ACTA SCIENTIFIC CLINICAL CASE REPORTS

Volume 6 Issue 1 January 2025

Case Report

Primary Lymphoma of the Appendix: A Case Report

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Received: October 08, 2024

Published: December 10, 2024

Abstract

Introduction: Primary lymphomas of appendix are extremely rare tumors. The incidence is 0.015% of all gastrointestinal lymphomas.

Presentation of Case: We present a case of a 58 year-old female patient who presented with lower abdominal pain since 15 days with one episode of vomiting and intermittent fever.

Discussion: The patient received exploratory laporatomy. The definitive histopathological examination revealed Non- Hodgkin's lymphoma involving appendix, ileum and lymph nodes in mesocolon. The neoplasms of appendix usually manifest clinically with sign and symptoms of acute appendicitis from luminal obstruction (30-50%). Preoperative diagnosis is difficult and often occurs through histopathological examination.

Conclusion: Primary appendiceal lymphoma is rare and there are no clear guidelines for therapy. Primary surgical resection followed by post-operative chemotherapy showed high efficacy. The histopathological examination of all appendectomy is essential.

Keywords: Appendicitis; Appendix; Case Report; Lymphoma

Introduction

Primary lymphoma of the appendix is an extremely rare entity that accounts for approximately 0.015% of the lymphomas of the gastrointestinal tract. There are very few published reports of the pathology. Burkitt's lymphoma is more frequent in children, whereas the majority of cases in adults correspond to large B-cell non-Hodgkin lymphoma.

The majority of articles found in the international literature correspond to case reports. In the case series by Marcelo Zamorano., *et al.* from a total of 7626 appendectomy specimens, there were only 25 appendiceal tumors and just 2 of them were primary lymphomas [5]. In another case series that included 5307 appendectomy specimens, Daniel Esmer., *et al*. found a total of 31 cases (0.58%) of appendiceal tumor, none of which corresponded to appendiceal lymphoma [4]. A total of 1060 appendectomy specimens were reviewed over a 10-year period at the Kamineni Hospitals in India and only one case of appendiceal lymphoma was found, illustrating the rareness of the pathology [10].

Case Report

A 58 years old female came to Opd with chief complains of pain in lower abdomen since 15 days. Patient was apparently alright 15 days back when she presented with pain in lower abdomen which was insidious in onset, progressive in nature, initially at periumbilical area further radiating to right lower quadrant, associated with one episode of bilious vomiting and intermittent episodes of fever. No any other complains on presentation. No significant medical or

surgical history in the past. Patient reached menopause 6 years ago and obstretic score was P3A0L3 with normal vaginal delivery.

On examination, general physical examination was within normal limits. Per abdominal examination revealed soft, mild tenderness in right iliac fossa and localised guarding in right lumbar and right iliac fossa. Per rectal examination was normal. Xray erect abdomen was done and showed no signs of obstruction. Clinical examination favoured towards appendicitis.

Patient was admitted and blood investigations were done along with usg abdomen pelvis followed by cect abdomen. Blood reports showed Hb 13 g/dl, pcv 39.7%, TLC 10,700/ul, platelets 4.29 lakhs/cumm, Pt 14.0 sec, INR 1.1, Bsl® 140mg/dl, Urea 15 mg/dl, Creatinine 0.7 mg/dl, Potassium 4.3 mEq/L, sodium 134 mEq/L. Usg abdomen pelvis report suggested proximal 2/3rd segment of appendix is dilated and mildly ballooned up with maximum diameter at base measuring approx 2cm with base shows invagination into caecum extending for approx 2cms. Multiple, enlarged nodes are noted in nearby mesentry –largest 22mm and many smaller, discrete nodes are seen in periumbilical mesentry. On usg diagnosis was given favouring mucocole of appendix/meckels diverticultis with abscess/subacute small bowel perforation with localised abscess. CECT abdomen report shows well defined globular, intraluminal, exophytic, mildly and homogenously enhancing, soft tissue density lesion of size 5.5x3.8x6.3cm is noted arising from distal ileal loop with rim of oral contrast around it. Another small similar morphology lesion of size 2.1x2.7x3cm is also noted involving base and proximal body of appendix. Multiple non necrotic, discrete lymph nodes are noted in mesentry at umbilical region with adjacent mesenteric fat stranding, largest lymph node measures 2.3 x 1.4cm.

Patient was operated - exploratory laporatory was done. Intraoperative findings showed 3x3 cm appendicular mass with 6x5cm distal ileum mass – 5cms proximal to ileocaecal junction with multiple mesenteric lymphadenopathy of largest 4cm, so we performed right hemicolectomy followed by ileotransverse anastomosis. Resected part was sent for histopathology examination. HPR report (B/4627/2020) was suggestive of Non- Hodgkin's lymphoma involving appendix, ileum and lymph nodes in mesocolon and proximal and distal surgical margins free from tumour.

Intra-operative photographs

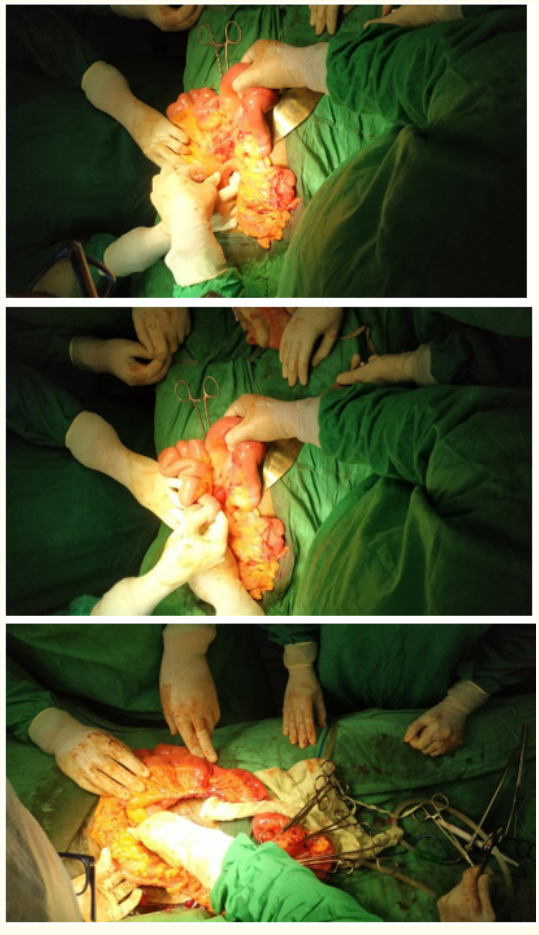

Figure 1

Microscopic photos

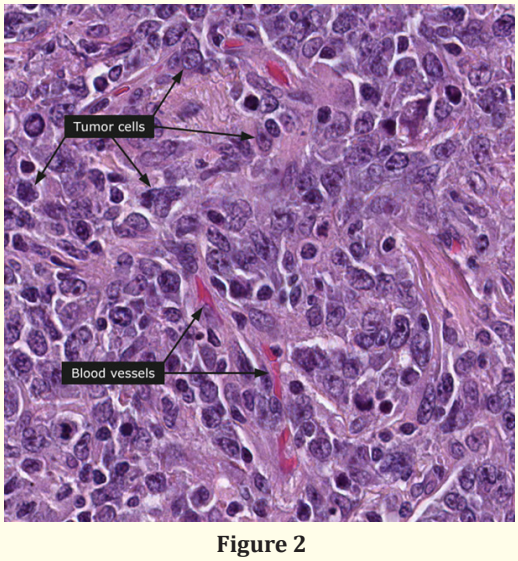

Figure 2

Discussion

Malignant lymphoma comprises approximately up to 1% to 4% of the gastrointestinal malignant neoplasms [1]. Primary appendicular lymphoma is even rarer and often a postoperative diagnosis with the reported incidence of approximately 0.015% in a large series of 710,000 human appendectomy specimens [2]. The most common presenting symptom was pain in the right iliac fossa occurring over a period of few months and frequently associated with a palpable mass. Muller., *et al*. reported four cases of lymphoma presenting as acute appendicitis [3]. Our case also presented with abdominal pain and all underwent laparotomy surgery with suspicion to appendicitis. Men are more likely to develop appendiceal lymphoma over women by 1.5:1 with a median age onset of 18 years [4]. Although, there are numerous reports of acute surgical presentations of appendicular Burkitt's lymphoma, all of the cases have been reported in children and young adults [8]. Although, there are no classical imaging features of appendiceal lymphoma, enlargement of appendix beyond 1.5 cm in diameter on CT-scan should be viewed with suspicion, and a diameter above 2.5 cm should be even more concerning. In the appendiceal lymphoma, as with all lymphoma, chemotherapy for treatment is required [5].

Appendiceal neoplasm is uncommon and consists of a heterogeneous group of pathologies and many present as appendicitis, but may be encountered as an unexpected finding at laparascopy or laparotomy, or on pre- or post-operative imaging[7]. Knowledge of the different types of neoplasm and appropriate treatment allows the surgeon to provide patients with optimal care and referral to a specialist center if appropriate.

Conclusion

Primary appendiceal lymphoma is rare and there are no clear guidelines for therapy [6]. Acute appendicitis is a frequent presentation of primary appendiceal lymphoma. Primary surgical resection followed by post-operative chemotherapy showed high efficacy [9]. The histopathological examination of all appendectomy is essential.

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