Diffuse large B-cell lymphoma associated ileocecal intussusception in adulthood

Difuzní velkobuněčný B-lymfom asociovaný s ileocekální intususcepcí v dospělosti

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Summary

Background: Intussusception in adults is considered a rare condition, accounting for 5% of all cases of intussusceptions and approx. 1-5% of bowel obstruction. Almost half intussusceptions of the bowel are associated with malignant disease; thus, we should also treat the underlying malignancy. Case description: A 52-year-old male presented with colicky right lower abdominal pain for a 6-month period. He had a weight loss of 20kg within 6 months. Physical examination revealed a tender right lower abdominal mass. Colonoscopy showed a mass that filled the ileocecal. The digestive surgeon performed laparoscopic right hemicolectomy with end-to-end anastomosis. Histopathology examination showed diffuse proliferation of large tumor cells with centroblastic-like features prominently in submucosal area, with normal epithelial mucosa. The immunohistochemistry result concluded the final diagnosis of diffuse large B-cell lymphoma. RCHOP chemotherapy regimens were administered every 3 weeks for 6 cycles. The response was complete remission. Discussion: Intussusception was preoperatively diagnosed by multi-slice spiral CT scans with the characteristic target or sausage sign, edematous bowel wall and mesentery in the lumen. After surgery, approximately 90% of adult intussusception cases have a demonstrable etiology. Malignant lymphoma, especially diffuse large B-cell lymphoma, of the ileocecal is one cause of the adult intussusception. Conclusion: Adult bowel intussusception is a rare clinical entity. Abdominal CT is considered as the most sensitive imaging modality in the diagnosis of intussusception. Diffuse large B-cell lymphoma is the most common cause of ileocecal intussusception.

Key words

diffuse large B-cell lymphoma – intussusception – malignant lymphoma – bowel obstruction

Souhrn

Východiska: Intususcepce u dospělých je považována za vzácné onemocnění; na všech případech intususcepce se podílí z 5% a u neprůchodnosti střev z 1–5%. Téměř polovina případů intususcepce střeva souvisí s malignitou; měli bychom tedy léčit i vlastní nádorové onemocnění. Popis případu: Muž ve věku 52 let přišel s kolikovitými bolestmi pravého podbřišku trvajícími 6 měsíců. Během této doby zhubl o 20 kg. Fyzikální vyšetření odhalilo měkkou masu v pravém podbřišku. Kolonoskopie ukázala masu, která vyplňovala ileocékum. Chirurg provedl laparoskopickou pravostrannou hemikolektomii s terminoterminální anastomózou. Histopatologické vyšetření ukázalo difuzní proliferaci velkých nádorových buněk s charakteristikami podobnými centroblastům, převážně v oblasti podslizničního vaziva, při normální sliznici epitelu. Výsledek imunochemického vyšetření potvrdil finální diagnózu difuzního velkobuněčného B-lymfomu. Pacientovi poté byla podávána chemoterapie v režimu RCHOP každé 3 týdny v 6 cyklech. Odpovědí byla kompletní remise. *Diskuze:* Intususcepce byla předoperačně diagnostikována podle snímků z vícevrstvové spirální CT s charakteristickým terčem neboli "sausage sign", edematózní střevní stěnou a mezenteriem v lumen. Po operaci mívá přibližně 90% případů intususcepce prokazatelnou etiologii. Jednou z příčin intususcepce u dospělých bývá maligní lymfom, zejména pak difuzní velkobuněčný B-lymfom. Závěr: Intususcepce střeva u dospělých je vzácnou klinickou entitou. Nejcitlivější zobrazovací technikou při diagnostice tohoto onemocnění je CT břicha. Nejčastější příčinou intususcepce ileocéka je difuzní velkobuněčný B-lymfom.

Klíčová slova

difuzní velkobuněčný B-lymfom – intususcepce – maligní lymfom – neprůchodnost střev

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Introduction

Intussusception of the bowel is defined as the telescoping of a proximal segment of the gastrointestinal tract within the lumen of the adjacent segment. If we compare to children, bowel intussusception in adults is considered a rare condition, accounting for 5% of all cases of intussusceptions and approx. 1-5% of bowel obstruction [1]. It can present with a variety of acute, intermittent, and chronic vague and nonspecific symptoms, thus making its preoperative diagnosis difficult. CT scan of the whole abdomen proved to be the most useful diagnostic radiologic method. The treatment option of adult intussusception is surgical resection. Almost half intussusceptions of the bowel are associated with a malignant disease; thus, we should also treat the underlying malignancy.

Case description

A 52-year-old male presented with colicky right lower abdominal pain for a 6-month period. He also complained constipation, nausea, and vomiting. He had a weight loss of 20 kg within 6 months. Physical examination revealed a tender right lower abdominal mass. Laboratory findings showed mild anemia with a hemoglobin level of 11 g/dL, leukocytosis with white blood cells count 14,620/µL, and normal thrombocyte with platelet count 280,000/µL. Renal, liver function, and blood glucose were within normal limits.



Fig. 1. Colonoscopy showed tumor in the ileocecal region.

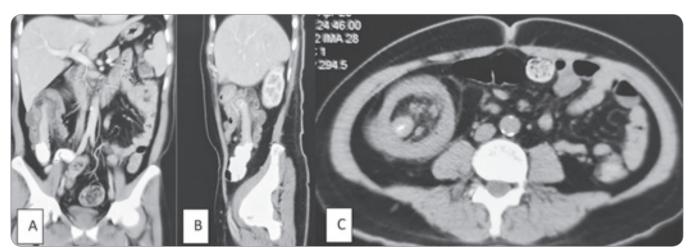


Fig. 2. A, B) Coronal and oblique CT-scan showed invagination of the terminal ileum and cecum into the lumen of the proximal transverse colon with sausage-like appearance; C) axial CT-scan of a target-like mass suggesting ileocecal intussusception with invaginated mesenteric fat and blood vessels.

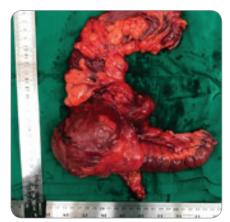


Fig. 3. A specimen from right hemicolectomy showed ileocecal intussusception.



Fig. 5. Complete remission after 6 cycles of RCHOP chemotherapy.

Lactate dehydrogenase (LDH) was also normal – 329 U/L (norm: 240–480 U/L). The level of carcinoembryonic antigen (CEA) was normal. Colonoscopy showed a mass that filled the ileocecal and did not allow the scope to enter (Fig. 1). CT scan of the whole abdomen showed the structure of ileum, mesenteric fat, and blood vessels invaginated into the structure of the cecum extension to the proximal of colon transversum with multiple lymphadenopathies around the mesenterium without sign of obstructive ileus (Fig. 2).

The digestive surgeon performed laparoscopic right hemicolectomy with end-to-end anastomosis. The resected colon section can be seen in Fig. 3. The result of histopathology examination showed diffuse proliferation of large tumor cells with centroblastic-like features prominently in submucosal area, with normal epithelial mucosa. The immunohistochemistry result (Fig. 4) showed positive result for B-cell marker CD20 and negative for T-cell marker CD3, epithelial marker AE1/3 and high Ki67 prolifera-

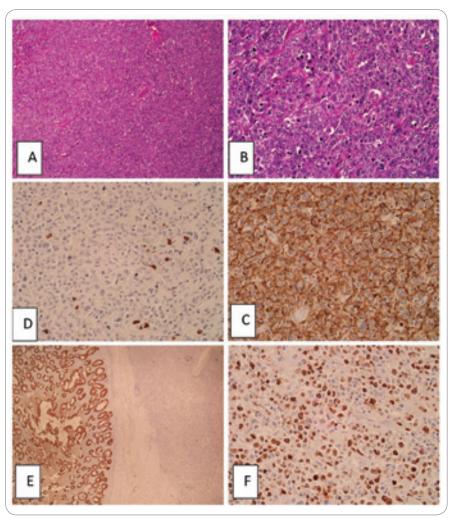


Fig. 4. Diffuse large B-cell lymphoma. A) The tumor showed diffuse pattern (hematoxylin & eosin $100\times$); B) the tumor cells are large with pleomorphic nuclei and centroblastic-like features (hematoxylin & eosin $100\times$); C) CD20 is positive (immunohistochemistry $400\times$); D) CD3 is negative (immunohistochemistry $400\times$); E) AE1/3 is negative at tumor cells (immunohistochemistry $100\times$); F) Ki67 proliferation index 60% (immunohistochemistry $400\times$).

tion index (60%). The final diagnosis was diffuse large B-cell lymphoma.

We administered the RCHOP regimen every 3 weeks for 6 cycles. The response was complete remission (Fig. 5). The patient tolerated the chemotherapy well, although he developed neutropenia, dyspepsia, nausea and vomiting, diarrhea, alopecia, and peripheral neuropathy during chemotherapy.

Discussion

Intussusception occurs if a proximal portion of the bowel invaginates into the distal bowel [2]. It can be classified into three types based on its location: (1) enteroenteric, when confined to the small

bowel; (2) colocolonic, when involving the large bowel; (3) enterocolonic, which can be ileocecal or ileocaeco-colonic [3]. It looks like that this patient suffered from ileocecal intussusception. The presentation of adult intussusception can be acute, subacute, or chronic non-specific symptoms therefore the initial diagnosis is often delayed. Yakan et al reported a retrospective review of adult patients with a diagnosis of intestinal intussusception that pain was the most common presenting symptoms (85%) followed by nausea, vomiting, constipation, rectal bleeding, and diarrhea [4]. This patient presented with chronic colicky right lower abdominal pain, constipation, nausea, vomiting, rectal bleeding, and weight loss.

Intussusception was preoperatively diagnosed by multi-slice spiral CT scans with the characteristic target or sausage sign, edematous bowel wall and mesentery in the lumen. Abdominal CT scan has been reported to be the most useful imaging technique to diagnose intussusception, with a diagnostic accuracy is 58–100%. Additional valuable information, such as metastasis or lymphadenopathy, is readily obtained by CT and may point to an underlying pathology. CT scan of the whole abdomen in this patient showed a characteristic finding of intussusception.

Treatment of adult intussusception usually requires resection of the involved bowel segment with primary anastomosis [4,5]. After surgery, approximately 90% of adult intussusception cases have

a demonstrable etiology. Malignant lymphoma, especially diffuse large B-cell lymphoma, of the ileocecal is one cause of the adult intussusception [5]. The digestive surgeon performed laparoscopic right hemicolectomy with primary anastomosis. Since the result of histopathology and immunohistochemistry was diffuse large B-cell lymphoma, we administered chemotherapy RCHOP to the patient resulting in a good response.

Conclusion

Adult bowel intussusception is a rare clinical entity. Its diagnosis is usually delayed due to nonspecific symptoms. Abdominal CT is considered as the most sensitive imaging modality in the diagnosis of intussusception. Diffuse large B-cell lymphoma is the most common cause of ileocecal intussusception. Laparoscopic hemicolectomy of the seg-

mental intussusception followed by chemotherapy RCHOP is the preferred treatment.

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