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CASE REPORT

Primary Small Intestinal Non-Hodgkin's Lymphoma: A Case Report

Lymphome non Hodgkinien Primaire de l'Intestin Grêle : Un Rapport de Cas

¹I. A. Udo, ²C. C. Nwafor

ABSTRACT

BACKGROUND: Primary intestinal lymphoma has not been previously reported in our unit, and we consider it to be a very rare cause of acute small bowel obstruction.

METHODS: We present an adult male with features of recurrent small intestinal obstruction who previously underwent umbilical hernia repair for the same pain. A plain x-ray and ultrasound scan showed features of intestinal obstruction but did not suggest an aetiology of his symptoms.

RESULTS: He was resuscitated and underwent an exploratory laparotomy and resection of an obstructing ileal mass with mesenteric nodes. Primary anastomosis of healthy ileum was done and the post-operative period was uneventful. The tissue was reported as low-grade B-cell non-Hodgkin's lymphoma (NHL). He was placed on CHOP with a satisfactory response.

CONCLUSION: Small intestinal lymphoma is a rare cause of intestinal obstruction. **WAJM 2023; 40(2): 232–234.**

Keywords: Primary intestinal lymphoma, Recurrent intestinal obstruction, Laparotomy.

RÉSUMÉ

CONTEXTE: Le lymphome intestinal primaire n'a pas été rapporté précédemment dans notre unité, et nous le considérons comme une cause très rare d'obstruction aiguë de l'intestin grêle.

MÉTHODES: Nous présentons un homme adulte présentant les caractéristiques d'une obstruction récurrente de l'intestin grêle et ayant déjà subi une réparation de hernie ombilicale pour la même douleur. La radiographie et l'échographie ont montré des caractéristiques d'obstruction intestinale mais n'ont pas suggéré l'étiologie de ses symptômes.

RÉSULTATS: Il a été réanimé et a subi une laparotomie exploratoire et la résection d'une masse iléale obstructive avec des ganglions mésentériques. Une anastomose primaire de l'iléon sain a été réalisée et la période postopératoire s'est déroulée sans incident. Le tissu a été déclaré comme étant un lymphome non hodgkinien (LNH) à cellules B de bas grade. Il a été placé sous CHOP et avec une réponse satisfaisante.

CONCLUSION: Le lymphome de l'intestin grêle est une cause rare d'obstruction intestinale. **WAJM 2023; 40(2): 232–234.**

Mots clés: Lymphome intestinal primaire, Obstruction intestinale récurrente, Laparotomie.

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INTRODUCTION

The small intestine is a very rare primary site for malignant tumours compared to the shorter large intestine, and all tumours occur predominantly in males.¹ Primary small intestinal lymphoma is even rarer. The rare nature of primary small intestinal lymphoma coupled with its non-specific presentations may lead to difficulty with its early diagnosis and treatment. The optimal treatment for primary small intestinal lymphoma is undetermined; surgery, chemotherapy, or a combination of both modalities are employed variously.

CASE REPORT

A 47-year-old man presented with a three-hour history of progressively severe and centrally located colicky abdominal pain associated with abdominal distension, vomiting and inability to pass stool or flatus. Similar symptoms and presentations had been recurrent in the past three years, associated with nausea and vomiting. Symptoms often resolve spontaneously with the passage of loose dark stools. He did not volunteer a history of weight loss. He previously underwent an umbilical hernia repair a year to current symptoms, on account of similar complaints but symptoms continued at intervals.

He was ill looking and in severe pain, well hydrated and not pale or jaundiced. There were no enlarged peripheral lymph nodes. His abdomen was centrally distended, tender and resonant. There was a poorly delineated mass in the peri-umbilical region. His bowel sounds were absent and rectal examination unremarkable. Haematological and biochemical parameters were normal. A plain abdominal x-ray showed multiple centrally located air-fluid levels and an ultrasound scan showed gaseous distension. A clinical diagnosis of small intestinal obstruction secondary to intussusception was made and he underwent urgent exploratory laparotomy.

Intra-operatively, the ileum and jejunum were dilated with an 8cm x 8cm x 6cm fleshy mass in the middle portion of the ileum extending into the mesentery with multiple enlarged mesenteric lymph

nodes up to the root of the mesentery (Figure 1). There was evidence of a constriction 4cm proximal to the mass. Primary resection of the mass with 5cm margins to include the mesenteric nodes and anastomosis was done. The mass was approximately 3cm–4cm thick with a fish-flesh appearance. The lumen contained altered blood.

His post-operative course was unremarkable and he was discharged on the 5th day. Histology of the tissue showed non-Hodgkin's lymphoma (Figure 2). Immuno-histochemistry showed strongly positive CD20 tumour

cells that were CD3 negative, bcl6 negative, CD 10 negative, cmc negative, with KI 67 proliferative index of approximately 5%. A final diagnosis of low-grade B-cell NHL was made. He received cyclophosphamide 750mg/m², Adriamycin 50mg/m², vincristine 1.2mg/m² and prednisolone 40mg and has remained without complaints at fifteen months.

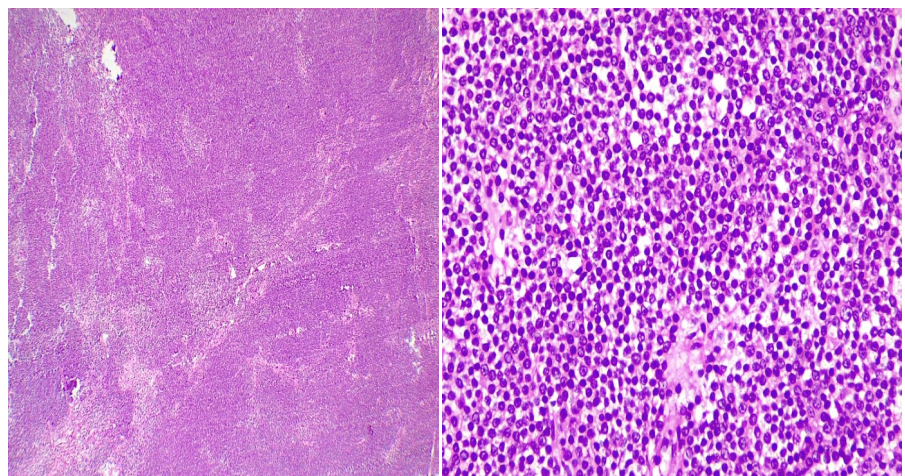
DISCUSSION

Malignant tumours of the small bowel occur rarely in Nigeria.^{2,3} We previously reported on benign small intestinal tumours presenting with recurrent small intestinal intussusception.⁴ This is our first report of small intestinal lymphoma in an adult presenting with recurrent intestinal obstruction. The clinical diagnosis of intussusception was correct but the specific lesion causing it was not known; small intestinal lymphoma was not considered as a primary cause owing to its rarity in our practice.

The diagnosis of primary intestinal lymphoma could be difficult owing to its rarity and lack of specific clinical features. Abdominal x-ray and ultrasound scan could identify features of intestinal obstruction but the specific diagnosis was elusive. Small bowel enema showing mucosal effacement and CT scan showing intestinal mass do suggest a malignant disease.⁵ These later imaging modalities, which are more sensitive and specific, are



Fig. 1: Small Intestinal Mass with Multiple Mesenteric Lymph Nodes and Proximal Loop Dilatation.



Figs: 2A & 2B: x40 & x400 Magnification, showing Diffuse Small to Intermediate Sized Cells, with Hyperchromatic Nuclei and Scattered Mitotic Figures. No Intestinal Mucosa Seen.

not routinely employed in our practice to investigate non-specific abdominal pain, but emerging challenges indicate a need to increasingly employ them where and when indicated.

The incidence of extra-nodal lymphoma is reported to be increasing more rapidly in the Middle East, Europe and the US compared to the nodal disease.⁶ There is a need to increase surveillance for this disease among patients presenting with non-specific abdominal pain in our practice, every small intestinal tumor must be fully investigated. Studies show a wide geographical variation in the primary site for extra-nodal NHL, but the gastrointestinal tract, the ileum in particular, is considered to be the most common site with high-grade B-cell lymphoma being the most common sub-type in all patients.⁷⁻⁹

The optimum treatment for small intestinal NHL is not determined. Surgical resection with a safe margin of the involved segment of the small intestine and of the mesenteric nodes is practiced as a first-line treatment for small intestinal NHL. Adjuvant chemotherapy or

radiotherapy is also practiced. Our primary approach in the index case was surgical resection, especially because he was obstructed. However, there are reports that question the role of surgery in uncomplicated disease, preferring chemotherapy alone.⁶

Summary

Small intestinal lymphoma is a known rare cause of small intestinal obstruction in adults. Surgery is a common primary modality of treatment but its role in uncomplicated cases is questioned.

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