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Mantle cell lymphoma presenting as acute appendicitis

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ABSTRACT

INTRODUCTION: Most cases of appendicitis associated with lymphoma reported in literature are in patients with Burkitt's or large B-cell lymphoma. Mantle cell lymphoma only makes up 4% of all lymphoma cases in the U.S. We report a case of a patient with mantle cell lymphoma presenting with acute appendicitis.

PRESENTATION OF CASE: A 75 year old male with a history of left cervical lymphadenopathy biopsied to be mantle cell lymphoma presented with right lower abdomen pain for 3 days. An outpatient CT scan revealed acute appendicitis. Laparoscopic appendectomy was performed without any complication. The histologic examination showed mantle cell lymphoma occluding the lumen of appendix.

DISCUSSION: Typically, appendicitis is caused by obstruction of the lumen of appendix by fecalith or lymphoma. A previously reported case of a patient with mantle cell lymphoma who developed appendicitis received chemotherapy before appendectomy. The author could not determine how mantle cell lymphoma contributed to appendicitis because the lumen of appendix was not occluded by the lymphoma, likely from cytoreduction from chemotherapy. We have a patient with mantle cell lymphoma before the patient received chemotherapy who presented with appendicitis. The appendiceal specimen shows the lumen filled with mantle cell lymphoma.

CONCLUSION: This is a rare case of mantle cell lymphoma causing obstruction of appendiceal lumen and subsequently appendicitis, unaffected by chemotherapy.

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1. Introduction

Lymphoma infiltrating the appendix and causing appendicitis has been reported in literature, with reports usually in children with Burkitt's lymphoma and large B-cell lymphoma.^{1–6} However, the incidence of lymphoma causing appendicitis is such a rare event that not much is known about pathogenesis. Of all lymphoma prevalence, a subtype of lymphoma known as mantle cell lymphoma comprises only 4% in the U.S. and 7–9% in Europe.⁷ As rare as for any lymphoma to cause appendicitis, it is rarer for mantle cell lymphoma to be found in appendix specimen after an appendectomy. There is only one reported case in the literature of mantle cell lymphoma associated with appendicitis.⁸ We are reporting a case of a patient with mantle cell lymphoma before a chemotherapy treatment presenting with acute appendicitis.

2. Presentation of case

A 75 year old male with a history of mantle cell lymphoma diagnosed 3 years prior went to his primary medical doctor with a complaint of right lower quadrant abdominal pain for 3 days. The primary medical doctor advised the patient to obtain a CT scan of abdomen and pelvis at an outpatient facility. The pain started as a vague right lower abdominal pain that worsened as time has passed. The patient denied any nausea, vomiting, or anorexia. He stated that he had a low grade fever a couple of days ago. After the CT scan result, the doctor told the patient to go to an emergency room. Physical examination was significant for mild tenderness in right lower quadrant of abdomen but was soft and with no guarding. The patient had a significant lymphadenopathy in his left side of neck. White blood cell count was within normal with a slight shift. The outpatient CT scan showed acute appendicitis with no signs of perforation and prominent abdominal and pelvic lymph nodes (Fig. 1). For acute appendicitis, the patient was taken to OR for laparoscopic appendectomy on the same day.

In May 2011, the patient presented with left cervical lymphadenopathy for which he was taken to OR for lymph node biopsy. The lymph node specimen revealed mantle cell lymphoma with immunohistochemical stains positive for CD20, Pax-5, CD5, CD43, BCL-1, and BCL-2 while negative for CD10 and BCL-6.

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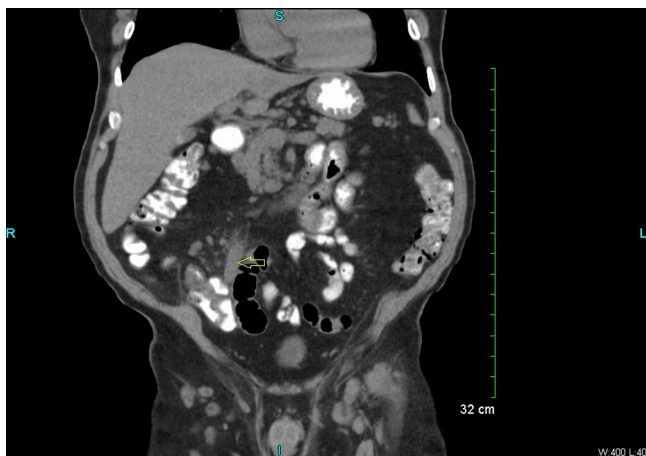


Fig. 1. CT scan of abdomen and pelvis in a coronal view. Arrow shows an acutely inflamed appendix with no sign of perforation.

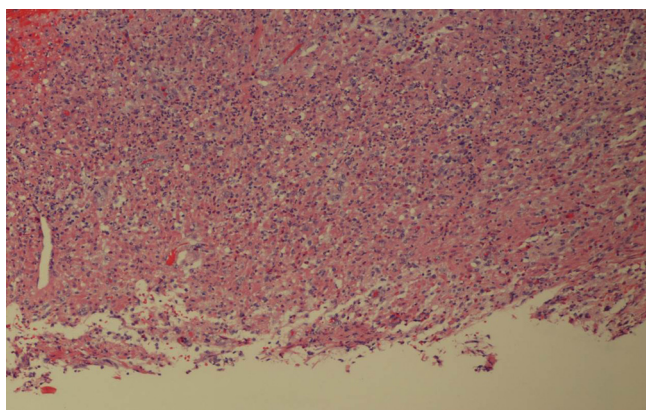


Fig. 2. Cross section of appendix showing areas of acute appendicitis (10× objective).

A laparoscopic appendectomy was performed without any complication. The appendix was adhered to terminal ileum and omentum. Careful adhesiolysis was carried out to free the appendix. Using a LigaSure device to divide the mesoappendix, the base of appendix was visualized and the appendix was transected using an EndoGIA stapling device. The patient recovered post-operatively without any complication and was discharged on the post-op day 2. The pathology of appendix specimen revealed acute appendicitis and periappendicitis with the lumen of appendix filled with and appendiceal wall replaced with small to intermediate size lymphocytes (Fig. 2). The immunohistochemical stains were positive for CD20, Pax-5, BCL-1 and BCL-2 and negative for CD3, CD5, CD10, and BCL-6, consistent with mantle cell lymphoma (Figs. 3–6).

On the follow up appointment in office, the patient is scheduled to meet his oncologist to have a regimen of chemotherapy for the treatment of mantle cell lymphoma.

3. Discussion

Mantle cell lymphoma is an aggressive subtype of non-Hodgkins B-cell lymphoma. The pathogenesis of lymphoma causing appendicitis has not been elucidated by any high quality study. Being that the number of lymphoma related appendicitis is so few that mostly case reports are found in literature.^{1,8,9} How lymphoma causes appendicitis is not known definitively. Case reports show evidences that intravascular tumor invasion leading to ischemia of appendix and subsequently appendicitis or obstruction of appendiceal lumen

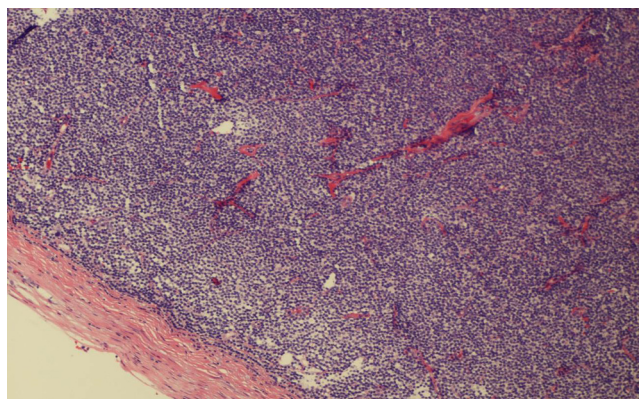


Fig. 3. Mantle cell lymphoma involving appendix-lumen filled with monotonous atypical lymphoid infiltrate (10× objective).

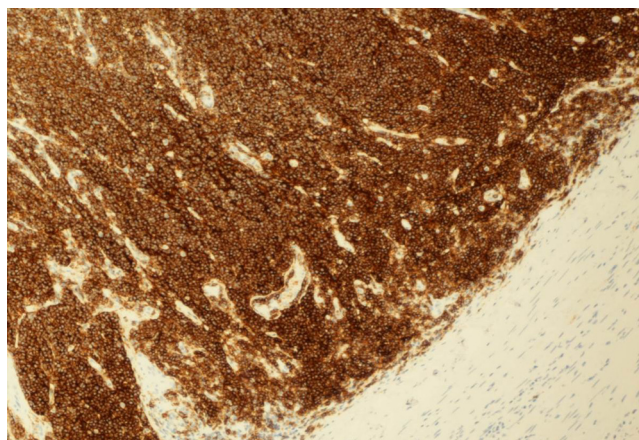


Fig. 4. Appendiceal specimen stained with immunohistochemical stain CD 20 (B-cell) diffusely positive (10× objective).

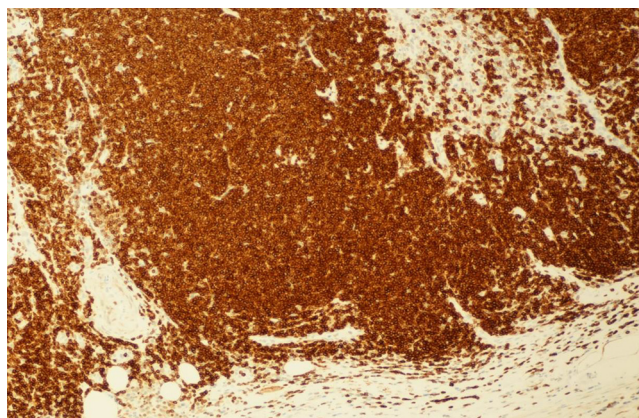


Fig. 5. Appendiceal specimen stained with immunohistochemical stain BCL-2 diffusely positive (10× objective).

leading to appendicitis.^{2,4,5,9} A common extranodal manifestation of lymphoma is in gastrointestinal tract, other than bone marrow or spleen, due to gastrointestinal tract containing numerous lymphocytes in mucosa.⁷ Finding of multiple lymphomatous polyposis in gastrointestinal tract is closely related to mantle cell lymphoma.^{7,10} Enlarged lymphoma in the lumen of appendix or extrinsic compression on appendix by lymphomatous polyposis can obstruct the lumen of appendix and cause appendicitis. In a previous case report by Linden, a patient with a known history of mantle cell lymphoma for which the first of six cycles of chemotherapy has been

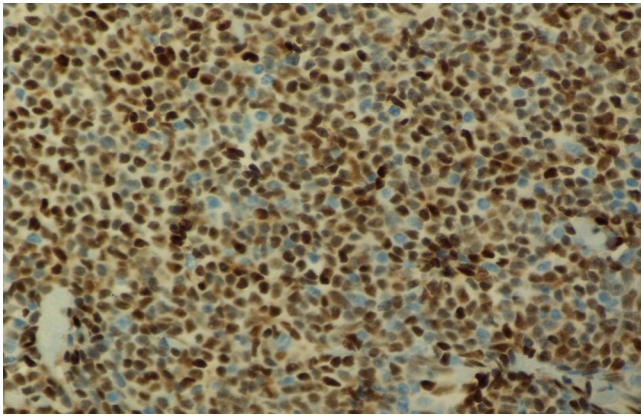


Fig. 6. Appendiceal specimen stained with immunohistochemical stain BCL-1 diffusely positive (40× objective).

given presented 17 days after the chemotherapy has started with an acute appendicitis. The histologic examination after the appendectomy did not reveal mantle cell lymphoma to be extensively encompassing or occluding the lumen of specimen. The author surmised that a reason for the finding of less than expected invasion of lymphoma in the appendiceal specimen could be due to chemotherapy given to the patient prior to appendectomy. The author could not conclude how mantle cell lymphoma caused appendicitis, but suspect that cytoreduction of lymphoma invasion in the appendiceal wall from chemotherapy could have caused perforation of appendix. We have a patient presented with acute appendicitis before initiation of chemotherapy. The appendiceal specimen after appendectomy showed completely occluded lumen by mantle cell lymphoma. This case is unique in that this is the only reported case of mantle cell lymphoma presenting as acute appendicitis before initiation of chemotherapy showing an appendiceal specimen lumen completely filled with mantle cell lymphocytes. Although not conclusive, we have a glimpse of pathogenesis of mantle cell lymphoma resulting in appendicitis.

Key learning points

- Mantle cell lymphoma associated with appendicitis is extremely rare.
- Obstruction of the lumen of appendix by mantle cell lymphoma predisposes to the development of appendicitis.
- This case is the first mantle cell lymphoma to cause appendicitis, unaltered by introduction of chemotherapy.

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4. Conclusion

Classically appendicitis is caused by obstruction of lumen of appendix by a fecalith leading to inflammation of appendix. Unlike the previously reported case of mantle cell lymphoma with appendicitis affected by chemotherapy, this interesting case shows mantle cell lymphoma occluding the lumen of appendix unaffected by chemotherapy. Mantle cell lymphoma behaves as an obstructing mass in appendiceal lumen, similar to a fecalith, leading to appendicitis.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interest

The authors have no conflict of interest.

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None.

Ethical approval

Not applicable.

Author contributions

Max Chae – a main author of this case report.

Sampath Kumar – a co-author of this case report.

Muhammad Cheema – a pathologist who prepared histologic specimens and provided images of histologic slides.