivascular epithelioid cell tumor of the ascending colon mesentery in a child: case report and review of the literature

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Abstract Perivascular epithelioid cell tumor (PEComa) is a rare mesenchymal tumor. Perivascular epithelioid cell tumors of the gastrointestinal tract are very rare, with only about 20 previous reported cases. We present a 5.5-year-old boy with PEComa of the right colon. Treatment consisted of tumor resection only, without additional adjuvant therapy. Two years after surgery, he remains free of tumor. To the best of our knowledge, this is the youngest reported child with PEComa of the colon. We review the literature concerning PEComas in children, especially those of the gastrointestinal tract. We emphasize the importance of correct immunohistochemistry diagnosis, recommended treatment, and surveillance of this unique family of tumors.

Primary malignant tumors of the gastrointestinal (GI) tract are very rare in children. It is estimated that they account for 1.2% of pediatric malignancies [1]. Recent development in immunohistochemistry allows for the classification of new tumors that could not be defined precisely in the past. Perivascular epithelioid cell tumors (PEComas) are an example of such a recently defined family of rare tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells (PECs). These tumors have been reported in various locations in children. Perivascular epithelioid cell tumors are generally low-grade malignancies; however, there are reports of more aggressive behavior. Because of their relative rarity, little is known about the natural history and prognostic factors of PEComas. We report a very young child with PEComa arising at the right colon and discuss the recommended treatment modalities and follow-up of this rare mesenchymal GI tract tumor.

1. Case report

A 5.5-year-old boy was admitted to the emergency department complaining of abdominal pain and fever. Prior medical history and development were normal. Four days before admission, he developed abdominal pain, vomiting,

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and fever up to 38°C. On admission, the child appeared deeply suffering. Vital signs were normal (temperature: 36.6°C, pulse: 111 beats per minute, blood pressure: 110/60 mm Hg). Physical examination was remarkable for right abdominal tenderness and rigidity, without hepatosplenomegaly. There was no skin rash or hypopigmentation. Laboratory examinations showed moderate leukocytosis of 14,500 white blood cells per cubic millimeter and elevated C-reactive protein of 13.5 mg% (reference range: 0-1 mg%).

Abdominal radiography revealed a right midabdominal indiscrete lobular opacity and mild distension of an adjacent bowel loop, suggestive of partial bowel obstruction. Abdominal sonography showed free fluid and enlarged mesenteric lymph nodes; the appendix was not visualized. Contrast-enhanced abdominal computed tomography (CT) was suspicious for a mass within the bowel that seemed to cause an intussusception (Fig. 1).

The child was taken to the operating room. At laparotomy, there was clear free abdominal fluid, without evidence of infection. The appendix and the ileum were normal. Upon palpation, a soft tissue mass was found in the mesentery of the right colon, extending into the bowel lumen (Fig. 2). Resection of the ascending colon and anastomosis of the ileum to the transverse colon were performed. The postoperative course was uneventful.

Upon opening the lumen of the right colon, a 5-cm ulcerating, polypoid lesion was found. Macroscopically, the mass consisted of gray-white firm tissue that invaded all layers of the bowel wall into the serosa and the surrounding pericolic fat. Microscopic examination revealed a spindle cell sarcoma-like tumor. On immunohistochemical staining, the tumor cells were strongly positive for HMB-45 (Fig. 3). They were also positive for desmin and vimentin, and were negative for CK, EMA, CD21, CD68, myogenin, actin, C-kit, CD34, MART-1, PDGFR, and S-100. The histology and immunohistochemical staining were consistent with a malignant form of perivascular epithelioid cell neoplasm (PEComa). Twenty-four mesenteric lymph nodes were negative for metastases, and the resection margins were clear of tumor.

Results of postoperative evaluation including chest and abdominal CT and bone scan were negative. The child did

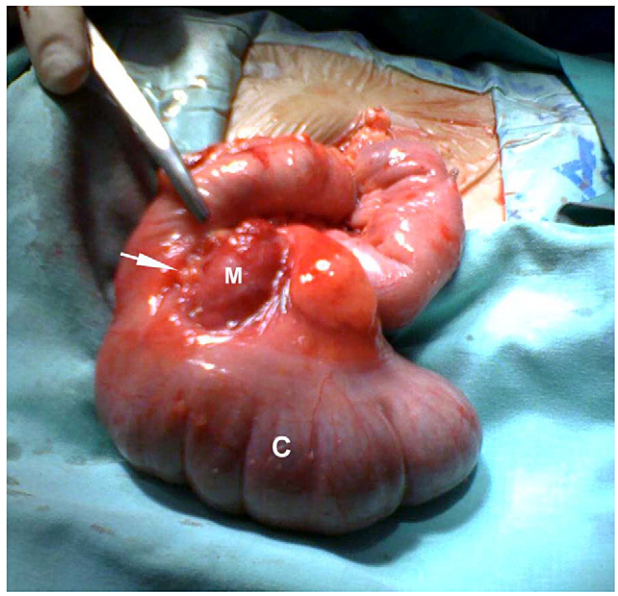

Fig. 2 Intraoperative picture showing a mesenteric mass (M), extending into and expanding the lumen of the right colon (C). One of several adjacent mesenteric lymph nodes is also demonstrated (arrow).

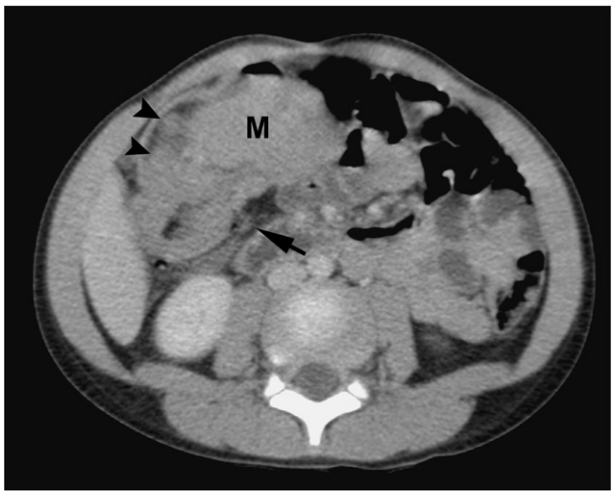

Fig. 1 Contrast-enhanced CT demonstrating a mass within the intestinal lumen (M). The mass cannot be differentiated from the medial bowel loop wall and seems to cause an intussusception. Moderate infiltration of mesenteric fat is present in the inferomedial aspect of the intussuscepted bowel loop (arrow), and small lymph nodes are seen in its lateral aspect (arrowheads).

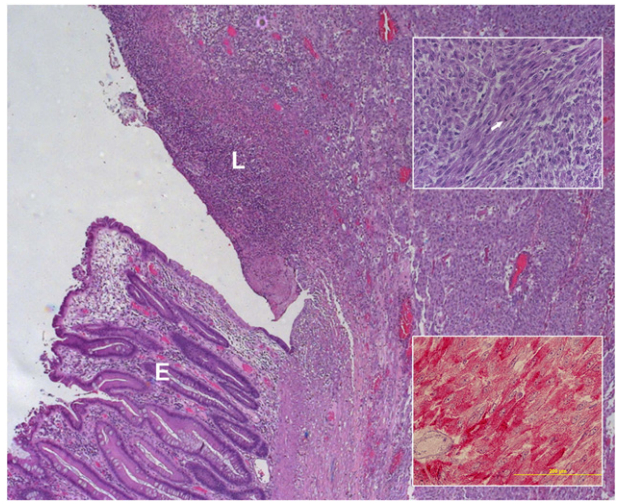

Fig. 3 Low-power view of ulcerating lesion in colon (L), adjacent to colonic epithelium (E). Upper insert: high-power view (original magnification ×40) with mitosis (arrow); lower insert: high-power view (original magnification ×40). Immunohistochemical staining of lesion with HMB-45.