Case Report

Florid Reactive Lymphoid Hyperplasia of Terminal Ileum - A Rare Case

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ABSTRACT

Florid reactive lymphoid hyperplasia in terminal ileum can present to a surgeon as acute abdominal pain. Only few cases were reported in literature. Our case represents that a rare case of florid reactive lymphoid hyperplasia can present to a surgeon as acute appendicitis. During the operation the gross appearance may mimic Crohn’s disease. A limited resection may clinch the diagnosis of florid lymphoid hyperplasia / Crohn’s disease. In florid reactive lymphoid hyperplasia limited resection may be curative.

Keywords: Acute appendicitis, terminal ileum, reactive florid hyperplasia.

INTRODUCTION

Florid lymphoid hyperplasia is a rare condition with an unknown aetiology. This condition was first reported by Briquet in 1838 and later it was investigated radiologically by Marina-Fiol and Rof-Carballo in 1941 and given a new name of “enteritis follicularis”. [1] Five cases of florid lymphoid hyperplasia of the ileocecal region are described. [2] Disease may involve the stomach, the entire small intestine, and the large intestine, [3] but is more common at the terminal ileum and rectum. [4,5] Lymphoid hyperplasia of the gastrointestinal tract represents a rare disease that is grossly characterized by the presence of numerous visible mucosal nodules measuring up to, and rarely exceeding, 0.5 cm in diameter. [3] The common presentation is abdominal pain with or without loose stools or gastrointestinal bleeding. [1] Here we report our case which presented as acute appendicitis. It is therefore important to have a working knowledge of the clinical manifestations of the disease, so that a systemic approach to the assessment of these patients can result in rational perioperative management.

CASE DESCRIPTION

A young 14 years girl, student, weighing 35kg, presented to the emergency department with a history of pain in the right iliac fossa for 4 days and a worsening pain over the last two days. She had no other gastrointestinal (GI) symptoms such as nausea/vomiting, altered bowel habits, bleeding per rectum, or weight loss. Her past and family histories were unremarkable. She was moderately built and nourished. All the vital parameters were normal. On physical examination there was minimal abdominal distension with severe tenderness and guarding in the right iliac fossa. Systemic examination revealed normal cardiovascular and respiratory systems. A
A clinical diagnosis of acute appendicitis was made. Blood investigations revealed increased inflammatory markers such as white blood cell count (13.3×10^9/l), neutrophils (9.5×10^9/l) and C reactive protein (45 IU). All other blood investigations were within normal limits. Plain abdominal x-ray showed normal gas pattern with no obvious radiological abnormality. Abdominal ultrasonography also confirmed the clinical diagnosis of inflamed appendix measuring 13mm with inflamed adjacent mesentery.

Open appendectomy was planned under spinal anaesthesia and accordingly written informed consent was obtained. During surgery, after appendectomy was performed as per the routine practice in our institution while screening for Meckel’s diverticulum we found a surprise. The Meckel’s diverticulum extended from umbilicus to urinary bladder with internal herniation of terminal ileum which was causing subacute obstruction. Hence, Meckel’s diverticulectomy was performed with release from umbilical and urinary bladder attachements and an end to end ileo-ileal anastomosis was done. The specimen was sent for histopathological evaluation.

Intraoperatively the patient’s vitals were stable, tolerated the procedure well, as well recovered satisfactorily postoperatively and were discharged home without any complications, with a further clinical follow-up after 6 months.

On microscopic examination, there was pronounced lymphoid hyperplasia of the terminal ileum within an area of Payer’s patch, with increased small lymphoid cells and reactive lymphoid follicles. The mucosal surface was focally ulcerated and there were transmural lymphoid aggregates. Eosinophils and occasional neutrophils were also present within the small bowel wall and on the serosal surface. Submucosal lymphatic tissue was distended and filled with lymphocytes. These findings eventually lead to the diagnosis of Florid Lymphoid Hyperplasia of terminal ileum and appendix.
DISCUSSION

To define lymphoid hyperplasia is abnormal proliferation of secondary follicles that occurs principally in the cortex of the lymph node without broaching the capsule due to stimulation of the B cell compartment. Histologically, hyperplastic lymphoid follicles with large germinal centres are seen in the lamina propria and superficial submucosa. There is enlargement of the mucosal B cell follicles caused by hyperplasia of the follicle centres; surrounded by a normal appearing mantle zone. In benign hyperplasia there is no tendency for interfollicular infiltrate to invade the follicles. Follicular hyperplasia usually occurs in children and young adults as in our case was a 14 year girl. In immunocompetent individuals, the lesion is often located only in the distal ileum and proximal colon. In immunodeficient patients, the nodules are frequently more generalized, affecting the upper small intestine in addition to distal small intestine and colon. The condition is hypothesized to be the result of an antigenic response, Yersinia infection, Adenovirus infection, Shigella, Giardia infection. The disorder is also found to occur in patients with dysgammaglobulinemia, isolated IgA deficiency, ulcerative colitis and regional enteritis. In our case, however we did not find any source of infection.

The most important tools to differentiate lymphoid hyperplasia from malignant lymphoma are immunohistochemical analysis (both kappa and lambda chains diagnostic) and monoclonal immunoglobulin gene arrangement. The malignant potential of this condition is still unclear. Our case did not show any evidence of malignancy. The potential for malignancy associated with lymphoid hyperplasia is the most conflicting factor when deciding therapeutic measurements.

CONCLUSION

To conclude, florid lymphoid hyperplasia is a rare entity which may present to a clinician with clinical features suggestive of acute appendicitis, intestinal obstruction, Crohn’s disease or malignant tumour of the small/large bowel. Local surgical excision is the treatment of choice.

Thus, reactive follicular hyperplasia is a rare benign disease with challenging issues to the surgeon in terms of diagnosis and management.

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How to cite this article: Bokil C, Naik P. Florid reactive lymphoid hyperplasia of terminal ileum- a rare case. Int J Health Sci Res. 2016; 6(2):405-408.

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