Ileocolic Intussusception as a Presenting Sign of Primary Lymphoma of the Colon

Milica Nestorović1, Goran Stanojević1, Vesna Brzački2, Vesna Milojković3, Branko Branković1, Vanja Pecić1, Ivan Pešić1, Igor Smiljković4

1Clinic for General Surgery, Clinical Centre, Niš, Serbia; 2Clinic for Gastroenterology and Hepatology, Clinical Centre, Niš, Serbia; 3Centre of Radiology, Clinical Centre, Niš, Serbia; 4Clinic for Vascular Surgery, Clinical Centre, Niš, Serbia

INTRODUCTION

Primary colorectal lymphomas are very rare and they account for 10-20% of gastrointestinal (GIT) lymphomas and only 0.2-0.6% of colorectal malignancies. Male predominance has been reported with a maximal incidence between 50 and 70 years. Intestinal intussusception or invagination in adults is also a rare condition. Only 5% of intussusceptions occur in adults, comprising only 1-5% of the cases with intestinal obstruction. In almost 90% of adult intussusceptions there is a pathological lesion that can only be clarified intraoperatively, making surgery unavoidable in most cases. We report a case of a young male with primary lymphoma of the colon presenting as ileocolic intussusception with the diagnosis established prior to surgery.

CASE REPORT

A 26-year-old man was admitted to the Clinic for Gastroenterology and Hepatology in Niš on August 26, 2009, with diarrhoea, diffuse abdominal pain and vomiting, especially after meals. Symptoms occurred five days prior to admission. The patient had no relevant past medical history. He did not smoke or drink. There were no malignancies in his family. Physical examination revealed a tender and distended abdomen without muscular defence or presence of palpable masses. There were no palpable lymph nodes. Erect plain abdominal radiography demonstrated the presence of air-fluid levels in the small bowel. The patient was hospitalized with the diagnosis of incomplete intestinal obstruction.

SUMMARY

Introduction Intussusception is a rare phenomenon in adults. It is caused mainly by malignant neoplasm. Primary lymphoma of the colon is a rare malignancy of the large intestine. The association of intussusception in adult and primary colorectal lymphoma is a diagnostic challenge, since they occur with a variety of atypical symptoms.

Case Outline We report a case of ileocolic intussusception in a 26-year-old man induced by primary lymphoma of the cecum. He was admitted to our hospital for incomplete intestinal obstruction. After thorough diagnostic work-up (plain abdominal radiography, abdominal ultrasonography, multi-slice computerized tomography, colonoscopy with biopsy), the patient underwent surgery. Intraoperative findings confirmed lymphoma as the cause of intussusception. The right hemicolectomy was carried out with end-to-side ileo-transverse anastomosis.

Conclusion Primary colorectal lymphomas should be considered in differential diagnosis of intussusceptions in adults. The treatment of choice is a radical resection where all oncological standards must be fulfilled.

Keywords: adult; colon neoplasm; extra-nodal lymphoma; ileocolic intussusception; non-Hodgkin lymphoma

Correspondence to:
Milica NESTOROVIC
Španskih boraca 4/9
18000 Niš
Serbia
milica20@yahoo.com
anastomosis. The postoperative course was uneventful. The pathology studies of the specimen confirmed extra-nodal diffuse large B-cell lymphoma of the colon. The predominant lymphoid population was immunoreactive for LCA, CD20, CD79a, CD43, Ki 67 (proliferation index 60-70%) and were negative for CK, ENA, CD5, CD23, CD10, bcl-2, bcl-6 and TdT. All of the fifteen examined lymph nodes tested negative for lymphoma. According to the Ann Arbor classification for extra-nodal lymphomas [4], it was classified as stage IE (lymphoma limited to the colon). The patient was then recommended to undergo three cycles of chemotherapy with CHOP (cyclophosphamide–vincristine–doxorubicine–prednisone) protocol.

**DISCUSSION**

Due to the rarity of primary colorectal lymphoma, the medical literature on this subject consists mainly of case-control and retrospective studies. Primary colorectal lymphoma accounts for only 1.4% of all lymphomas [5]. It represents an extra-nodal form of non-Hodgkin lymphoma. Incidence in adults takes third place, after gastric and small bowel lymphomas, unlike in younger subjects where intestinal localization is predominant [6]. Higher incidence is noted in the setting of immunosuppression like inflammatory bowel disease, HIV infection and after organ transplantation, but with a lack of strong evidence since there are no large studies [7]. All histological categories of lymphomas may occur in the GIT, but the most frequent subtypes (>90% of cases) are mucosa-associated lymphoid tissue (MALT) lymphoma and diffuse large B-cell lymphoma [8]. T-cell non-Hodgkin lymphomas more often appear among patients under the age of 18 years, and they are characterized by multifocal ulcerous lesions and have worse prognosis than B-cell lymphomas [9]. The most common sites of primary colorectal lymphomas are the cecum, as in this case, and the rectum, due to a greater amount of lymph tissue in these parts of GIT. The symptoms of the disease are non-specific mimicking colorectal cancer [10]. Atypical presentation delays diagnosis so that treatment begins in the advanced stages of the disease [7]. According to the literature, in 33.3 – 53.8% of patients with primary colorectal lymphoma, the diagnosis is established postoperatively after emergency surgery for acute intestinal obstruction or perforation [7, 9, 11].

Intussusception may occur in patients with primary lymphoma of the colon. In a large retrospective study carried out in Korea, intussusception was registered in 30.8% of patients with large B-cell colorectal lymphomas [12]. Intussusception involving the colon in adults most often occurs in flexible regions such as the sigmoid, the transverse colon and the cecum. The symptoms vary considerably, and are often associated with chronic obstruction. As a result, it is difficult to diagnose adult colonic intussusception, and most cases are diagnosed after patients undergo surgery. As in this case, adult intussusception should be considered when abdominal ultrasonography shows the typical concentric hyperechoic double ring coupled with the thickening of the intestinal walls, the „doughnut sign” [13]. The accuracy of ultrasonography in detecting intussusception is reported to be 60%. In the same group of patients the accuracy increased up to 91.7% in the presence of a palpable abdominal mass [14]. Masking by gas-filled loops and operator dependency
are major disadvantages of ultrasonography. CT allows a more detailed view. The reported diagnostic accuracy of CT scans varies from 58% to 100% [3, 14]. Few reports have described magnetic resonance imaging for adult intussusception and general characteristics are similar to those on computerized tomography [15]. Colonoscopy, which was performed in this case, is also useful in the diagnosis of intussusception [2]. In the case presented here, the diagnoses of both primary colonic lymphoma and intussusception were established preoperatively. Making the diagnosis prior to laparotomy plays an important role in determining therapeutic strategy.

In elective conditions, in cases of colorectal lymphomas most authors propose radical resection [5, 7]. The same treatment is mandatory for tumour induced intussusception. However, it remains a controversial matter whether or not to reduce intussusception before resection [3]. Although sometimes it is possible to reduce it by simple manipulation, this manoeuvre should be avoided in cases with suspected malignancy in order to prevent bowel perforation and tumour cell dissemination [16]. Since most lesions lead distally, they can easily be seen by colonoscopy. This diagnostic procedure might be helpful even during operation in distinguishing benign from malignant lesions before attempting to reduce intussusceptions [17].

The pathology findings in this case confirmed B-cell lymphoma graded IE. Approximately 50% of patients have regional lymph node involvement at the time of the diagnosis, which is considered a poor prognostic factor [18].

The treatment of primary malignant lymphomas requires a multidisciplinary approach, combining surgery and chemotherapy, with radiation reserved only for selected cases. Relaying on the literature, it is difficult to assess precisely the value of adjuvant therapy or chemotherapy alone. A significant increase of survival is reported among patients with stage II extra-nodal lymphoma after adjuvant chemotherapy (117.4 vs. 47.9 months) [5]. Some authors recommend preventive chemotherapy in patients with stage IE, with a reported 10-year survival of 80% [19]. The experience from our institution showed that in 87.5% of patients with adjuvant chemotherapy survival was 43.8 months, while in the group with surgery alone, it was 2.5 months [7]. Several prospective trials have shown that adding rituximab to standard CHOP regimen (R-CHOP) has shown higher response rates and better progression-free, event-free, disease-free and overall survival. The ideal time for administration of R-CHOP, the cycle length and the number of cycles are yet to be established [20].

In conclusion, the combination of primary lymphoma of the colon and adult intussusception is unusual. Primary colorectal lymphomas should be considered in the differential diagnosis of intussusceptions in adults. Making a diagnosis is difficult; however, by using available imaging techniques like MSCT, it can be done prior to surgery. In the case of malignant neoplasm as the leading point of intussusception, reduction is not recommended. The treatment of choice is radical resection where all oncological standards must be fulfilled.

REFERENCES
Илеоколична интусусцепција као презентујући знак примарног лимфома дебелог црева

Милица Несторовић1, Горан Станојевић1, Весна Брзачки2, Весна Милојковић3, Бранко Бранковић1, Vaња Пешић1, Иван Пешчић1, Игор Смиљковић4

1Клиника за општу хирургију, Клинички центар, Ниш, Србија;
2Клиника за гастроентерологију и хепатологију, Клинички центар, Ниш, Србија;
3Центар за радиологију, Клинички центар, Ниш, Србија;
4Клиника за васкуларну хирургију, Клинички центар, Ниш, Србија

КРАТАК САДРЖАЈ

Увод Интусусцепција или инвагинација се не јавља често код одраслих болесника. Најчешће је узрокована злоћудним тумором. Примарни лимфом дебелог црева је редак колоректални малигнитет. Истовремено постојање интусусцепције и примарног колоректалног лимфома код одраслих је су дијagnostички изазов, јер се оба стања испољавају разноврсним неспецифичним симптомима.

Приказ болесника Код двадесетишестогодишњег болесника дијagnostикована је илеоколична интусусцепција изазвана примарним лимфомом цекума. Болесник је примљен у нашу болницу као хитан случај због непотпуних опструкција црева. Након темељних дијagnostичких испитивања (нативна графија трбуха, упреваччни преглед абдомена, мултипласна компјутеризована томографија, колоноскопија са биопсијом) болесник је оперисан. Интраоперијационо је потврђен узрок интусусцепције, а потом је урађена десна хемиколектомија са кривањем термино-латералне анастомозе између илеума и трансверзалног колона.

Закључак У диференцијалној дијагнози интусусцепција код одраслих болесника у обзир треба узети и примарни лимфом дебелог црева. Метода избора у лечењу ових болесника је радикална резекција уз испуњавање свих онколошких стандарда.

Кључне речи: одрасла особа; неоплазма дебелог црева; екстранодални лимфом; илеоколична интусусцепција; нехонкински лимфом

Примљен • Received: 16/03/2010
Принхваћен • Accepted: 10/06/2010