

Epithelioid hemangioendothelioma as a cause of intestinal intussusception in an adult female patient: case report

Hemangioendotelioma epiteliode como causa de intususcepción intestinal en una paciente adulta: caso clínico

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Abstract

Introduction: Epithelioid hemangioendothelioma is a rare vascular tumor with an epithelioid and histiocytoid appearance. Intestinal intussusception can manifest as chronic abdominal pain, representing only 1-5% of intestinal obstructions in adults.

Case report: 65-year-old female who is attended with chronic abdominal pain. We performed a computed tomography showing the incursion of the ileum into the right colon. She was taken to the operating table, with the finding of ileo-colic intestinal intussusception due to small bowel tumor, with subsequent anatomopathological results of epithelioid hemangioendothelioma. **Conclusions:** The diagnosis and management process with an appropriate postoperative outcome is described.

Keywords: Hemangioendothelioma. Vascular tissue neoplasm. Abdominal pain. Intussusception. Intestinal invagination.

Resumen

Introducción: El hemangioendotelioma epiteliode es un tumor vascular poco frecuente de aspecto epiteliode e histiocitoide. La intususcepción intestinal suele ser causa de dolor abdominal crónico y corresponde al 1-5% de las obstrucciones intestinales en el adulto. **Caso clínico:** Mujer de 65 años que acude con dolor abdominal crónico. Se realiza tomografía computarizada y se observa incursión del íleon en el colon derecho. Es llevada a mesa operatoria con hallazgo de intususcepción intestinal ileocólica por tumoración de intestino delgado, con resultado anatomopatológico de hemangioendotelioma epiteliode. **Conclusiones:** Se describe el proceso de diagnóstico y manejo, con apropiado desenlace posoperatorio.

Palabras clave: Hemangioendotelioma. Neoplasias de tejido vascular. Dolor abdominal. Intususcepción. Invaginación intestinal.

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Introduction

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor of an epithelioid and histiocytoid appearance originating from endothelial or pre-endothelial vascular cells¹. It represents < 1% of vascular tumors and was first described in 1975 by Dail and Liebow as pulmonary EHE; the term “epithelioid hemangioendothelioma” was first introduced back in 1982 by Weiss and Enzinger to describe a vascular tumor of the bone and soft tissue with characteristics between hemangioma and angiosarcoma². The estimated prevalence is < 1 case per million inhabitants³. It is usually diagnosed in women in the 6th decade of life, showing as abdominal pain and iron deficiency anemia when located in the small intestine, and weight loss, painful hepatomegaly, portal hypertension, or platelet sequestration (Kasabach-Merritt syndrome) when located in the liver⁴.

Nonspecific abdominal pain is the most common cause of acute surgical admission for abdominal pain. Its etiology cannot be established in up to 40%-65% of patients, which can lead to a chronic course of the disease⁵. In acute abdominal pain, studies often focus on excluding conditions such as acute appendicitis, intestinal obstruction, and similar ailments; in chronic pain, the main focus is on malignant diseases, inflammatory bowel disease, acid-peptic disease, gynecological diseases, and hepatobiliary disease⁵.

Intestinal intussusception is a rare type of intestinal obstruction, defined as the introduction of a proximal intestinal loop (intussusceptum) into a distal loop (intussusciens), resulting in the obliteration of its lumen. Although it is a common cause of intestinal obstruction in children, intestinal intussusception in adults is quite rare, with an estimated incidence of 2 cases per million inhabitants per year⁶. Of all intussusception cases, only 5% occur in adults, and they represent only 1% up to 5% of all intestinal obstructions in adults⁷. The most common locations are the junctions between segments of the intestine that can move freely and the segments of the intestine that are adhered or fixed to the retroperitoneum⁸.

The clinical presentation of intestinal intussusception in adults is usually nonspecific. The classic triad of abdominal pain, vomiting, and “currant jelly” stools is rarely evident, which is more common in children, leading to delays in diagnosis. However, intestinal intussusception is an important differential diagnosis to consider because most cases in adults are caused by

structural lesions, commonly malignant neoplasms⁹. The use of computed tomography as a preoperative diagnostic method has been widely developed in recent years for this type of pathology¹⁰.

Case report

A 65-year-old woman, originally from Mérida, Venezuela, with a surgical history of open appendectomy, presents with a current illness of 4 months characterized by insidious onset and a 3-month history of mild, sharp abdominal pain, initially generalized but later localized to the right iliac fossa, for which she consulted a physician, who prescribed medical therapy with partial improvement of the condition. As symptoms persisted and a palpable mass appeared in the right iliac fossa, she visited the gastroenterology department of our center, where a colonoscopy revealed the presence of a subepithelial tumor in the cecal region.

She also presented with altered bowel habits, alternating between periods of constipation and diarrhea, recent onset of fever (15 days), and intolerance to oral intake (solids), for which she was referred to the general surgery department.

On physical examination, she was found to be in stable clinical condition, afebrile, hydrated, with adequate skin and mucosal coloration; abdominal examination revealed a flat abdomen, present bowel sounds, soft and depressible, with a palpable 5 cm × 5 cm painful mass in the right iliac fossa, without any signs of peritoneal irritation; rectal examination revealed a normal anus, preserved sphincter tone, smooth rectal walls with no palpable tumors, and scant soft stools inside; gynecological examination showed no abnormalities; the rest of the physical examination was unremarkable.

A computed tomography scan with double contrast (oral and intravenous) was requested, showing loss of the usual configuration in the ileocecal region with significant enlargement of the cecum, and the ileum was observed to protrude into the right colon along a long tract from the transverse colon to the splenic flexure (Fig. 1), where a sac-like formation filled with oral contrast was observed. During the arterial phase, the cecal region showed no changes in density, and vascular pathways were observed throughout the colon along with adipose tissue, but double enhancement was evident in the region of the splenic flexure of the transverse colon. Findings were suggestive of intussusception secondary to neoplasm.

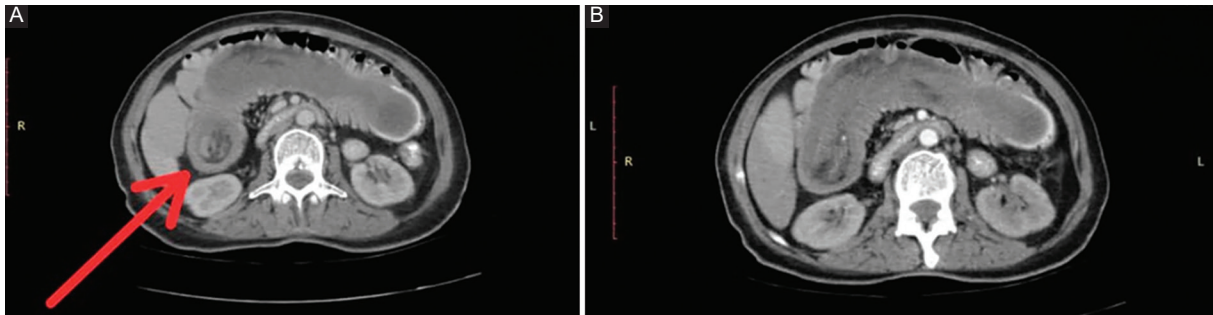


Figure 1. A: loss of usual configuration of the ileocecal region (arrow). B: incursion of the ileum into the right colon across a long tract extending from the transverse colon to the splenic flexure.

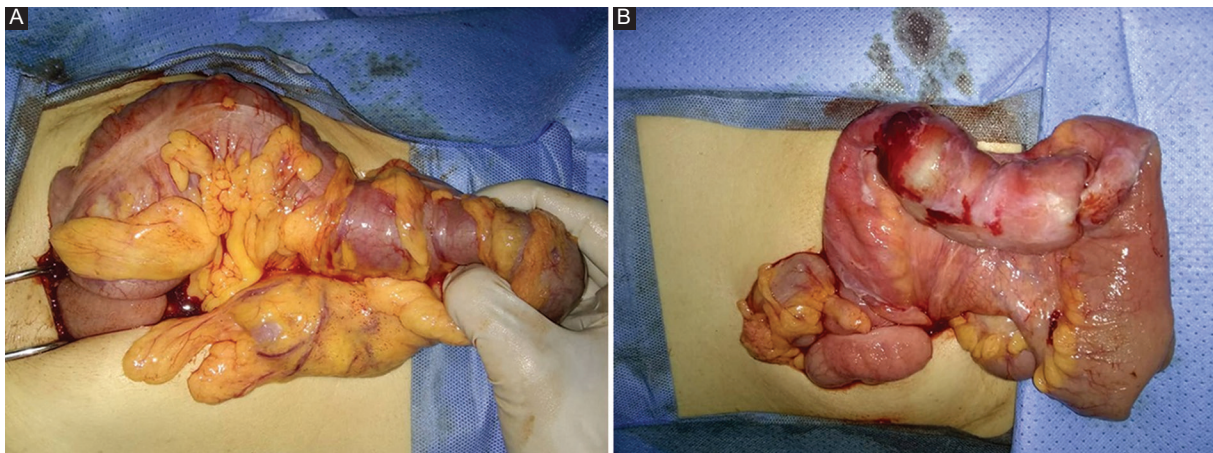


Figure 2. A: ileo-colic intussusception up to the hepatic flexure of the transverse colon. B: tumor found 10 cm away from the ileocecal valve, and 10 cm in length found in the small intestine, invading up to the indurated serosa and compromising the intestinal lumen.

Given the tomographic findings and physical examination, the patient was taken to the operating room, where a small amount of free serous fluid was found in the abdominal cavity. Ileocolic intussusception was evident up to the hepatic flexure of the transverse colon (Fig. 2A). Upon reducing the intussusception, a 10 cm tumor was observed 10 cm from the ileocecal valve in the small intestine, invading up to the serosa, indurated, and compromising the intestinal lumen (Fig. 2B), with multiple mesenteric lymph nodes of varying sizes (no > 2 cm) in the right colon mesentery, without evidence of liver lesions. The Cattell-Braasch maneuver was performed, right hemicolectomy up to the emergence of the right colic artery, closure of the transverse colon stump, and construction of a side-to-side ileotransverse anastomosis, washing and drainage of the cavity, and layered closure of the abdominal cavity. The surgical act ended uneventfully, and the patient had a favorable clinical course. Oral

intake was started 48 hours postoperatively. She was discharged 5 days postoperatively, the drain was removed at 10 days, and the patient continued follow-up in the general surgery department without any complications.

The studied sample showed a neoplastic lesion characterized by the proliferation of tubular structures with endothelial-like lining exhibiting eosinophilic cytoplasm, vesicular nuclei without nucleoli, intracytoplasmic vacuoles, presenting endothelial lumens, some with erythrocytes, and isolated mitoses (3 in 40 high-power fields). This area showed an increase in fibrous connective tissue. At the periphery, a significant polymorphonuclear neutrophil and lymphocytic inflammatory infiltrate with necrosis, cellular debris, and extravasated erythrocytes was observed. Special stains of PAS (Periodic Acid-Schiff) and Gomori's trichrome showed the presence of fibrous connective tissue around the described neoplastic lesion areas;

sections of the large intestine with mucosal lining showed no significant alterations, as did the mesentery. Findings in the small intestine were consistent with EHE of intermediate malignancy.

Discussion

Intussusception is classified, according to its location, into 4 different categories:

- Entero-enteric: confined to the small intestine.
- Colo-colonic: affects only the large intestine.
- Ileo-colic: defined as the introduction of the terminal ileum into the ascending colon.
- Ileo-cecal: the ileocecal valve is the main point of the intussusception and is somewhat difficult to distinguish from the ileo-colic variant¹¹.

We present the case of an adult patient with ileo-colic intussusception, which is very uncommon in this age group. The exact pathophysiology of intestinal intussusception (primary or idiopathic) is currently unknown in 8% up to 20% of cases, being more likely to occur in the small intestine⁷. In this case, it was due to an EHE, a very rare tumor representing only 1% up to 3% of all malignant gastrointestinal neoplasms¹², while the most common malignant tumors in the gastrointestinal tract are adenocarcinomas (40%), carcinoids (25% up to 30%), lymphomas (15% up to 20%), sarcomas (12%), and metastases¹³.

EHE is a rare and locally aggressive tumor described in the small intestine as a cause of upper GI bleeding⁴. Although we conducted an extensive literature review, we did not find any described cases as a cause of intestinal intussusception in adult or pediatric patients. There are 4 different histological types of hemangioendothelioma: epithelioid, spindle cell, Kaposiform, and malignant papillary intravascular or Dabska tumor⁴. In descending order of frequency, their location is usually the skin, liver, spleen, the GI tract (mainly the jejunum and ileum), bone, lung, and head and neck. Approximately 50% of patients with internal organ involvement present with associated skin lesions. In adults, the spindle cell and epithelioid variants are the most common, affecting one or several organs nodularly or diffusely⁴.

The patient underwent successive follow-up using other imaging methods (abdominal ultrasound and chest X-ray), without evidence of tumors in other organs or skin involvement, concluding that the localized EHE was a primary tumor affecting only 1 organ. The abdominal pain presented can be attributed to the

intussusception, which showed an atypical clinical expression.

Conclusions

Untreated intestinal intussusception can be life-threatening, causing progressive intestinal distension resulting in increased intraluminal pressure, which can lead to microvascular ischemia, tissue necrosis with intestinal perforation, and subsequently peritonitis¹⁴. Distant metastases of EHE have been described by hematogenous dissemination; they primarily invade the liver, but also the skin, serous membranes, spleen, tonsils, retroperitoneum, and kidneys, and exceptionally colonic metastases have been described¹⁵.

When lesions are small and limited in number, some authors recommend surgical resection or a preventive approach in asymptomatic patients. Although successful curative resection achieves good results, the role of adjuvant chemotherapy and radiotherapy is ambiguous. Radiotherapy is generally chosen after surgical resection of localized EHE to control residual disease, while chemotherapy is preferred in cases of disseminated disease, but its beneficial effect is not yet confirmed¹.

In this patient, early diagnosis based on presumption, coupled with more specific diagnostic imaging modalities (computed tomography) led to early surgical resolution, correcting not only the intussusception but also resecting a tumor of intermediate malignancy.

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Conflicts of interest

None declared.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

References

1. Sardaro A, Bardoscia L, Petruzzelli M, Portaluri M. Epithelioid hemangioendothelioma: an overview and update on a rare vascular tumor. *Oncol Rev*. 2014;8:82-91.
2. Rosenberg A, Agulnik M. Epithelioid hemangioendothelioma: update on diagnosis and treatment. *Curr Treat Options Oncol*. 2018;19:19.
3. Lau K, Massad M, Pollak C, Rubin C, Yeh J, Wang J, et al. Clinical patterns and outcome in epithelioid hemangioendothelioma with or without pulmonary involvement. *Chest*. 2011;140:1312-8.
4. Tortajada-Laureiro L, Poza-Cordón J, Froilán-Torres C, Gómez-Senent S, Jin Han E, Pascual-Turrión J, et al. El hemangioendotelio y otros tumores de intestino delgado como causa de hemorragia digestiva de origen oscuro. *Rev Esp Enferm Dig*. 2013;105:239-40.
5. Rathore MA, Andrabi SI, Mansha M. Adult intussusception — a surgical dilemma. *J Ayub Med Coll Abbottabad*. 2006;18:3-6.
6. Hadid T, Elazzamy H, Kafri Z. Bowel intussusception in adults: think cancer! *Case Rep Gastroenterol*. 2020;14:27-33.
7. Marinis A, Yiallourou A, Samanides L, Dafnios N, Anastasopoulos G, Vassiliou I, et al. Intussusception of the bowel in adults: a review. *World J Gastroenterol*. 2009;15:407-11.
8. Sachs M, Encke A. [Entero-enteral invagination of the small intestine in adults. A rare cause of "uncertain abdomen"]. *Langenbecks Arch Chir*. 1993;378:288-91.
9. Lu T, Chng YM. Adult intussusception. *Perm J*. 2015;19:79-81.
10. Yalamarthy S, Smith RC. Adult intussusception: case reports and review of literature. *Postgrad Med J*. 2005;81:174-7.
11. Weillbaeche D, Bolin JA, Hearn D, Ogden W. Intussusception in adults. Review of 160 cases. *Am J Surg*. 1971;121:531-5.
12. Jensen DM. Current diagnosis and treatment of severe obscure GI hemorrhage. *Gastrointest Endosc*. 2003;58:256-66.
13. Blanchard DK, Budde JM, Hatch GF, Wertheimer-Hatch L, Hatch KF, Davis GB, et al. Tumors of the small intestine. *World J Surg*. 2000;24:421-9.
14. Nkembe NM, Mbanga C, Agbor VN. Complicated idiopathic ileo-ileal intussusception in a 45-year-old male: a case report. *Int J Case Rep*. 2018;2:1-4.
15. Singly B, Pommaret E, Sailhan F, Coriat R. Colonic metastasis of an epithelioid hemangioendothelioma: an unusual cause of periumbilical pain and weight loss. *Dig Liver Dis*. 2011;43:e10.