CLINICAL CASE

EVOLUTIVE PECULIARITIES OF AN ILEO-CECAL VALVE TUMOR

Elena Neșțian¹, M. A. Șoitu¹, Rodica Bîrlă^{1,2}, Daniela Dinu^{1,2}, Cristina Iosif³, Mihaela Ungureanu⁴, S. Constantinoiu²

¹The University of Medicine and Pharmacy “Carol Davila”, Bucharest, Romania

²General and Esophageal Surgery Clinic, “Sf. Maria” Clinical Hospital, Bucharest, Romania

³Department of Pathology, “Sf. Maria” Clinical Hospital, Bucharest, Romania

⁴Department of Intensive Care Unit, “Sf. Maria” Clinical Hospital, Bucharest, Romania

Corresponding author: Rodica Bîrlă

Phone no: 0040722301289

E-mail: birlarodica@yahoo.com

Abstract

Ileo-cecal valve tumors are often clinically manifested either through transit disorders or anemic syndrome. The 88-year-old patient is admitted for abdominal pain in the left flank, vomiting, asthenia and lack of intestinal transit for feces, with a progressive onset of one week. Clinical examination: generalized affected state, wide abdomen, painful left flank and hypochondrium, lack of intestinal transit and pallor. Paraclinic: severe hypochromic microcytic anemia (hemoglobin: 5.8 g / dl). Plain radiography: hydroaeric levels in the left hypochondrium. Abdominal CT: ileo-colic intussusception that completely occupies the ascending colon to the hepatic flexure, with no visible tumoral lesions. After hydroelectrolytic and hemodynamic balancing with restoration of intestinal transit, colonoscopy is performed: 5 cm ulcero-vegetative mass at the ileo-cecal valve level - biopsy with histopathological examination: moderately differentiated adenocarcinoma. Patient refuses surgery. After 3 weeks, the patient returns to the clinic for the recurrence of digestive symptoms and emergency surgery is performed: laparotomy by median suprasubumbilical incision, finding: ileo-ceco-appendico-colic invagination up to the middle third of the transverse colon, defective attachment of ascending colon, ileum dilated with thick wall. The invagination is reduced and the tumoral mass is discovered. Right hemicolectomy with ileo-transverso-anastomosis is performed. Postoperatively, the patient shows a simple evolution with discharge on the 9th postoperative day. The ileo-colic invagination in adults is most often the appearance of a tumor located on mobile segments of the intestine. The right colon invagination is possible when there is a coalescence defect, anatomical peculiarity encountered in the presented case.

Keywords: *colic intussusception, adenocarcinoma, colic coalescence defect*

Introduction

The ileo-cecal valve is a narrow one-way passage from the small bowel to the colon. Any tumor pertaining to the valve will likely cause bowel obstruction. Due to its rarity, accurate pre-surgical diagnosis will prove challenging.

Furthermore, the patients commonly present themselves at later stages when signs of obstruction start to appear: transit disorders, diffuse abdominal pain, nausea, vomiting, diarrhea, abdominal distension and anemic syndrome due to occult tumoral bleeding. Obstruction can be caused by tumoral growth or

by its complications, such as intussusception, which can only occur on mobile intestinal segments. In up to 44% of all colonic intussusceptions in adults, the leading cause is a malignant tumor and up to 80% of colo-colic intussusceptions involve only the large bowel [1].

We report the evolutive peculiarities of an ileo-cecal valve tumor that causes recurrent intussusception on an abnormal right colon with a Toldt 1 coalescence fascia defect.

Case presentation

A female patient aged 88 with no significant past medical history was admitted for left flank pain, nausea, vomiting, absence of passage of feces and asthenia with progressive onset during the previous week. At clinical examination she displayed influenced general status, mild abdominal distension, diffuse spontaneous abdominal pain, left flank and upper quadrant pain on palpation, lack of intestinal transit, tegument and mucosal pallor. Laboratory tests discovered severe hypochromic microcytic anemia (hemoglobin: 5,8 g/dl, mean cell volume 60,1 fl, mean cell hemoglobin 16,1 pg).

Plain radiography was performed emergently and hydroaeric levels were found in the left hypochondrium. The abdominal CT scan showed an ileo-colic intussusception occupying the ascending colon up to the hepatic flexure, without any visible tumoral lesions (Figure 1).

The patient received 3 blood transfusions in order to correct the severe anemia. Her clinical evolution was favorable under conservative treatment with restoration of intestinal transit and digestive tolerance. Colonoscopy revealed a full-length paradoxically normal aspect of the large bowel; only at the site of the cecum a 5 cm ulcero-vegetative tumor originating from the inferior lip of the ileo-cecal valve was identified and biopsied. The pathology report documented a moderately differentiated adenocarcinoma (Figure 2).

Figure 1 – Imaging findings. Abdominal CT scan displaying the heterogeneous soft tissue mass represented by the ileo-colic intussusception which occupies the right iliac fossa (A) and extends as far as the hepatic flexure (C). A. The pathognomonic ‘sausage sign’ (dotted circle) generated by the ileal loop (white arrow) telescoped into the cecum (black arrow), seen longitudinally. B. The pathognomonic ‘target sign’ (dotted circle) on axial slice through the intussusception. Notice the radiolucent (dark) layer of fatty mesentery (yellow arrow) pertaining to the telescoped loop surrounded by the walls of the colon (blue arrow). C. Extension of the lesion to the hepatic flexure.

Figure 2 – Microscopic aspect. H&E stain of the biopsied specimens demonstrating moderately differentiated adenocarcinoma, with tubular and cribriform pattern, areas of intracellular mucosecretion and associated necrotic detritus. Original magnification $\times 10$. Courtesy of Cristina Iosif, MD, Department of Pathology, ‘Sf. Maria’ Clinical Hospital, Bucharest, Romania.

Surgery was proposed to remove the ileo-cecal valve adenocarcinoma, but the patient refused it and was discharged.

The patient was readmitted 3 weeks later with similar symptomatology and imaging and consented to undergo surgery. A median suprasubumbilical laparotomy was performed to approach the tumoral mass. Intraoperatively, a reducible ileo-ceco-appendico-colic intussusception as far as the middle third of the transverse colon was discovered, together with the ascending colon coalescence defect. The enlarged ileal loops displayed wall thickening and increased peristalsis. The intussusception was reduced and the cecal tumor exposed (Figure 3).

Figure 3 – Intraoperative view. A. The ileo-ceco-appendico-colic intussusception extending as far as the transverse colon. The segment distal to the occlusion is empty. B. Exposure of the ileo-cecal valve tumor after the reduction of telescoped ileal loop. C. Enlarged ileal loop with parietal edema.

Notice the viability of the segment lacking ischemic injury.

Right hemicolectomy with side-to-side ileo-transverso-anastomosis was performed. The histopathological assessment revealed tumor-free resection margins of the specimen, as well as lack of serosa or lymph nodes invasion (pT3, pN0, G2). Post-operatively, the evolution was uneventful, with discharge on the 9th postoperative day.

Discussions

Intussusception is a pathological condition in which a bowel segment, the intussusceptum, telescopes into the lumen of an adjacent bowel segment, the intussusciens, with the help of peristaltic wave. It was first reported in 1674 by Barbette of Amsterdam [2], [3] and a detailed record was presented in 1789 by John Hunter [4].

Adult intussusceptions usually have a lead point, a lesion that drags the bowel wall itself into an adjacent bowel lumen when a peristaltic wave tries to mobilize it. In contrast to the 90% idiopathic cases in the pediatric population, only 15,1% of adult intussusceptions are idiopathic [5], [6], [7].

In our particular case, the intussusception occurred as a result of an anatomical defect: the absence of Toldt 1 coalescence fascia that immobilizes the ascending colon. Cadaveric studies found that 11-22% of the subjects had a sufficiently mobile right colon to allow intussusception to occur. Associated risk factors are: prior abdominal surgery with development of adhesions, pregnancy, intestinal malrotation or other obstructive colonic lesions [8].

Another challenge is early accurate diagnosis. Pediatric intussusceptions have more specific presentations with passage of mucus and blood, either red or as clots (‘red currant jelly’ aspect), accompanied by abdominal cramps and palpable abdominal mass. In contrast, the adult forms are non-specific and vague: abdominal pain, nausea, vomiting, diarrhea, bleeding or melena and constipation [7].

Therefore, the surgeon must employ the use of imaging studies. Abdominal computed

tomography is the preferred investigation and describes the presence of a pathognomonic target or sausage shaped lesion [7], although a recent meta-analysis on 1229 patients reports only a 77,8% rate of accurate diagnosis [6].

Colonoscopy should be included in the pre-operative workup in order to localize the lesion and scan for other synchronous pathologies that might have been missed on the CT scan [7]. The majority of colonic (78,8%) and ileo-cecal (61,7%) intussusceptions are caused by primary adenocarcinoma, as in our case [6].

Due to the scarcity of reports, a standardized treatment plan does not exist and cases must be judged independently. The treatment options documented in the literature are: initial reduction followed by resection (for enteric location), primary resection without reduction (for colonic locations) and reduction only; the intermediate nature of the ileo-colic location warrants for a selective approach [6].

The decision to reduce the intussusception needs to be based on the following: ischemic lesions, enteral edema and the general viability of the bowel segment. In our case, the lesion was easily reducible and the segments involved did not display ischemic lesions. Some authors suggested that colonoscopic reduction is therefore cautioned, as there is the risk of perforation [9]. Colonoscopy was performed on our patient only after restoration of intestinal transit and digestive tolerance.

Considering that a malignant lead point can be found in 46,5% of colonic and respectively, 36,9% of ileo-colic intussusceptions, it would be unwise not to resect the lesion unless malignancy can firmly be ruled out [6]. 78,8% of the malignant tumors of the colon are primary adenocarcinoma therefore en bloc resection is recommended to avoid intraluminal seeding [10].

This case report documents a unique association of features displayed by an ileo-cecal valve adenocarcinoma. First of all, the unusual clinical presentation of the right colon tumor with obstructive signs was not the result of intraluminal occlusion, but of a complication: the intussusception, which occurred due to the coalescence defect. Secondly, the CT scan failed to identify the leading point; only colonoscopy revealed the tumoral mass. Thirdly, the intussusception had spontaneous

resolution (the patient recovered clinically and refused surgery on first admission), but recurrent behavior (she was readmitted later with similar symptomatology). Last, but not least, the telescoped loops were easily reducible intra-operatively and had not suffered ischemic lesions, limiting the extent of surgical resection and allowing a more favorable outcome.

Conclusion

Adult intussusception is more frequently related to a tumor, either malignant or benign, arising on a mobile bowel segment. The Toldt 1 fascia coalescence defect, an anatomical variation encountered in our case, permitted the right colon intussusception to occur. Due to the high incidence of malignant tumors associated with ileo-colic and colonic locations, resection ought to be performed. The association between the mobile right colon and a malignant lesion of the ileo-cecal valve (pre-operatively certified by pathological exam) leading to recurrent intussusception was the argument in deciding to perform right hemicolectomy on our patient.

References

- [1] L. K. Eisen, J. D. Cunningham and A. H. Aufses, "Intussusception in adults: institutional review," *J. Am. Coll. Surg.*, vol. 188, pp. 390–395, Apr. 1999.
- [2] D. de Moulin, "Paul Barbette, M.D.: a seventeenth-century Amsterdam author of best-selling textbooks," *Bull. Hist. Med.*, vol. 59, pp. 506-514, 1985.
- [3] P. Barbette, N. Geneva and F. Miegé, "Oeuvres chirurgiques et anatomiques," in Miegé F, Editor, 1674.
- [4] I. Noble, "Master surgeon: John Hunter," *Messner J*, p. 185, 1971.
- [5] T. Yoldas, A. C. Karaca, S. Ozturk, M. Unver, C. Caliskan and M. A. Korkut, "A rare cause of colonic obstruction "colonic intussusception": Report of two cases," *Case Rep. Surg.*, vol. 2015, pp. 1-3, Feb. 2015.
- [6] K. D. Hong, J. Kim, W. Ji and S. D. Wexner, "Adult intussusception: A systematic review and meta-analysis," *Tech. Coloproctol.*, vol. 23, pp. 315-324, Apr. 2019.
- [7] A. Wilson, G. Elias and R. Dupiton, "Adult colocolic intussusception and literature review,"

Case Rep. Gastroenterol., vol. 7, pp. 381-387, Sept. 2013.

[8] N. N. Mahmoud, J. I. S. Bleier, C. B. Aarons, E. C. Paulson, S. Shanmugan and R. D. Fry, "Colon and Rectum," in Sabiston Textbook of Surgery: The Biological Basis of Modern Surgical Practice, C. Townsend, Ed. Philadelphia: Elsevier, 2017, pp. 1336-1339.

[9] G. Yörük, K. Aksöz, Z. Buyraç, B. Ünsal, O. Nazli and N. Ekinici, "Adenocarcinoma of the ileo-

cecal valve: Report of a case," Turk. J. Gastroenterol, vol. 15, pp. 268-269, Dec. 2004.

[10] C. Balalau, N. Bacalbasa, I. Motofei, P. Banu, R. Scaunasu, S. Voiculescu, O. D. Balalau and V. D. Constantin, "Preoperative diagnostic difficulties of ileocecal valve adenocarcinoma – A case report," Archives of the Balkan Medical Union, vol. 52, pp. 117-121, Mar. 2017.