



# Primary small bowel lymphoma presenting as invagination

Nidal İflazoğlu<sup>1</sup>, Aslı Mengeloğlu<sup>2</sup>, Nilay Şen Korkmaz<sup>3</sup>, Bilgin Karaalioglu<sup>4</sup>, Murat Yüklü<sup>5</sup>

## ABSTRACT

One of the causes of invagination in adults is primary small bowel lymphoma. Primary small bowel lymphomas are rare, present themselves with complications due to diagnostic difficulties, and are diagnosed only after surgical intervention. In case of invagination, one of the complications of these tumors, namely primary small bowel lymphoma, should also be considered as a cause in the diagnosis. In this paper, the diagnosis and therapy of a rare case of primary polypoid-type small intestinal lymphoma demonstrating findings of obstruction due to invagination have been presented and discussed in the light of the literature.

**Keywords:** Lymphoma, invagination, small bowel

## INTRODUCTION

An invagination is a condition in which the proximal part of the intestine folds into the distal part of the intestine, similar to the way the parts of a collapsible telescope slide into one another. One of the causes of invagination in adults is small bowel lymphoma (1, 2). Obstruction, perforation, bleeding, and invagination are the well-defined complications of small bowel lymphoma (2).

In this paper, the diagnosis of and therapy for a rare case of primary polypoid-type small bowel lymphoma demonstrating findings of obstruction due to invagination are presented and discussed in the light of the literature.

## CASE PRESENTATION

A 34-year-old male patient presented to the General Surgery Outpatient Clinic with complaints of weakness, weight loss (10 kg), and abdominal pain in the last 3 months and abdominal swelling and vomiting in the last 10 days. The patient looked pale, anergic, and anemic. The patient's life history and family history presented no significance. On physical examination, his blood pressure was 90/60 mm Hg, his pulse was 102/min, and his body temperature was 36.5°C. The thorax was found to be normal. Abdominal examination with deep palpation yielded a suspect mass in the right lower quadrant. The result of rectal examination was normal. His blood count showed the following: white blood cells, 4.2 K/mm<sup>3</sup>; hemoglobin, 8.5 g/dL; hematocrit, 27.2%; and platelets, 140,000 /µL. The results of his renal and hepatic function tests were within normal limits. Blood tumor markers (such as CEA and CA 19-9) and the results of a complete urine test were found to be normal. Abdominal computed tomography (CT) showed a mass in the right side of the abdomen (Figure 1). The subsequent colonoscopy identified an atypical mass lesion in the hepatic flexura, obstructing the lumen almost completely (Figure 2). Multiple biopsy specimens were obtained from the mass. The pathology report of the specimens was malignancy of an unknown nature. Upon findings of obstruction and determination of malignancy, the patient underwent conventional surgery. The exploration showed the ileum (with a palpable mass in the lumen) to have been invaginated into the right colon; moreover, it also showed palpable enlarged lymph nodes in the ileocolic mesothelium. The liver and spleen were evaluated and found to be normal. Subsequently, in accordance with the cancer surgery principles (total mesocolic excision+proper surgical boundaries), right hemicolectomy, partial ileum resection, and ileotransversostomy were performed.

In the following histopathological examination of the ileum specimen, invaginating from the ileocecal valve into the colon, that is, in the resected 18-cm long small intestinal and 25-cm long large intestinal material, macroscopically, there was a tumoral mass localized in the terminal ileum. The mass was 4×3×2.5 cm in size, had a smooth surface, was of fish-meat consistency in the sectional surface, and showed invasion to the serosa and the surrounding fatty tissue. Tumoral cells were observed in the microscopic examination of the material. The cells showed a diffuse distribution pattern; were round and of medium size; and had narrow basophilic cytoplasm, markedly hyperchromatic nuclei, and apparent multiple nucleoli (Figure 3). In the immunohistochemical examination of the material, CD45, CD20, CD79a, bcl-6, and CD10 were found to be positive, but CD3, CD5, CyclinD1, and bcl2 were determined

## Cite this paper as:

İflazoğlu N, Mengeloğlu A, Şen Korkmaz N, Karaalioglu B, Yüklü M. Primary small bowel lymphoma presenting as invagination Turk J Surg 2018; 34(4): 331-333.

<sup>1</sup>Department of General Surgery, Kilis State Hospital, Kilis, Turkey

<sup>2</sup>Department of Pathology, Kilis State Hospital, Kilis, Turkey

<sup>3</sup>Department of Pathology, Afyon State Hospital, Afyon, Turkey

<sup>4</sup>Department of Internal Medicine, Şanlıurfa Mehmet Akif İnan Training and Research Hospital, Şanlıurfa, Turkey

<sup>5</sup>Department of General Surgery, Afyon State Hospital, Afyon, Turkey

## Corresponding Author Nidal İflazoğlu

e-mail: nidal1933@yahoo.com

Received: 16.02.2015

Accepted: 10.06.2015

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Available online at  
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to be negative (Figure 4, 5). The Ki-67 proliferation index was 99%. Thus, the diagnosis was reported as Burkitt's lymphoma.

After establishing the diagnosis of small bowel lymphoma, systemic involvement according to Dawson's criteria was checked, and no peripheral lymphadenopathy was found (3). A thorax CT revealed no lymph node of pathological size. The examination results of a peripheral blood smear and bone marrow aspiration biopsy were also normal. Following all of these examinations, the patient received the final diagnosis of primary small bowel lymphoma. According to Musshoff's staging system, the patient was placed in Stage 2E1 because he had serosal involvement and regional lymph node positivity (4). In the postoperative stage, the patient developed no complications and was discharged with no problems from the hospital on the sixth postoperative day. The patient gave consent for this publication when he visited the hospital for controls.

### DISCUSSION

Invagination occurs often and idiopathically in childhood (5). In adults, invagination is seldom seen and is due to an underlying pathology in up to 80% of the cases. Of the patholo-

gies underlying invagination, about 40% are either primary or secondary malignant diseases. Polyps, lipomas, malignant masses, and adhesions due to past surgery are the causes or starting points of well-defined invaginations (1, 6, 7). In the present study, compatible with the literature, our patient had primary small bowel lymphoma as the starting point of his invagination.



Figure 1. Computed tomographic examination showed mass image in the right abdomen



Figure 2. Atypical mass lesion in the hepatic flexura of the colon, a colonoscopy figure

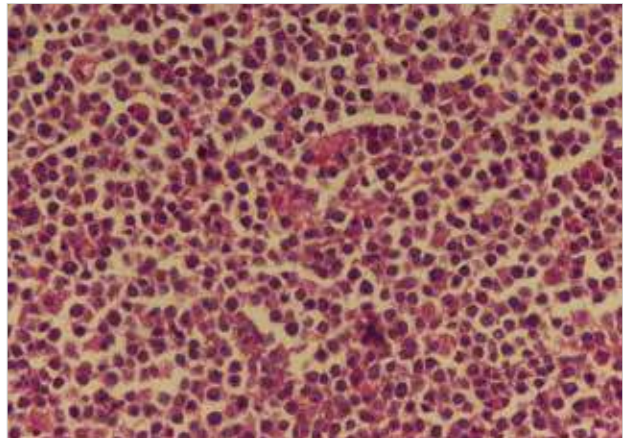


Figure 3. Microscopic examination of the material (H&E, 200x)

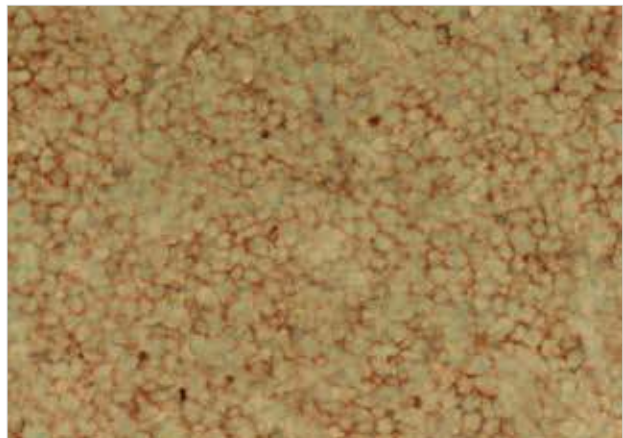


Figure 4. Immunohistochemical examination of the material (CD20, 400x)



Figure 5. Immunohistochemical examination of the material (CD45, 400x)

Primary lymphomas make up about 1%-4% of all gastrointestinal system tumors. Of these, about 20%-30% are primary small bowel lymphomas (8). According to Barakat's endoscopic classification, primary small bowel lymphomas are described as granulopapular, nodulopolypoid, ulcerative, infiltrative, and mixed (9). Retrospectively, it can be said that our patient had the nodulopolypoid type of lymphoma.

Small bowel lymphomas can have different clinical courses; generally, however, they present with nonspecific findings and cause difficulty in endoscopic or radiological diagnosis due to their localization and consequently are diagnosed when complicated or by histopathological examination following surgery. Many presented cases in the literature have been diagnosed months after the symptoms start (10). The fact that our patient was also diagnosed months after the symptoms had begun as a complicated case points to the diagnostic difficulty of the disease.

Histopathologically, about 90% of small bowel lymphomas are B-cell non-Hodgkin lymphoma, and the next most common types are T-cell non-Hodgkin lymphoma and Hodgkin lymphoma (6). Our patient was diagnosed with Burkitt's lymphoma, which is one of B-cell non-Hodgkin lymphomas.

In recent years, primary small bowel lymphomas are increasingly seen, particularly in immunocompromised patients, and frequently cause co-morbidity in cases with transplantation history, inflammatory intestinal diseases, and some immunodeficiency syndromes (11, 12). Our patient had no such co-morbidity based on his anamnesis and clinical findings.

The treatment of primary small bowel lymphomas is still controversial and lacks a clear optimum strategy. This situation may be due to the limited number of cases and the lack of high-quality prospective studies. Various combinations of surgery, chemotherapy (CT), and radiotherapy (RT) are the options for treatment. Although surgery and RT are thought to provide local control of the tumor, these two therapeutic approaches are being increasingly displaced by alternatives of combined chemotherapy. Surgery is recommended for complicated cases during the process of diagnosis/therapy or for post-therapy residual disease. It has been reported that chemotherapy alone is equivalent to surgery in uncomplicated cases in the early stage (13, 14). Since our patient was a complicated case, we performed surgery and planned to initiate chemotherapy.

## CONCLUSION

Primary small bowel lymphomas are rare, present themselves with complications due to diagnostic difficulties, and are diagnosed only after surgical intervention. In case of invagination, one of the complications of these tumors, namely, primary small bowel lymphoma, should also be considered as a cause in the diagnosis. We are of the opinion that with the advances in diagnostic methods and technical facilities, primary small bowel lymphomas can be diagnosed early and treated before complications develop.

**Informed Consent:** Written informed consent was obtained from patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept - N.İ., A.M.; Design - N.İ., A.M., N.Ş.K.; Supervision - N.İ., B.K., M.Y.; Resource - A.M., N.Ş.K.; Materials - N.İ., B.K., M.Y.; Data Collection and/or Processing - N.İ., N.Ş.K.; Analysis and/or Interpretation - B.K., M.Y.; Literature Search - N.İ., B.K., M.Y.; Writing Manuscript - N.İ., B.K., M.Y.; Critical Reviews - N.İ., A.M., N.Ş.K.

**Conflict of Interest:** The authors have no conflicts of interest to declare.

**Financial Disclosure:** The authors declared that this study has received no financial support.

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