

# Primary Burkitt's Lymphoma Of The Appendix Presenting As Acute Abdomen: A Case Report

Maneesh Khanna<sup>1</sup>, Shankar Rao Buddhavarapu<sup>1</sup>

1. Department of Radiology, NAH, Hamad Medical Corporation, Doha; State of Qatar

\* Correspondence: Dr. Maneesh Khanna, Department of Radiology, NAH, Hamad Medical Corporation, Doha; State of Qatar, P.O Box: 3050 (✉ [mkhanna@hmc.org.qa](mailto:mkhanna@hmc.org.qa))

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## ABSTRACT

Leukemic and lymphomatous involvement of the appendix is rare and even rarer is its presentation as appendicitis. Burkitt's lymphoma is a high grade B-cell neoplasm. Its non-endemic form typically presents as abdominal mass in children. This rapidly growing tumour may cause symptoms due to mass effect or direct involvement of the bowel. Clinical presentations like acute abdomen can be secondary to intestinal obstruction, intussusception or sometimes perforation.

We describe here a case of an adult male with an unusual presentation of appendiceal Burkitt's lymphoma mimicking acute cholecystitis or appendicitis.

## CASE REPORT

### Case Report:

A 49-year-old male presented to the emergency department with right upper abdominal pain of 3 days duration. He had dull and aching pain in the same area for the last 2 to 3 months, which increased in severity and frequency over the last 1 month. There was associated nausea and vomiting. No history of fever, jaundice, loss of weight or change in bowel habit was present. There was also no history of diabetes, hypertension or renal problem.

**General Physical Examination** was unremarkable.

**Abdominal Examination:** showed tenderness in the right hypochondrium and iliac fossa. Abdomen was soft. No guarding, rigidity or rebound tenderness was noted. Bowel sounds were normal.

**Laboratory Investigation:** Hematocrit was normal. WBC count was  $10.8 \times 10^3/\mu\text{L}$  (N:  $4-11 \times 10^3/\mu\text{L}$ ). Total bilirubin was  $12\mu\text{mol/L}$  (N:  $3.5-24\mu\text{mol/L}$ ); ALT was  $30\text{U/L}$  (N:  $0-40\text{U/L}$ ); AST was  $35\text{U/L}$  (N:  $0-37\text{U/L}$ ); Alkaline phosphatase was  $99\text{U/L}$  (N:  $40-129\text{U/L}$ ), serum amylase was  $23\text{U/L}$  (N:  $13-53\text{U/L}$ ), and serum lipase was  $36\text{U/L}$  (N:  $13-60\text{U/L}$ ).

A diagnosis of acute appendicitis or cholecystitis was made clinically.

Abdominal ultrasound revealed a tubular blind ending aperistaltic hypoechoic mass arising from the right iliac fossa extending superiorly upto the subhepatic region with faintly echogenic central mucosal line indicative of its bowel origin (Fig. 1). There were echogenic foci with posterior shadowing within the superior aspect of the mass suggestive of calculi (Fig. 2). No luminal collection was present. Marked mural thickening, increased echogenicity of adjoining fat suggestive of inflammation and enlarged lymph nodes were also noted (Fig. 1, 2). Possibility of an appendicular mass lesion was suspected.

Liver, gall bladder (Fig. 3), spleen and both kidneys were unremarkable.

Plain and contrast enhanced CT of the abdomen with oral and rectal contrast was subsequently performed, which showed a long tubular soft tissue attenuation enhancing mass (Plain and contrast CT attenuation of 35HU and 62HU respectively) in the right anterior pararenal space, posterior and lateral to cecum and ascending colon. The lesion was extending superiorly from right iliac fossa to the subhepatic region. The vermiform morphology was maintained. Lumen

was not visualized. The superior aspect of the mass showed another tubular hypodense structure in continuity with the primary mass with hypodense centre, enhancing wall and luminal calcific density (Fig. 4a, 4b, 4c, 4d, 7a, 7b). There was associated cecal wall thickening and beaking (Fig. 6). In addition extensive perilesional mesenteric fat stranding with thickened pararenal fascia and multiple enlarged draining lymph nodes (Fig. 5) were also seen.

The location of the mass and relative preservation of the vermiform morphology in spite of the marked enlargement suggested the possibility of a primary appendicular lesion. An additional tubular lesion at the superior aspect of the mass was considered preoperatively to be due to changes of post obstructive appendicitis involving predominantly the tip of the appendix.

Liver, gall bladder, pancreas, spleen, both kidneys, and retroperitoneum were unremarkable.

Diagnostic laparoscopy was subsequently performed which revealed large appendicular mass invading cecum with adhesion to the undersurface of liver. The mass measured 7 x 5 x 3.5 cm, was hard in consistency with large mesenteric lymph nodes (largest measuring 3 x 3 cm sized) along right colic artery. Appendix showed inflammation.

Laparotomy with right hemicolectomy and side to side ileotransverse anastomosis was performed. Complete perilesional lymph node resection was also performed.

Macroscopic examination of the specimen showed a dilated firm sausage like appendix measuring 3 x 3 x 7.5 cm. Cross section revealed a firm fleshy mass totally replacing the appendiceal lumen reaching up to the serosa. No lumen was identified. The mass was bulging under the mucosa into the cecal lumen, which seemed uninvolved. Mesoappendix had multiple enlarged lymph nodes. Microscopic examination revealed sheets and cords of highly neoplastic lymphoblasts with high N/C ratio, hyperchromasia, apoptotic bodies, and nuclear moldings with several mitotic figures. Numerous tingible body macrophages giving starry sky appearance were seen (Fig. 8a). Immunoperoxidase stains were positive for CD20, CD79a, CD10 and KI-67 (expressed in 100% of lymphoma cells) (Fig. 8b) and negative for CD3, CD30, CD43, Bcl-2 and MUM-1.

Final diagnosis of appendicular Burkitt's lymphoma with lymph node involvement was made.

The patient was put on Chemotherapy (CODOX-M / IVAC regime) and has received 2 cycles. Follow up CT chest and abdomen over 10 months showed no local recurrence, metastasis or lymphadenopathy.

### DISCUSSION

Burkitt's lymphoma is a highly aggressive non-Hodgkin B- cell lymphoma mostly presenting as extranodal disease. Children and immunocompromised patients are most often affected by the disease (1).

Burkitt's lymphoma is classified into three clinical forms:

- a) Endemic form is common in Africa and presents mostly as rapidly growing tumour involving facial bones.
- b) Non-endemic form, typically presents as abdominal mass or ascites.

c) The third form occurs in immunocompromised patients and it is commonly seen in patients with HIV/AIDS. It most commonly presents as diffuse lymphadenopathy (1).

Malignant lymphoma comprises approximately 1 to 4% of all the gastrointestinal malignant neoplasms (2). Primary appendicular lymphomas are even rarer and often a postoperative diagnosis with the reported incidence of approximately 0.015% in a large series of 71,000 human appendicular specimens evaluated (3).

Muller has reported four cases of leukemia or lymphoma presenting as acute appendicitis (4). Domizio has described 119 cases of primary small bowel lymphomas some of which produced acute abdomen (5). Chirletti has described 18 patients with hematologic disorders presenting as acute abdomen, 5 with acute appendicitis, 3 had necrotizing enterocolitis, 3 had hemoperitoneum, 3 had cholecystitis, 2 had splenic infarcts and 2 had bowel obstruction(6).

Although, there are numerous reports of an acute surgical presentation of appendicular Burkitt's lymphoma, all the cases have been reported in children and young adults. Information of appendiceal lymphoma is limited to scattered case reports and even fewer reports focus on imaging features of the disease.

Although there are no classical imaging features of appendiceal lymphoma, enlargement of appendix beyond 15 mm in diameter on CT should be viewed with suspicion(7). A diameter above 2.5 cm should be even more concerning (8). The presence of periappendiceal stranding on CT can be due either to inflammatory changes from secondary appendicitis or direct serosal extension of lymphomatous cells.

Differential diagnosis of a diffusely enlarged appendix can be: 1. Benign conditions like appendicitis in which the diameter of the appendix usually does not exceed 15mm. (7).

2. Mucinous epithelial neoplasms generally are associated with mucocoeles, seen on CT as cystic dilatation of the lumen with or without calcifications. Areas of focal nodular solid enhancement of the wall favours presence of mucinous cystadenocarcinoma. (9).

3. Neuroendocrine tumors (like Carcinoid, Paraganglioma and Ganglioneuroma) show infiltrative growth pattern. Carcinoid typically shows a small (usually less than 1 cm sized) mass involving the distal appendix rather than circumferential enlargement (10). The other neuroendocrine tumors are rare but may resemble enlargement of the appendix by lymphoma (11).

Our case was unusual in that the primary appendiceal Burkitt's lymphoma in an adult patient to our knowledge has not been reported before. It's unusual high location and acute presentation made it a clinical dilemma mimicking cholecystitis.

### TEACHING POINT

CT feature showing marked mural thickening of the appendix (more than 1.5cm) with enlarged lymph nodes should be viewed with suspicion for primary appendiceal malignancy.

ABBREVIATIONS

WBC: White blood cell  
 CT: Computed Tomography  
 N/C: Nuclear-cytoplasmic ratio  
 CD: Cluster differentiation  
 Bcl-2: B cell leukemia-lymphoma 2  
 KI-67: Histochemical marker for cell proliferation  
 MUM-1: Melanoma associated antigen mutated 1  
 ALT: Alanine aminotransferase  
 AST: Aspartate aminotransferase  
 N: Normal  
 HIV: Human immunodeficiency virus  
 AIDS: Acquired immune deficiency syndrome  
 CODOX-M/IVAC: Cyclophosphamide, Doxorubicin,  
 High-dose Methotrexate / Ifosfamide, Etoposide and  
 High dose Cytarabine

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FIGURES



**Figure 1:** Longitudinal ultrasound of the right iliac fossa/right lumbar region shows a long tubular blind ending mass with marked mural thickening, irregular outline and a faint central echogenic line likely mucosa (white arrow). Increased fat echogenicity is seen posterior to the lesion suggestive of surrounding inflammation (black asterisk).



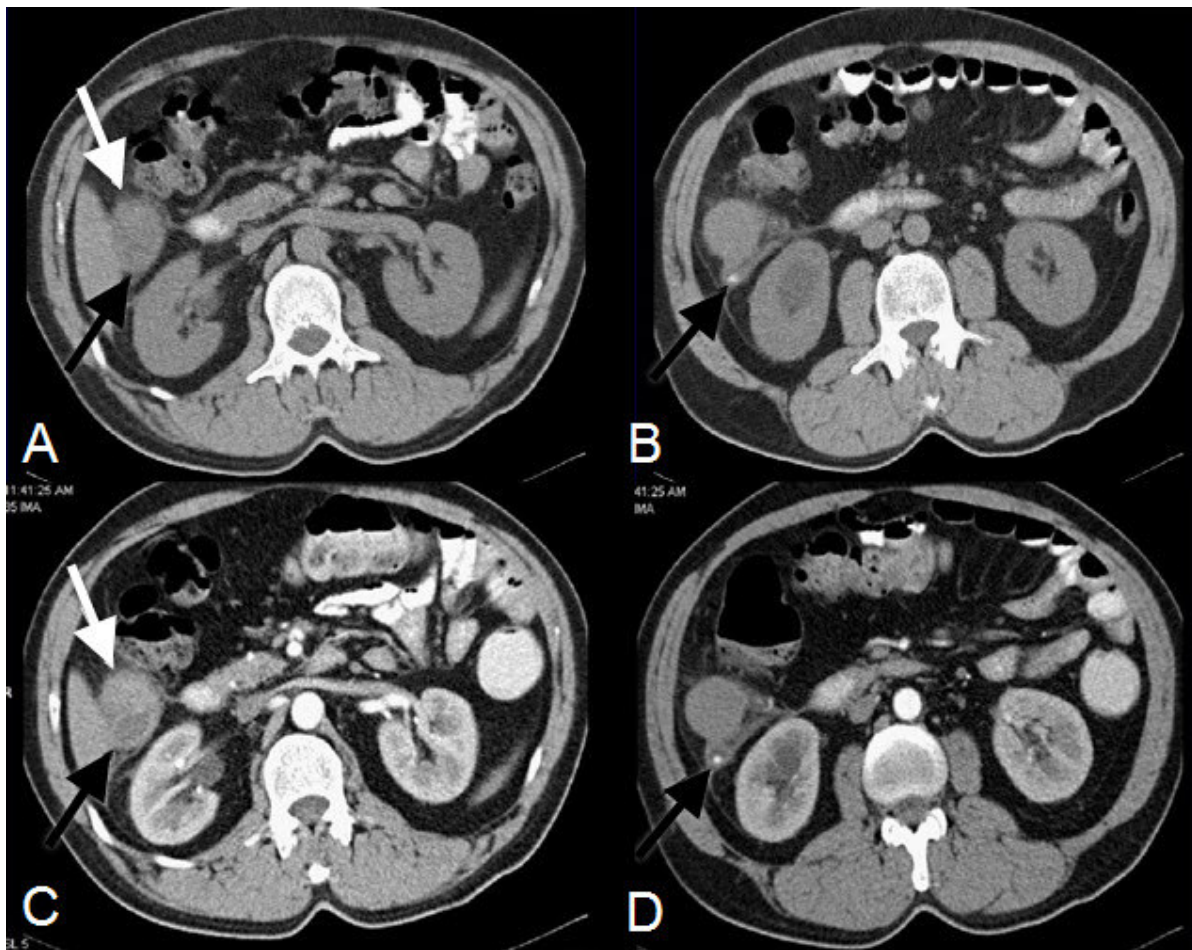
**Figure 2:** Longitudinal ultrasound of the right iliac fossa/right lumbar region shows calculi in the superior aspect of the lesion (bold white arrow). Enlarged oval perifocal lymph node (black thin arrow) posterior to the tubular mass (white thin arrow) is also noted.



**Figure 3:** Ultrasound of the right upper abdomen shows normal gall bladder lumen and wall thickness. No pericholecystic fluid is noted.

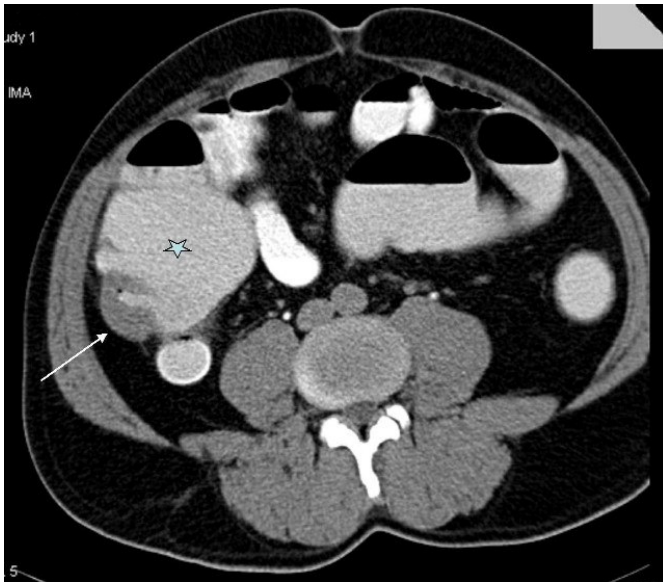


**Figure 5:** Axial contrast enhanced CT image reveals the rounded soft tissue density mass in the right anterior pararenal space (long white arrow) with surrounding fat stranding, pararenal fascia thickening (short white arrow) and multiple enlarged draining lymph nodes (broken white arrow). Note is made of a right renal cyst.



**Figure 4:** Axial plain ( 4a, 4b) and contrast enhanced (4c, 4d) CT at the level of the kidneys reveal an enhancing rounded soft tissue attenuation mass (white arrow) with tubular hypodense area showing peripheral mural enhancement and calcification (black arrow). Note is made of surrounding fat stranding and loss of fat planes with adjacent liver.

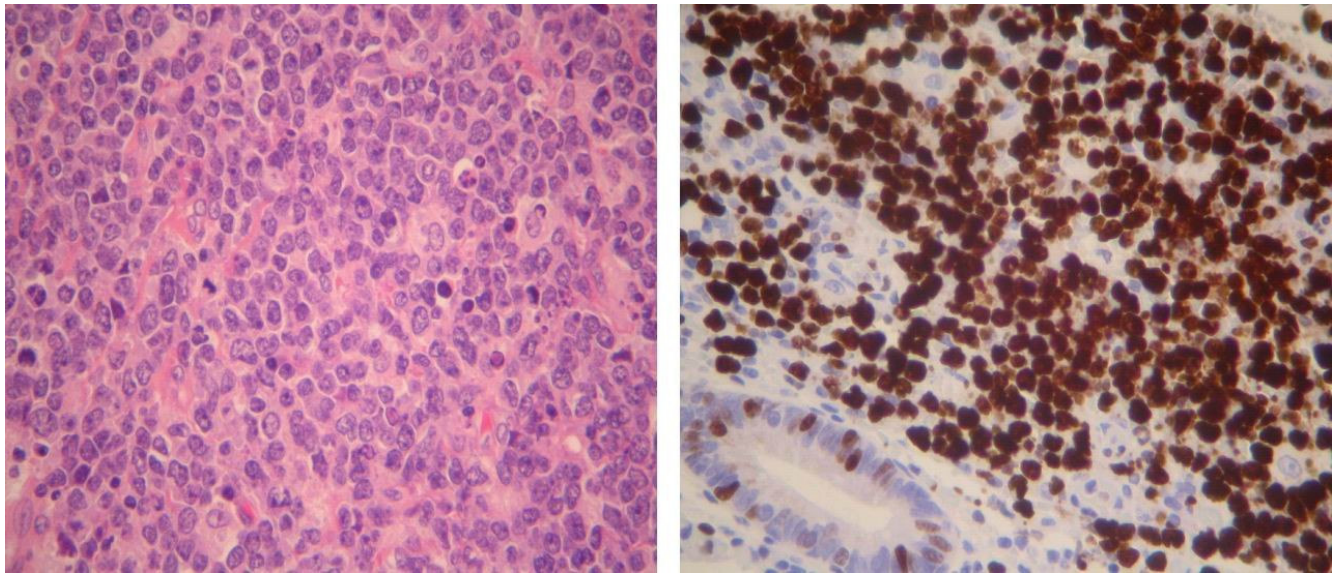




**Figure 6:** Axial contrast enhanced CT image with rectal contrast reveals mural thickening (white arrow) lateral wall of cecum (asterix).



**Figure 7:** Coronal reformatted images of a contrast enhanced CT of the abdomen reveal large tubular soft tissue mass (left, white arrow) extending from cecum (asterix) superiorly with non visualized lumen within the mass. There are multiple enlarged perifocal lymph nodes (left, broken white arrow) and surrounding fat stranding. A tubular hypodense area on top of this mass with calcific density is noted. (right, bold black arrow).



**Figure 8:**

Left image: High power view showing lymphoma cells with numerous mitotic figures and apoptotic bodies (Hematoxylin & Eosin x400). Sheets and cords of highly neoplastic lymphoblasts with high N/C ratio, hyperchromasia, apoptotic bodies, and nuclear moldings with several mitotic figures. Numerous tingible body macrophages giving a starry sky appearance.

Right image: Immunohistochemistry using antibody against Ki-67 (proliferative marker - expressed in 100% of lymphoma cells), very characteristic of Burkitt's lymphoma (Immunohistochemistry x400).

KEYWORDS

Lymphoma; Appendicitis; Cholecystitis; Burkitt's lymphoma; Appendix; CT

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