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CASE REPORT

# Unusual presentation for primary appendiceal lymphoma: A case report

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**Abstract** Primary appendiceal neoplasms are uncommon, being found in approximately 0.5%–1.0% of appendectomy specimens at pathologic evaluation. Primary appendiceal Burkitt's lymphomas are rare occurring in 0.015% of all gastrointestinal lymphomas. Almost all reported cases of appendiceal lymphoma have proved to be non-Hodgkin lymphoma. The majority of appendiceal lymphomas are of B-cell. Patients were almost entirely males. Acute appendicitis is the most common clinical manifestation. This report describes a rare case of primary appendiceal lymphoma in a patient presented with hematuria and dull aching right lower abdominal and back pain.

**Keywords** Appendix · Burkitt's · Lymphoma

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## Introduction

Primary appendiceal Burkitt's lymphomas are rare tumours. Acute appendicitis is the most common manifestation for most appendiceal tumors. Goblet cell carcinoid tumor and non-Hodgkin lymphoma of the appendix usually infiltrate the entire appendix. Computed tomography (CT), is effective in the evaluation of appendiceal neoplasms and appears to be the modality of choice whenever an appendiceal mass is suspected.

## Case report

A 49-year-old male was admitted to the hospital complaining of two attacks of gross haematuria and moderately severe, dull aching lower right abdominal and back pain for 2 weeks. The pain was not radiating, not related to meals or urination, and not associated with fever, weight loss or change in bowel habits. It responded to over-the-counter (OTC) analgesics and was occasionally associated with nausea and vomiting. No history of renal diseases or stones. Physical examination was significant only for lower abdominal tenderness with more on the right iliac fossa and suprapubic region without any signs of peritoneal irritation. Labs were within normal limits with a white blood cell count of 10.8 thousand/ml. Kidneys, Yreters and Bladder (KUB) and intravenous pyelography were negative. Abdominal ultrasonography showed acute appendicitis with appendicular mass formation. A CT scan of abdomen with contrast revealed retrocaecal elongated thick wall phlegmanous appendicitis or neoplastic involvement of appendix and caecum with multiple enlarged mesenteric lymphadenopathies anterior and below the third part of the duodenum. The largest node measures 3.3 × 3.2 cm with central necrosis (Fig. 1). Cystoscopy was negative.

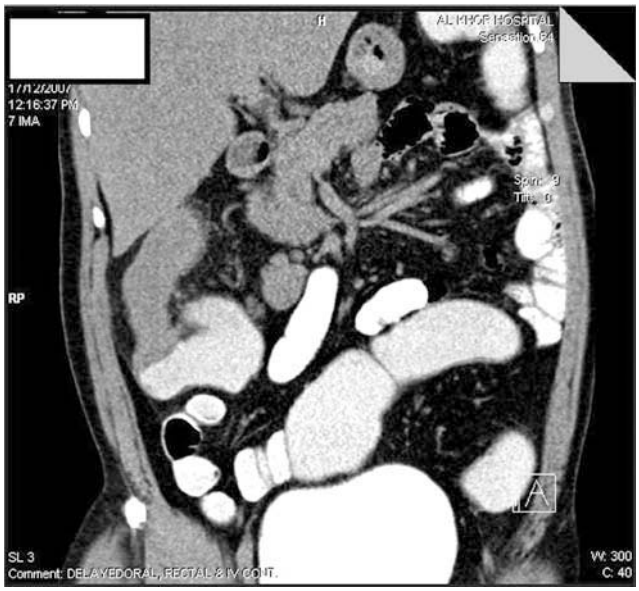


**Fig. 1** Abdominal CT with contrast showing the appendicular mass

The patient was prepared for and consented to laparoscopic exploration which revealed a large appendicular mass about 7 × 5 cm invading the caecum and adherent to the underlying surface of the liver with a large mesenteric lymph node about 3 × 3 cm along the right colic artery. Because of extensive adhesions at the right iliac fossa, the procedure was converted into a laparotomy through a midline incision. The appendicular mass was hard in consistency, invading the caecum, was subhepatic and adherent to the underlying surface of the liver. We suspected either appendiceal or caecal tumour.

Surgery proceeded with a right haemicolectomy and ilieotransverse anastomosis using EndoGIA linear stapler.

Mesenteric lymph nodes along the right colic artery were excised. On the fourth postoperative day the patient developed mild wound sepsis. A swab culture was taken and sent for bacteriological examination which revealed *Enterococus fecalis*, sensitive to ampicillin and trimethoprim/sulfa, which treatment was applied with good response. Pathological examination revealed Burkitt's lymphoma of the appendix. The tumor infiltrated the muscularis propria up to serosa within 1 mm of the serosal margin. Proximal and distal resection margins were free of malignancy. One mesenteric lymph node was involved and measured 2.7 × 2.4 × 2.4 cm with extranodal extension. The other 14 lymph nodes were free of malignancy (Figs. 2, 3, 4, 5). Carcinoemberyonic antigen was ordered and it was 0.7 µg/l. Patient was discharged after nine days with oncological referral. Postoperative treatment with chemotherapy was carried out. Bone marrow aspirate revealed cellular marrow with erythroid hyperplasia, no abnormal cells seen. One month later a follow-up CT of abdomen and pelvis revealed no evidence of any tumour remnants.

## Discussion

The gastrointestinal tract lymphoma represents 4–20% of non-Hodgkin's lymphoma and 30–45% of extranodal cases [1]. The stomach is most commonly involved, followed by the small intestine, pharynx, colon and esophagus. Lymphoma of the ileum is the most common extragastric site. Primary appendiceal lymphomas are rare occurring in 0.015% of all gastrointestinal lymphomas [2]. The median age at diagnosis for gastrointestinal tract NHL is 55 years. The disease is more common in men [3]. Acute appendicitis from luminal obstruction is the most common manifestation for most appendicular tumours [4]. Other clinical manifestations include an asymptomatic palpable mass, incidental imaging findings, intussusception, gastrointestinal bleeding, ureteral obstruction or hematuria, and increasing abdominal girth from rupture of a malignant mucocele, resulting in pseudomyxoma peritonei. Detection of these neoplasms at preoperative imaging is important because it may change the surgical approach and obviate additional surgery [4]. All cases have demonstrated prominent enlargement of the appendix with relative maintenance of its vermiform appearance. The abnormal appendix could be mistaken for an abnormal small bowel loop or an extra intestinal process such as lymphadenopathy. Diffuse mural thickening is typically hypoechoic at US and has soft-tissue attenuation at CT. The hypoechoic appearance at US may mimic the cystic dilatation seen in mucoceles of the appendix. Aneurysmal dilatation of the appendiceal lumen may be an important associated finding [3]. On CT, the diameter of the lymphomatous appendix usually measures 3 cm or larger, which is out of proportion to the size expected for non-tumoral appendicitis and should allow one to make a presumptive diagnosis of appendiceal neoplasm. Although the CT appearance is not

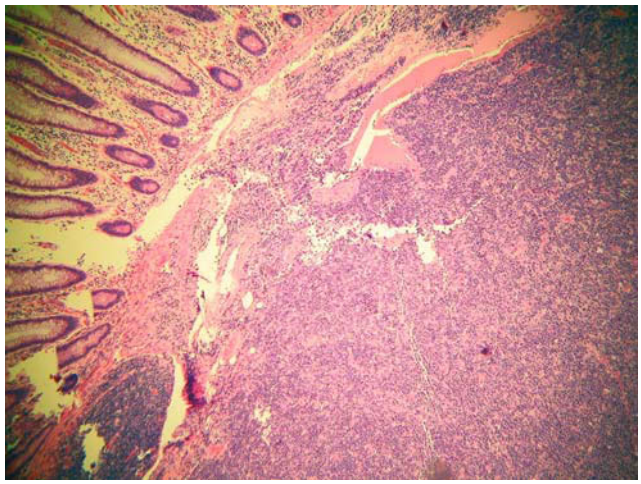

**Fig. 2** Microscopic view showing tumour on the right half and mucosa of the appendix in the left half of the slide (Haematoxylin and Eosin ×100)

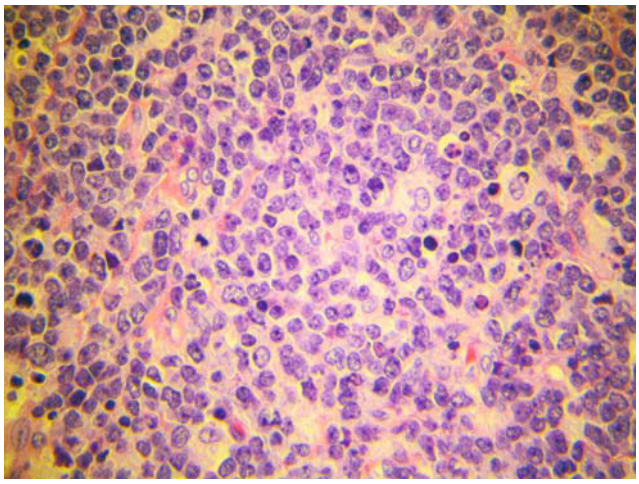

**Fig. 3** High power view showing the lymohoma cells with numerous mitotic figures and apoptotic bodies (Haematoxylin and Eosin ×400)

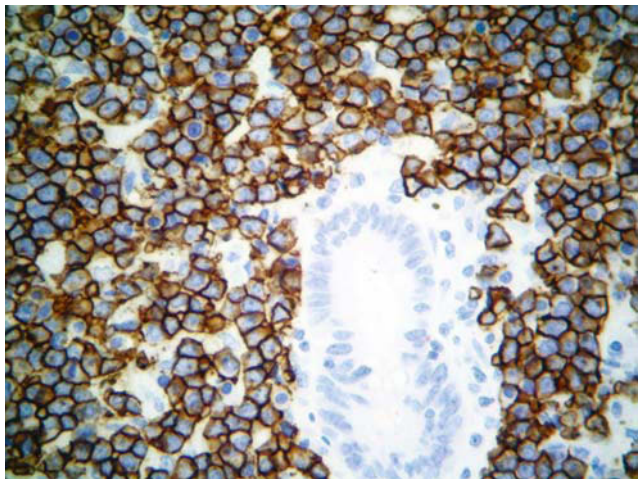

**Fig. 4** Immunohistochemistry using antibody against CD20 (B-cell marker), notice the negative staining in the mucosal crypts (Immunohistochemistry ×400)

pathognomonic and can be seen occasionally with other primary appendiceal neoplasms, NHL should be the lead differential diagnosis. Specificity for lymphoma will be increased in the setting of abdominal lymphadenopathy or aneurysmal dilatation of the appendiceal lumen [3].

Surgery alone can be considered adequate treatment for patients with low-grade NHL disease that does not infiltrate beyond the submucosa. Patients with advanced gastrointestinal NHL show a long-term outcome similar to that of patients with advanced NHL arising outside the gastrointestinal tract [5]. Primary surgical resection followed by postoperative CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone) chemotherapy showed high efficacy in intestinal diffuse large B cell lymphoma (DLBL) patients [6]. The efficacy of the surgery-chemotherapy combination in obtaining a good remission rate for localised early primary intestinal lymphoma and indicates that this combination represents the only means for managing complications [7]. Age is the only factor that revealed a statistically significant impact on outcome, while tumour burden and LDH levels represented the most important prognostic factors affecting outcome.

## Summary

This case emphasizes that careful histopathological examination of all appendectomy specimens should be mandatory. Despite the fact that lymphoma of the appendix are rare and uncommon presentation of it does not necessarily mean that it is important, our case illustrates that it must be included in the differential diagnosis of appendicular mass and that surgeons have to be aware of these conditions. Preoperative diagnosis is difficult but would improve the prognosis of appendicular lymphoma. Surgery-chemotherapy combination is the best treatment option currently available for appendiceal lymphoma.

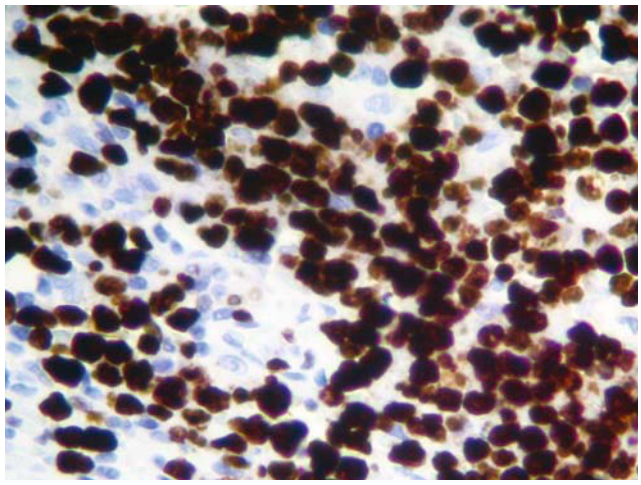

**Fig. 5** Immunohistochemistry using antibody against Ki-67 (proliferative marker) expressed in 100% of the lymphoma cells, very characterestic of Burkitt lymphoma (Immunohistochemistry ×400)

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