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ILEOCOLIC INTUSSUSCEPTION DUE TO LIPOMATOSIS OF THE ILEUM: A COMMON COMPLICATION OF A RARE CLINICAL ENTITY

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We report a case of intestinal ileal lipomatosis in a 56-year-old Caucasian male complicated with small bowel obstruction due to ileocolic intussusception with a lipoma serving as lead point. This rare disease is often only discovered incidentally as a consequence of mechanical complications and not well reported in the international literature, compared to intussusception due to an isolated lipoma. Computed tomography is the imaging modality of choice to depict complications of this distinct clinicopathological entity. Density measurements can confirm the fatty content and homogeneity analysis of the lesions can guide the radiologist in the differential diagnosis.

Key-word: Lipoma and lipomatosis.

Case report

A previously healthy 56-year-old male without significant medical history was referred by his general practitioner to the hospital with intermittent cramping abdominal pain and bloody stools. Apart from rebound tenderness in the right fossa the clinical examination was unremarkable. Inflammatory parameters were elevated. Colonoscopy was performed but interrupted because of intolerable pain. Additionally, Computed Tomography (CT) was carried out and revealed small bowel obstruction due to ileocolic intussusception with a low attenuating sharply demarcated mass as lead point (Fig. 1). Also, diffuse fatty infiltration of the submucosal layer and multiple other low attenuating submucosal masses protruding into the bowel lumen were seen scattered throughout the terminal ileum. The density varied between -80 and -120 Hounsfield units, in keeping with lipomatous tissue. Because of the small bowel obstruction, an urgent laparotomy was performed. Manual reduction was not possible and ileocaecal resection was performed. The postoperative course was uneventful. Macroscopical (Fig. 2) and histopathological (Fig. 3) analysis of the resected specimen confirmed the presence of numerous tumors composed of mature adipose tissue interspersed with some fibrous tissue. The diagnosis of intestinal ileal lipomatosis complicated with ileocolic intussusception was made.

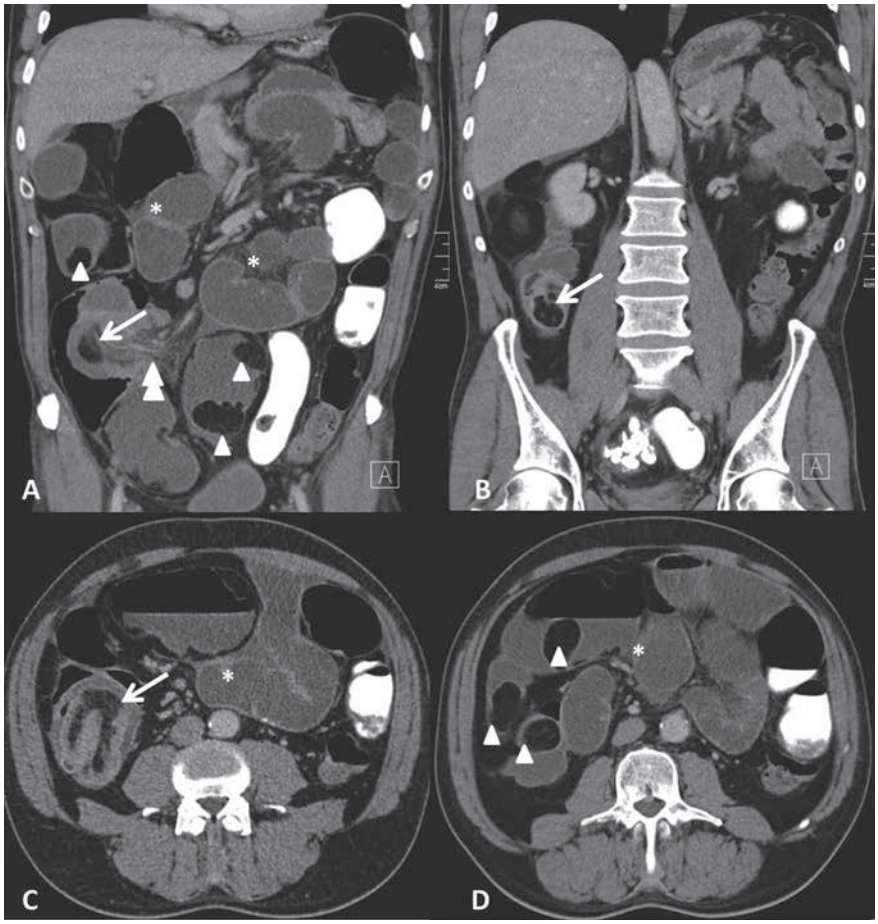

Fig. 1. — CT of the abdomen (A and B) coronal reformatted and (C and D) axial images show small bowel obstruction with dilated small bowel loops (asterisk) due to ileocolic intussusception (arrow in A and C) with infiltration of the mesenterial fat suggestive of ischemia (double arrowhead in A). A lead point could be identified (arrow in B). Also, multiple additional lipomas and diffuse submucosal fatty infiltration were seen throughout the terminal ileum (arrowheads).

Discussion

Benign tumors of the small bowel are relatively rare with lipoma being the second most common type in

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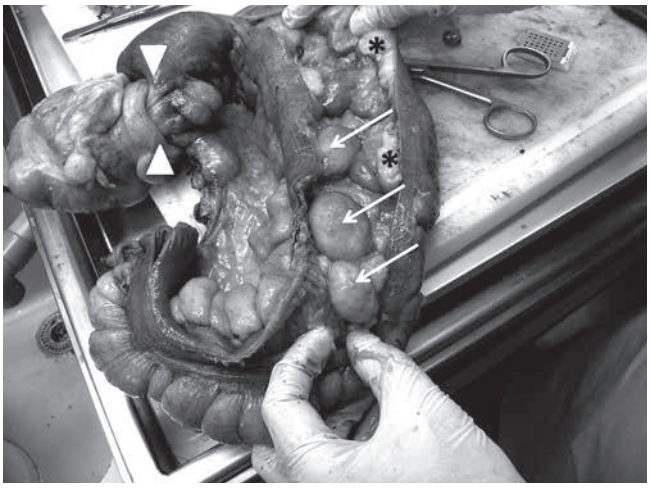

Fig. 2. — Macroscopic examination of the resected ileocaecal specimen shows intraluminal yellowish sharply delineated masses (arrows) in the terminal ileum and submucosal fatty infiltration (asterisk). Also note the ileocolic intussusception (arrowheads).

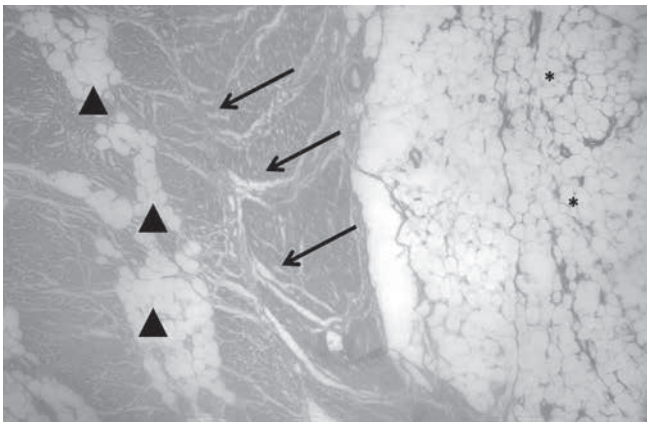

Fig. 3. — Histopathological examination of the resected specimen shows submucosal lipomatous tissue (asterisks) and muscularis propria (arrows) separated by lipocytes (arrowheads), in keeping with diffuse lipomatosis (magnification: 40x, H and E stain).

approximately 25% of the cases, only preceded by leiomyomas (1). Typical manifestations of lipomas on CT are symmetric, sharply delineated masses with a density ranging from -80 to -120 Hounsfield units compatible with fatty tissue. Solitary lipomas need to be differentiated from lipomatosis of the ileocaecal valve and intestinal lipomatosis. Lipomatosis of the ileocaecal valve involves a symmetrical fatty enlargement and is a very common finding (2). The term intestinal lipomatosis is currently used to describe diffuse infiltration and overgrowth of well differentiated fatty tissue in the submucosal layer. On histopathological examination, proliferation of adipose cells may extend to the serosal layer or mesenteric fatty tissue (3). The ileum is the most common site involved.

Intestinal lipomatosis was first described in 1906 by Hellström (4). The diagnostic criteria used nowadays are not clearly defined and range from a minimum of 4 to more than 100 lipomas (5). The etiology has not yet been elucidated. Some patients with intestinal lipomatosis have a family history, incriminating an inherited tendency. Association with hypercholesterolemia (1) or with macrodactylia fibrolipomatosis (6) has also been suggested. According to Climie et al, intestinal lipomatosis might be of hamartomatous origin since neuronal bundles were observed in the lipomas of their series (7), although this was not the case in our patient. There seems to be no gender predilection. Less than 50% of the reported cases were symptomatic with symptoms attributed to obstruction, intussusception, volvulus or (occult) bleeding (8, 9). When encountered incidentally, no treatment is necessary (3, 10). This rare clinical entity is not well reported in the international literature (11-13), compared to intussusception due to an isolated lipoma. Imaging modality of choice is CT since density measurements can confirm the lipomatous nature of the luminal masses. CT is also capable of differentiating lipoma from liposarcoma when evaluating the homogeneity of the fatty content and absence of increased density (3). In some cases – however – loss of fat density of lipomas may mimic malignancy (14).

The differential diagnosis of multiple intestinal lipomas includes intestinal manifestations of Proteus syndrome, neurofibromatosis, lymphoma, mesenteric masses or metastases (3). Proteus syndrome is caused by a germline mutation of the tumor suppressor gene *PTEN*, and is characterized by vascular malformations, lipomas, hyperpigmentation and cutaneous nevi (15). Some specific histopathological features of the fatty masses discriminate these lesions from the lipomas encountered in intestinal lipomatosis (5). Neurofibromatosis, lymphoma and metastases often comprise other more specific clinical features that help in the differential diagnosis.

Conclusion

Intestinal lipomatosis is a rare clinicopathological condition in which multiple lipomas are seen in the bowel wall. This condition is often detected as a consequence of mechanical complications, such as obstruction, intussusception or volvulus, but can also be discovered incidentally. CT is the imaging modality of choice to depict the complications and may contribute to the differential diagnosis since density measurements of the masses can demonstrate their lipomatous origin.

References

1. Yakabe S., Muranaka T., Sumii T., Takeshita M., Yamashita T., Tsuruta S., Saku M.,Yoshida K.: Jejunal lipomatosis with diverticulosis: report of a case. *Surg Today*, 1998, 28: 846-849.

2. Hoeffel C., Crema M.D., Belkacem A., Azizi L., Lewin M., Arrive L., Tubiana J.M.: Multi-detector row CT: spectrum of diseases involving the ileocecal area. *Radiographics*, 2006, 26: 1373-1390.
3. Chen Y.L., Huang T.Y., Chang W.C.: Lipomatosis of the small intestine. *Clin Res Hepatol Gastroenterol*, 2012, May 1, [Epub ahead of print].
4. Hellström N.: Kasuistische Beiträge zur Kenntnis des Intestinallipoms. *Dtsch Z Chir*, 1906, 84: 488-511.
5. Liu X., Wilcox C.M., Nodit L., Lazenby A.J.: Multiple gastrointestinal stromal tumors and lipomatosis. *Arch Pathol Lab Med*, 2008, 132: 1825-1829.
6. Mazziotti S., Salamone I., Vinci S., Pandolfo A.: Macrodactylia fibrolipomatosis associated with multiple small-bowel lipomas. *AJR Am J Roentgenol*, 2006, 186: 1195-1196.
7. Climie A.R., Wylin R.F.: Small-intestinal lipomatosis. *Arch Pathol Lab Med*, 1981, 105: 40-42.
8. Purohit V., Joshi R., Kalla A.R., Purohit R.C.: Colo-colic intussusception due to intestinal lipomatosis. *Indian J Gastroenterol*, 2003, 22: 151-152.
9. Tani T., Abe H., Tsukada H., Kodama M.: Lipomatosis of the ileum with volvulus: report of a case. *Surg Today*, 1998, 28: 640-642.
10. Ormson M.J., Stephens D.H., Carlson H.C.: CT recognition of intestinal lipomatosis. *AJR Am J Roentgenol*, 1985, 144: 313-314.
11. Kalund Jensen H., Knudsen N.: [Jejuno-ileal invagination caused by multiple lipomas]. *Ugeskr Laeger*, 1966, 128: 1347-1348.
12. Rivera Irigoin R., Fernandez Moreno N., Funez Liebana R., Medina Cano F., Navarro Sanchis E., Moreno Mejia P.P., Garcia Fernandez G., Navarro Jarabo J.M., de Sola Earle C.M., Fernandez Perez F.J., Moreno Platero J.J., Sanchez Cantos A.: [Ileo-ileal and ileocecal invagination due to intestinal lipomatosis]. *Gastroenterol Hepatol*, 2003, 26: 482-484.
13. Scotti M., Castrati G., Arrigoni G., Fiorone E.: [Intestinal invagination caused by multiple lipomas of the small intestine]. *Minerva Chir*, 1989, 44: 1405-1407.
14. Thompson W.M.: Imaging and findings of lipomas of the gastrointestinal tract. *AJR Am J Roentgenol*, 2005, 184: 1163-1171.
15. Vanhoenacker F.M., De Beuckeleer L.H., Deprettere A., De Moor A., De Schepper A.M.: Proteus syndrome: MRI characteristics of plantar cerebriform hyperplasia. *Skeletal Radiol*, 2000, 29: 101-103.