CASE REPORT

Ileocecal lipohyperplasia presenting as a chronic sideropenic anaemia

Hytham K S Hamid, Iftikhar Ahmed, Ahmed Mohamed, Tim O'Hanrahan

Department of Surgery, Sligo General Hospital, Sligo, Ireland

Correspondence to
Iftikhar Ahmed,
Iftikhar1.Ahmed@hse.ie

SUMMARY

Ileocecal lipohyperplasia is a relatively uncommon pathological entity characterised by submucosal infiltration of adipose tissue in the ileocecal valve (ICV). The majority of patients are asymptomatic, whereas, others present with serious complications. We report a 45-year-old man who presented with recurrent episodes of melena with secondary sideropenic anaemia for 6 years. Extensive radiological and endoscopic workup failed to detect the source of bleeding. During his last admission for abdominal pain, contrast-enhanced CT scan revealed a fatty mass at the ICV. The patient underwent limited right hemicolectomy, and histology confirmed ileocecal lipohyperplasia. The patient remained well 18 months later. This case highlights the difficulties encountered by clinicians in diagnosing this unusual colonic pathology.

BACKGROUND

Although formerly considered a rarity, lipohyperplasia of the ileocecal valve (ICV) is being reported with increasing frequency in the literature. Its incidence varies remarkably in published reports, ranging from less than 1% in clinical material to more than 13% in necroptic studies.¹⁻² The condition is most often clinically silent, and diagnosed as an incidental finding. At times, however, it may lead to debilitating and potentially fatal complications.³⁻⁴ Moreover, the lesions can clinically and radiologically mimic malignant neoplasms. We herein present a rare case of lipohyperplasia of the ICV, presented initially as chronic sideropenic anaemia. The clinical course and management are discussed with review of the pertinent literature.

CASE PRESENTATION

A 45-year-old man with a medical history of appendectomy and alcohol abuse, presented initially to gastroenterologists in February 2004 with exertional dyspnoea and easy fatigability. Laboratory tests showed hypochromic microcytic anaemia with haemoglobin of 6.4 g/dL. Gastroscopy and colonoscopy were reported normal, and a barium enema showed colonic and ileal diverticular disease and mild prominence of the ICV. The patient was transfused and his symptoms abated. Over a period of 4 years, his condition recurred three times with occasional melena. Further investigations including repeated colonoscopy, capsule endoscopy, isotope Meckel's scan and barium follow though did not show significant findings. At this stage a diagnosis of diverticular disease was considered as the cause of symptoms.

However, the patient remained clinically well with stable haemoglobin for the following 2 years, and no intervention was required.

In May 2010, he was admitted with severe right lower quadrant pain that had recurred periodically during the last 2 months. Physical examination was remarkable for pallor and moderate tenderness in the right iliac fossa. Laboratory tests disclosed haemoglobin 8.7 g/dL, mean cell volume 80 fL, mean cell haemoglobin 25.4 pg. Both carcinoembryonic antigen and carbohydrate antigen 19.9 were within normal range. Abdominal ultrasound was normal. Contrast-enhanced CT scan of the abdomen showed markedly prominent ICV with a fatty mass protruding into the caecum (figure 1). Colonoscopy demonstrated caecal diverticulae and a semipedunculated ileocaecal polypoid mass that could be partially reduced into the ileum upon air insufflation. The mass surface was nodular with multiple ulcerations and signs of recent bleeding (figure 2A,B). Biopsy revealed ulcerated mucosa with acute and chronic inflammation in the lamina propria, focal cryptitis and mucin depletion of the glands, consistent with a prolapsed mucosa.

In light of his recurrent symptoms, surgical intervention was deemed necessary. Limited right hemicolectomy was successfully performed. Gross pathology of the specimen showed a well-circumscribed polypoid mass at the ICV measuring 5×4×4 cm (figure 3). Upon histological examination, the surface mucosa showed acute and chronic inflammation with focal ulceration, and submucosal adipose tissue infiltration without

Figure 1 Contrast-enhanced CT scan showing prominent ileocecal valve with a fatty mass protruding into an empty caecum.

To cite: Hamid HKS, Ahmed I, Mohamed A, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-008052

Figure 2 Photographs showing a large smooth ileocecal lesion (A) that partially reduces into the ileum upon air insufflations (B), with signs of a recent bleeding.

encapsulation. The ileal and distal colonic mucosa showed no focal abnormalities.

OUTCOME AND FOLLOW-UP

The patient remained symptom free 2 years later.

DISCUSSION

The term lipomatosis of the ICV was first coined by Kelby in 1948, to denote a localised massive accumulation of

Figure 3 The surgical specimen showing ulcerated ileocecal mass involving both the upper and lower lips of the valve.

unencapsulated submucosal fat in the ileocecal region.⁵ Subsequently, Zettergren described the term “lipohyperplasia” as more appropriate to distinguish it from true neoplastic lesions.⁶ The precise aetiopathogenesis of this condition remains enigmatic. Initial mucosal prolapse, possibly due to faulty peristalsis, has been proposed as the precursor lesion for lipohyperplasia of the ICV. This creates a tissue vacuum and results in local ischaemia with consequent reactive lay down of fat.^{7 8} ICV lipohyperplasia has also been linked to obesity and metabolic disorders, representing a local manifestation of generalised conditions.² Other associations are abdominal operations and non-specific inflammatory conditions.⁹

Lipohyperplasia of the ICV usually presents in the sixth and the seventh decades of life, with females being affected more than males. The condition is often asymptomatic, and the diagnosis is quite frequently made accidentally, when a radiological examination is made. When symptomatic, ICV lipohyperplasia typically presents with common symptoms complex including right-sided abdominal pain, distension and tenderness in the right lower quadrant of the abdomen, collectively known as Bauhin’s or ICV syndrome.¹⁰ On rare occasions, patients can present dramatically with complications such as appendicitis and intestinal obstruction.^{3 11} Bleeding per rectum is an uncommon manifestation of ICV lipohyperplasia, and it is often attributed to anticoagulant therapy or other concomitant pathology.^{4 12}

Traditionally, preoperative diagnosis of ICV lipohyperplasia has been challenging because of the limitations of diagnostic methods used, the ambiguous clinical presentation of the condition and the inherent variable morphological characteristics of the ICV. The appearance of ICV lipohyperplasia on barium enema is non-specific, presenting as a well-demarcated prominent ICV. Similar imaging findings are encountered in benign and malignant ICV tumours, ileal mucosal prolapse and oedema, and inflammatory lesions.¹³ Colonoscopy is a useful adjunct to evaluate abnormally looking ICV and to rule out malignant disease. Nevertheless, its value in diagnosing ICV lipohyperplasia is limited by the subjective interpretation of findings and the low diagnostic yield of endoscopic biopsies given the submucosal location of the lesions.¹⁴ Recently, Yamamoto and colleagues have classified ICV lipohyperplasia into three types; localised, intermediate and diffuse, based on endoscopic features. Of note, the symptoms were independent of the size of the lesion, which may add to the difficulty of diagnosis.¹ The advent of two-dimensional CT scan has enabled non-invasive evaluation of the ICV and more precise diagnosis of colonic fatty lesions.¹⁵ The findings of submucosal fats should however be interpreted cautiously in conjunction with the endoscopic results and clinical symptoms, as the mere presence of submucosal fatty infiltration of the valve is considered a normal variant.

There are no firm guidelines in the literature with regard to the management of ICV lipohyperplasia, perhaps due to the relative rarity of the condition. Intervention is generally indicated in cases with unremitting symptoms or acute complications. Endoscopic resection has been advocated by some authors for lesions localised to one lip of the valve, with possible risk of perforation and haemorrhage.¹ Di Rienzo *et al*¹⁶ have recently described resection of the lesion with refashioning of the valve though a caecostomy, combined with ileocecal fixation. Limited right hemicolectomy remains the most preferable treatment option for symptomatic ICV lipohyperplasia.¹³ In our patient, the lack of preoperative tissue diagnosis and the presence of more than one demonstrable lesion that is capable of bleeding have prompted limited ileal and caecal resection.

Learning points

- ▶ Despite its infrequent occurrence, ileocecal valve lipohyperplasia should be considered as a possible cause in patients with obscure gastrointestinal bleeding.
- ▶ CT scan remains the gold standard method for diagnosis.
- ▶ Both endoscopic and surgical approaches can be used for treatment of this condition with satisfactory outcomes.

Contributors All authors contributed and agreed to the final manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Yamamoto H, Kusugami K, Ando T, *et al.* Endoscopic features of lipohyperplasia in the large intestine. *Dig Endosc* 1996;8:281–6.
- 2 Tawfik OW, McGregor DH. Lipohyperplasia of the ileocecal valve. *Am J Gastroenterol* 1992;87:82–7.
- 3 Walke L, Christie AJ. Lipohyperplasia of ileocecal valve, causing recurrent intussusception. *Henry Ford Hosp Med J* 1990;38:259–61.
- 4 Seabrook DB, Stevens R, Scholl V, *et al.* Lipomatosis of the ileocecal region. *Am J Surg* 1956;92:214–21.
- 5 Kelby GM. Submucous lipomas of the ileocecal valve. *Lancet* 1948;68:301–4.
- 6 Zettergren L. Lipohyperplasia of the ileocolic valve. *Acta Soc Med Upsal* 1953;59:61–8.
- 7 Hulten J. Lipomatosis of the ileocecal valve. *Acta Chir Scand* 1965;192:104–11.
- 8 Cabaud GP, Harris LT. Lipomatosis of the ileocecal valve. *Ann Surg* 1959;150:1092–8.
- 9 Boquist L, Bergdahl L, Andersson A. Lipomatosis of the ileocecal valve. *Cancer* 1972;29:136–40.
- 10 Lasser E, Rigler L. Ileocecal valve syndrome. *Gastroenterology* 1955;28:1.
- 11 Smith SR, Fenton L. Lipohyperplasia of the ileo-caecal valve causing appendicitis. *Aust N Z J Surg* 2000;70:76–7.
- 12 Ryan J, Martin JE, Pollock DJ. Fatty tumours of the large intestine: a clinicopathological review of 13 cases. *Br J Surg* 1989;76:793–6.
- 13 Skaane P, Eide TJ, Westgaard T, *et al.* Lipomatosis and true lipomas of the ileocecal valve. *Rofo* 1981;135:663–8.
- 14 De Beer RA, Shinya H. Colonic lipomas. An endoscopic analysis. *Gastrointest Endosc* 1975;22:90–1.
- 15 Jelbert A, Swinson S, Atkin K, *et al.* Imaging of the ileocaecal valve. *Tech Coloproctol* 2008;12:87–92.
- 16 Di Rienzo M, Mascitelli E, Angelucci D, *et al.* Surgical conservative treatment for Bauhin's syndrome. *Ann Ital Chir* 2002;73:189–95.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow