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Case Report

A case of esophageal crohn disease presenting as upper gastrointestinal hemorrhage

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Abstract: Crohn disease is a chronic inflammatory bowel disease which rarely affects esophagus. Early diagnosis, differential diagnosis and treatment of atypia CD are of great significance. Here we report a case of 41-year-old Chinese man who was admitted and presented with massive hematemesis as much as some 600 ml. His primary diagnosis was multiple esophageal ulcers accompany with bleeding. After detailed laboratory examination, gastrointestinal endoscopy, pathology examination, radiological investigations as well as clinical evidence, we further take esophageal Crohn disease into consideration. According to existing European consensus on management of CD, the patient was treated with prednisone, salazosulfapyridine and omeprazole. After systematic treatment and follow-up gastroscopy, the lesions have disappeared and the disease is in remission. As optimal treatment of esophageal CD has not been established, we hope this case may widen the thinking and strategy of diagnosis and treatment.

Keywords: Crohn disease, esophagus, hemorrhage, endoscopy

Introduction

Crohn disease (CD), an idiopathic and chronic relapsing immunologically mediated disease, arises from an interaction of genetics, environmental factors and gut microbiota [1, 2]. As one type of chronic inflammatory bowel disease, CD mostly affects ileum and colon. The incidence of CD is estimated to be 29.3 per 100 000 in Australia, 20.2 per 100000 in Canada and 10.6 per 100000 in northern Europe [3]. The incidence is lower in Asia, accounting for 0.54 per 100000 [4]. Additionally, the proximal CD, especially isolated esophageal CD, is rather rare [5, 6]. Here we report a case of esophageal CD in a patient who presented as upper gastrointestinal hemorrhage and was successfully treated with prednisone, salazosulfapyridine and omeprazole.

Case presentation

A 41-year-old Chinese man was admitted and presented with massive hematemesis as much as 600 ml, subsequent melena lasting for 4 days, retrosternal unwell felling, dizziness and abdominal distention. He has a history of recurrent aphthous ulcer, shapeless and frequent stool and arthralgia in recent 3 years. Furthermore, he has family histories of ulcerative colitis. Physical examination showed multiple aphthous ulcers in the mouth mucosa. Immediate gastroscopy revealed 3 ulcers in the esophagus, located 28, 37, 40 cm respectively from the upper incisors, measuring 1.0 × 1.0 (cm) (**Figure 1**). In addition, the surface of ulcer located 40 cm from the upper incisors is covered with a red scab and the primary diagnosis is multiple esophageal ulcers accompany with bleeding. For the further diagnosis, an upper endoscopy, biopsy, colonoscopy and chest CT and other relevant laboratory investigations were approached. The gastroscopy showed heterogeneous punch-out ulcers and a mucosa prominence in the esophageal mucosa with peripheral mucosa congestive and hydropic (**Figure 2**). In the lower esophagus there existed elongate longitudinal erosion which revealed a cobblestone-like appearance and accounted for one quarter of the circumference. The histological examination of biopsy specimens from lesions margin indicated moderate acute and chronic inflammation accompanied with non-caseous granulomas (**Figure 3**).

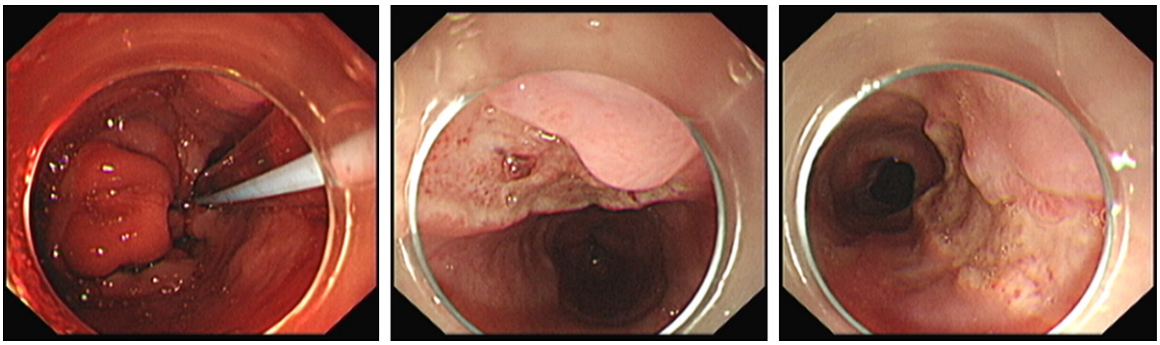

Figure 1. Gastroscopy view of the esophagus. Pictures revealed active bleeding and longitudinal ulcers.

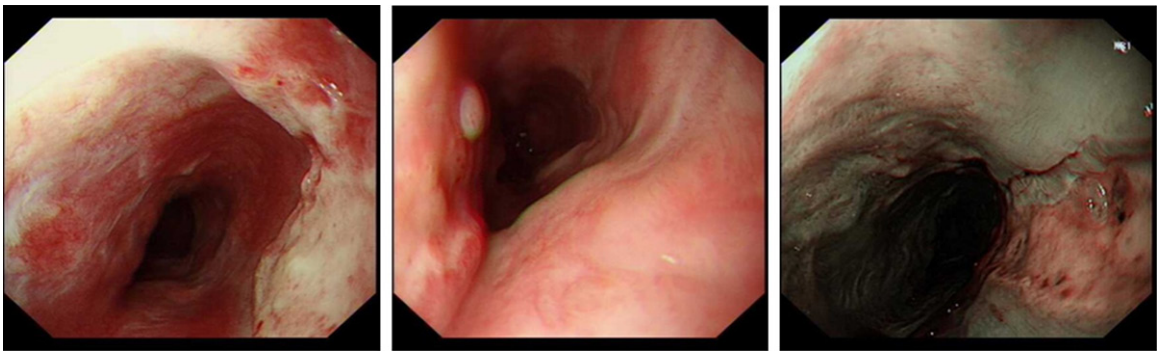

Figure 2. Gastroscopy view of the esophagus. Pictures showed heterogeneous punch-out ulcers and a mucosa prominence with peripheral mucosa congestive and hydropic.

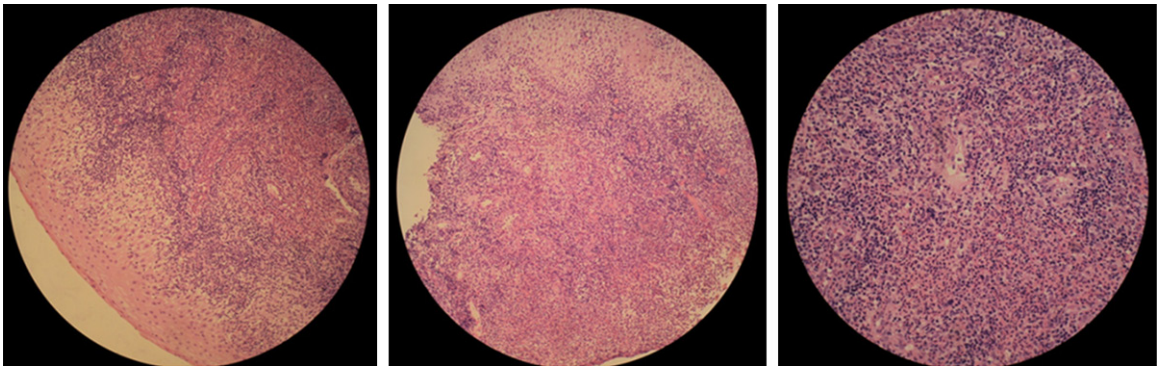

Figure 3. Pathology examination revealed moderate acute and chronic inflammation accompanied with non-caseous granulomas. HE staining.

Immunohistochemical tests showed CK516 and CD20 are positive while P53, Ki67 and CD147 were negative. Chest CT showed that the esophageal wall is slightly thickened (**Figure 4**). Colonoscopy of distal ileum demonstrated that enteric cavity was infested with erosive ulcers which made intestinal mucosa congestive and edematous. Moreover several inflammatory granulomas were observed in distal ileum. The ileocecal valve was congestive and erosive (**Figure 5**). Laboratory tests revealed that triglyceride was elevated and HBsAg, anti-HBe and anti-HBc were positive, whilst serum levels of CRP and ESR were normal. A series of laboratory investigations were further made to further distinguish CD from relevant tumors, autoimmune disease and tuberculosis. Digestive tumor markers (CA199, CA724, AFP,

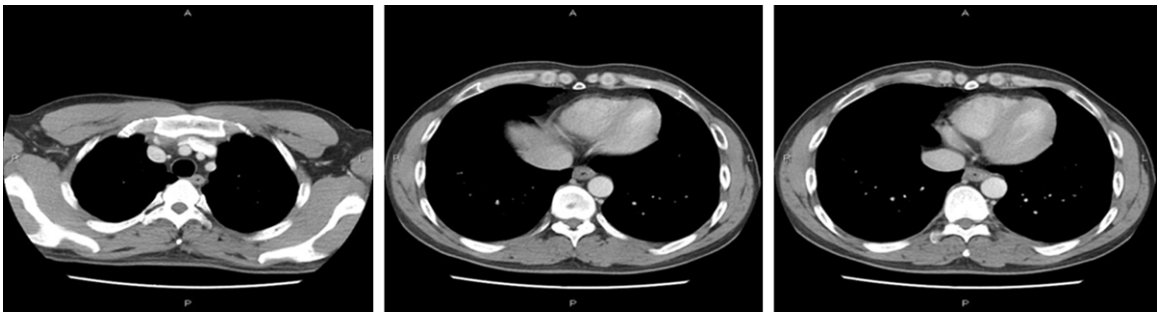

Figure 4. Chest CT scan showed that the esophageal wall is slightly thickened.

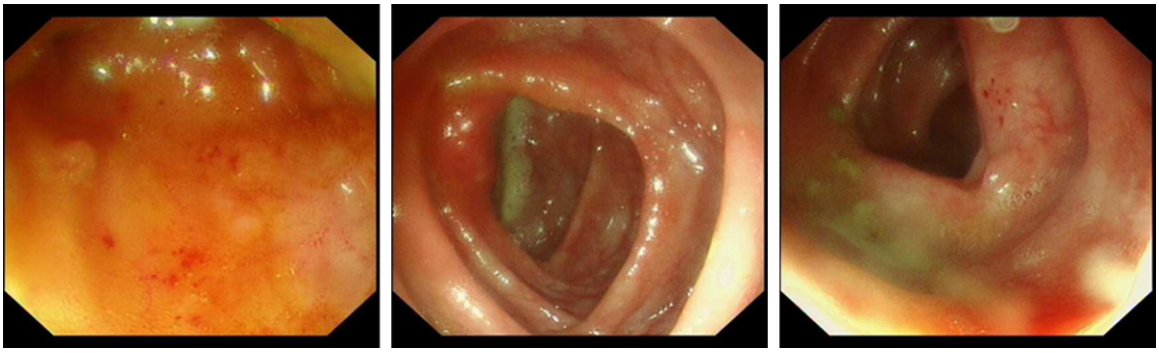

Figure 5. Colonoscopy of ileocecal junction and ascending colon. Inflammatory granulomas, congestion and erosive ulcers were observed.

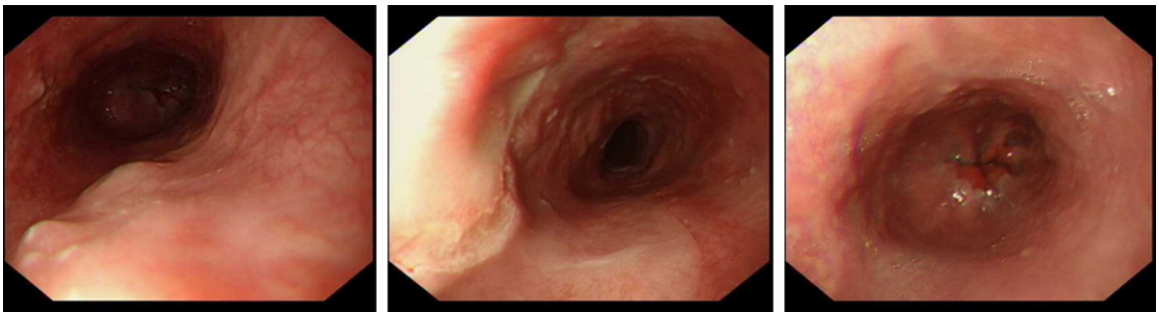

Figure 6. Gastroscopy of one month follow-up. One ulcer was reserved in esophagus wall. Other mucosal lesions were healed, only leaving several mucosal nodules.

CEA), autoimmune diseases related antibodies (HLA-B27) and mycobacterium tuberculosis IgG and IgM were showed negative. Then esophageal CD was considered by taking all these together.

The treatment was started with prednisone (30 mg q.d., p.o.), omeprazole (20 mg q.d., p.o.) and salicylazosulfapyridine (1.0 g q.i.d., p.o.). The follow-up gastroscopy in first month demonstrated that one ulcer was reserved in the esophagus wall, while other mucosal lesions were healed; only leaving several mucosal nodules (**Figure 6**). Two months later, repeated gastroscopy revealed that esophageal ulcers were disappeared and the mucosal lesions were completely repaired (**Figure 7**). Meanwhile, single balloon enteroscopy showed that ileocecal mucosa was hyperemic and edematous (**Figure 8**).

Discussion

CD may affect any part of the gastrointestinal tract from mouth to anus. However, it rarely affects esophagus. The clinical features of CD

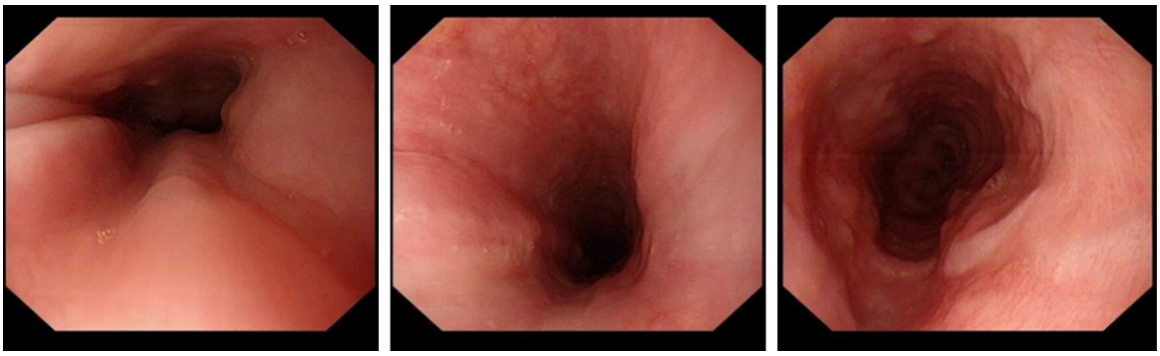

Figure 7. Gastroscopy of two months follow-up. Esophageal ulcers were disappeared and mucosal lesions were completely repaired.

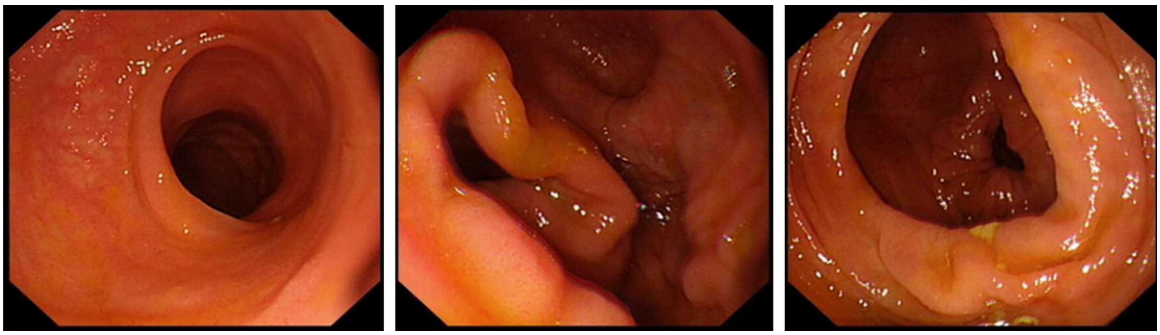

Figure 8. Single balloon enteroscopy of two months follow-up. Ileocecal mucosa was hyperemic and ulcers were disappeared.

vary according to lesion location and severity of the condition. Primary symptoms of esophageal CD are dysphagia, odynophagia and weight loss [6]. Meanwhile, major complications of esophageal CD is stenosis while fistulas is less common [5]. Our case is so infrequent that it affects esophagus and distal ileum. Moreover, the main presented symptoms are massive upper gastrointestinal hemorrhage companied with shapeless stool, arthralgia and repeated mouth ulcer.

Given the prognosis of proximal CD is worse [7] and long-term complications are difficult to manage, the diagnosis and treatment of CD is extremely crucial. As CD is a heterogeneous entity comprising of multiple various phenotypes [8], the diagnosis of CD is confirmed by clinical evaluation and a combination of endoscopic, histological, radiological or biochemical investigations [1] with exclusion of other possible causes. Particular attention should be paid to his risk factors of smoking and family history of ulcerative colitis. According to Crohn's Disease Activity Index (CDAI), phase of this case is the moderate phase without evidence of obstruction or abscess [1]. The histological finding reveals that the moderate acute and chronic inflammation is accompanied with granulomas. Gastroscopy, colonoscopy and chest CT are also approved of the diagnosis. In addition, laboratory and histological tests exclude the possible involvement of digestive tumor, autoimmune disease, Bechtel syndrome, tuberculosis, reflux esophagitis, esophageal lymphoma or infectious esophagitis (CMV, HSV, Candida albicans).

The treatment of esophageal CD is aimed at management of lesions and the underlying inflammatory process [9]. According to European consensus on management of CD, a proton pump inhibitor and systemic corticosteroids are recommended for the treatment of esophageal CD [10]. The patient was treated with prednisone, omeprazole and salicylazosulfapyridine which can relieve intestinal inflammation. In addition, it has reported that esoph-

ageal CD was successfully treated topically with swallowed aerosolized budesonide [11], adalimumab [12] and infliximab [13]. After systematic treatment for two months, the follow-up gastroscopy demonstrated that lesions had disappeared and esophageal mucosal lesions were healed. Single balloon enteroscopy showed that ileocecal mucosa was hyperemic and edematous, indicating this patient is in remission. A follow-up visit or reexamination at regular period as well as management of remission is needed for a patient of CD.

Conclusion

Early diagnosis and differential diagnosis of atypia CD, especially esophageal CD, are of great significance. We hope this case could make sense because the optimal treatment of esophageal CD has not been established and evidence-based therapy is mostly derived from case series. A comprehensive management of CD is needed which includes resolving existing inflammation, achieving mucosal healing and modifying external environment.

Disclosure of conflict of interest

None.

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References

[1] Van Assche G, Dignass A, Panes J, Beaugerie L, Karagiannis J, Allez M, Ochsenkuhn T, Orchard T, Rogler G, Louis E, Kupcinskas L, Mantzaris G, Travis S and Stange E. The second European evidence-based Consensus on the diagnosis and management of Crohn's disease: Definitions and diagnosis. J Crohns Colitis 2010; 4: 7-27.

[2] Ananthakrishnan AN. Epidemiology and risk factors for IBD. Nat Rev Gastroenterol Hepatol 2015; 12: 205-217.

[3] Molodecky NA, Soon IS, Rabi DM, Ghali WA, Ferris M, Chernoff G, Benchimol EI, Panaccione R, Ghosh S, Barkema HW and Kaplan GG. Increasing incidence and prevalence of the inflammatory bowel diseases with time, based on systematic review. Gastroenterology 2012; 142: 46-54, e42; quiz e30.

[4] Ng SC, Tang W, Ching JY, Wong M, Chow CM, Hui AJ, Wong TC, Leung VK, Tsang SW, Yu HH, Li MF, Ng KK, Kamm MA, Studd C, Bell S, Leong R, de Silva HJ, Kasturiratne A, Mufeena MN, Ling KL, Ooi CJ, Tan PS, Ong D, Goh KL, Hilmi I, Pisespongsa P, Manatsathit S, Rerknimitr R, Aniwan S, Wang YF, Ouyang Q, Zeng Z, Zhu Z, Chen MH, Hu PJ, Wu K, Wang X, Simadibrata M, Abdullah M, Wu JC, Sung JJ and Chan FK. Incidence and phenotype of inflammatory bowel disease based on results from the Asia-pacific Crohn's and colitis epidemiology study. Gastroenterology 2013; 145: 158-165, e152.

[5] van Hogezand RA, Witte AM, Veenendaal RA, Wagtmans MJ and Lamers CB. Proximal Crohn's disease: review of the clinicopathologic features and therapy. Inflamm Bowel Dis 2001; 7: 328-337.

[6] Decker GA, Loftus EV Jr, Pasha TM, Tremaine WJ and Sandborn WJ. Crohn's disease of the esophagus: clinical features and outcomes. Inflamm Bowel Dis 2001; 7: 113-119.

[7] Jess T, Winther KV, Munkholm P, Langholz E and Binder V. Mortality and causes of death in Crohn's disease: follow-up of a population-based cohort in Copenhagen County, Denmark. Gastroenterology 2002; 122: 1808-1814.

[8] Silverberg MS, Satsangi J, Ahmad T, Arnott ID, Bernstein CN, Brant SR, Caprilli R, Colombel JF, Gasche C, Geboes K, Jewell DP, Karban A, Loftus EV Jr, Pena AS, Riddell RH, Sachar DB, Schreiber S, Steinhart AH, Targan SR, Vermeire S and Warren BF. Toward an integrated clinical, molecular and serological classification of inflammatory bowel disease: report of a Working Party of the 2005 Montreal World Congress of Gastroenterology. Can J Gastroenterol 2005; 19 Suppl A: 5A-36A.

[9] Isaacs KL. Crohn's disease of the esophagus. Curr Treat Options Gastroenterol 2007; 10: 61-70.

[10] Dignass A, Van Assche G, Lindsay JO, Lemann M, Soderholm J, Colombel JF, Danese S, D'Hoore A, Gassull M, Gomollon F, Hommes DW, Michetti P, O'Morain C, Oresland T, Windsor A, Stange EF and Travis SP. The second European evidence-based Consensus on the diagnosis and management of Crohn's disease: Current management. J Crohns Colitis 2010; 4: 28-62.

[11] Zezos P, Kouklakis G, Oikonomou A, Pitiakoudis M and Simopoulos C. Esophageal Crohn's disease treated "topically" with swallowed aerosolized budesonide. Case Rep Med 2010; 2010: 2010.

[12] Musto D, Martorelli L, Cirillo A, Selvaggi F, Esposito P, Russo MI and Riegler G. Distal esophageal involvement in Crohn disease: short

treatment with adalimumab. Endoscopy 2010; 42 Suppl 2: E299.
[13] Chiba M, Sugawara T, Tsuda H, Abe T, Tokairin T and Kashima Y. Esophageal ulcer of Crohn's disease: disappearance in 1 week with infliximab. Inflamm Bowel Dis 2009; 15: 1121-1122.