

Primary Lymphoma of Appendix – A Case Report with Review of Literature

Nanda Patil¹, Shoaib Khoja², Vaidehi Dharmesh Nagar^{2*}, Sonam Billawaria²

¹Professor, Department of Pathology, Krishna Institute of Medical Sciences, Deemed University, Karad, NH4, Pune - Bangalore Highway, Dist.Satara, Agashivnagar, Malkapur, Maharashtra 415539, India

²Tutor, Department of Pathology, Krishna Institute of Medical Sciences, Deemed University, Karad, NH4, Pune - Bangalore Highway, Dist.Satara, Agashivnagar, Malkapur, Maharashtra 415539, India

DOI: [10.36347/sjmcr.2021.v09i01.007](https://doi.org/10.36347/sjmcr.2021.v09i01.007)

| Received: 28.12.2020 | Accepted: 12.01.2021 | Published: 14.01.2021

*Corresponding author: Vaidehi Dharmesh Nagar

Abstract

Case Report

Introduction: Primary lymphoma of appendix is extremely rare. Most of them are diagnosed accidentally from appendectomy specimens removed for appendicitis. **Case Report:** We report a case of primary Non Hodgkin's Lymphoma of appendix in a 58 year old female patient who presented with periumbilical pain in abdomen with history of weight loss. On the basis of radiological examination she was diagnosed as mucocele of appendix with soft tissue density of ileum. Right hemicolectomy was done and specimen was received for histopathological examination which revealed non Hodgkin's lymphoma involving entire appendix. **Conclusion:** Primary appendiceal lymphoma is very rare and difficult to diagnose clinically. Histopathological examination of all appendectomy specimens is essential for diagnosis and further management of patients.

Keywords: Appendix, Non Hodgkin's Lymphoma, Primary.

Copyright © 2021 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

BACKGROUND

Primary lymphoma of gastrointestinal tract is rare and accounts for 1-4% of all tumors of gastrointestinal tract [1]. Primary lymphoma of appendix is extremely rare, the incidence of this is 0.015% of all gastrointestinal lymphomas [2]. We present a case of primary diffuse large B cell lymphoma of appendix in a 58 year old female patient, clinically presented with acute appendicitis.

CASE REPORT

A 58 years old female patient presented with periumbilical pain in abdomen and constipation since 10 days. She gave history of weight loss. Hematological investigations revealed anemia (Hb -10.4 gm/dl). Total and differential count were normal. Chest X – ray findings were within normal limits.

CT Abdomen and pelvis revealed a lesion involving appendix and projecting into caecal lumen. Another well defined globular intraluminal exophytic soft tissue density involving ileum was also noted along with multiple enlarged mesenteric lymph nodes.

The patient was diagnosed clinically as mucocele of appendix. Liver and spleen were normal and there was no palpable lymphadenopathy.

Right hemicolectomy was performed and specimen was received for histopathological examination.

HISTOPATHOLOGICAL EXAMINATION

A specimen of right hemicolectomy was received consisting of part of ileum, caecum, appendix and a part of ascending colon with attached mesentery. Appendix was enlarged measuring 6.8 cm in length and 2.5 cm in diameter, cut section of entire appendix revealed extremely thickened wall which was grey white, firm and fleshy. Base of appendix from mucosal surface also showed fleshy polypoidal mass measuring 2.5 x 2 x 1.5 cm protruding in the lumen of caecum, cut section of which was grey white and fleshy (Fig-1).

In addition another grey white polypoidal mass was seen protruding from the mucosal surface of terminal ileum measuring 7.5 x 5 x 3.8 cm, cut section of which was grey white and fleshy. Attached mesocolon revealed multiple matted lymph nodes, on cutting open revealed fleshy appearance (Fig-2).

Microscopy of tumor mass revealed large neoplastic lymphoid cells arranged in sheets, showing high nuclear cytoplasmic ratio, enlarged vesicular nuclei with heterogenous chromatin and scanty cytoplasm (Fig-3).

The tumor was involving whole thickness of appendix. Multiple sections from mass in the terminal ileum and lymph nodes in the mesocolon revealed

similar microscopic features. Immunohistochemistry profile of tumor revealed positivity for CD 20, BCL 6, MUM 1, Ki67 proliferative index was 90 % (Fig-4). The negative markers were CD 3, CD 10 and Cyclin D1.

Considering these features, the diagnosis was given as Primary diffuse Large B cell Lymphoma arising from appendix.



Fig 1. A - Enlarged and thickened appendix.
B - Cut section - entirely involved by tumor showing grey white fleshy appearance.



Fig 2. - Multiple enlarged lymph nodes in attached mesocolon. Cut section - grey white and fleshy.

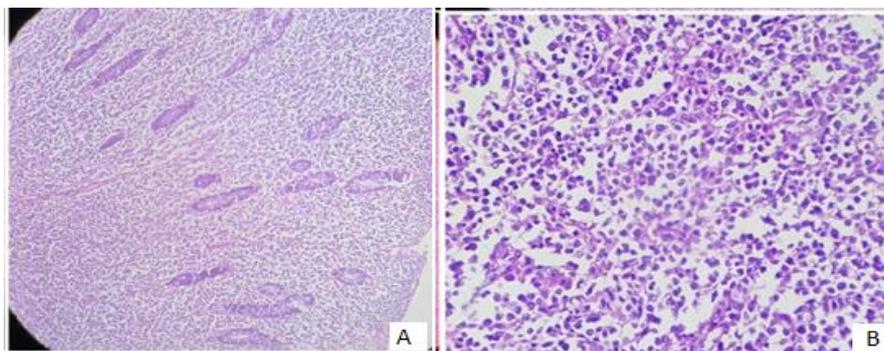


Fig 3. A - Infiltration of neoplastic lymphoid cells in appendix (100 X H&E)
B - Neoplastic lymphoid cells (400 X H&E)



Fig. 4

Fig. 4 - Ki67 index of tumor > 90%

DISCUSSION

Primary lymphoma of the intestine is almost a Non Hodgkin's Lymphoma (NHL) [3].

Diagnostic criterias for primary gastrointestinal lymphoma include:

1. Absence of palpable lymphadenopathy on clinical examination.
2. Absence of mediastinal lymphadenopathy on chest wall X-ray.
3. Normal differential WBC count
4. Disease confined to the intestine and adjacent lymph nodes without hepatosplenomegaly [4].

Our case also revealed Non-Hodgkin's Lymphoma confined to appendix with enlarged lymph nodes in mesocolon with absence of palpable lymph nodes, normal total and differential WBC count and no evidence of mediastinal lymphadenopathy. Radiological features can give clues towards diagnosis of NHL involving gastrointestinal tract. These are multifocal appearance, absence of obstruction despite of large tumor mass and absence of hypervascularization.

The lesions can be polypoid masses or circumferential infiltrations, ulcerated lesions or nodules [5]. Primary lymphoma of appendix is extremely rare. In our case tumor was in the form of polypoid mass with multifocal appearance involving entire appendix with its base and terminal ileum.

Diffuse large B cell Lymphoma and MALT Lymphoma are the two most common types of primary lymphomas of the intestine. Diffuse large B cell lymphoma can present grossly as polypoidal or ulcerative form, diffusely infiltrating tumor or mixed pattern [6]. In our case, the tumor was showing mixed pattern i.e. polypoid growth as well as the tumor was infiltrating entire wall of appendix.

Computed Tomography can be helpful for diagnosis of primary lymphoma of appendix. The clues to the diagnosis are diffuse appendiceal enlargement beyond 1.5 cm in diameter. Neuroendocrine tumors can also show similar pattern but usually tip of appendix is involved in such case. Abdominal lymphadenopathy associated with diffusely enlarged appendix helps in the diagnosis of lymphoma of appendix [7].

Clinical presentation of primary lymphoma of appendix is similar to other primary tumors of appendix in the form of symptoms of acute appendicitis, right iliac fossa pain or palpable mass in right lower quadrant [8], similar presentation was seen in our case. Surgical resection with postoperative chemotherapy is treatment of choice for primary lymphoma of appendix [9].

CONCLUSION

Primary lymphoma of appendix is very rare and manifests clinically with similar signs and symptoms as acute appendicitis. Preoperative diagnosis of this neoplasm is very difficult. CT scan findings of diffuse enlargement of appendix with abdominal lymphadenopathy may lead to suspicion of lymphoma of appendix. Histopathological examination of all appendectomy specimen is essential for diagnosis and management of patients.

REFERENCES

1. Shiwani MH. Primary malignant lymphoma of the appendix associated with acute appendicitis. *Journal of the College of Physicians and Surgeons--Pakistan: Jcsp*. 2006 Jan 1;16(1):79-80.
2. Radha S, Afroz T, Satyanarayana G. Primary marginal zone B-cell lymphoma of appendix.

- Indian Journal of Pathology and Microbiology. 2008 Jul 1;51(3):392-394.
3. Ponce J, Garangou AC, García FG, editors. Tratamiento de las enfermedades gastroenterológicas. Asociacion Española de Gastroenterología; 2011; 327-34.
 4. Dawson IM, Cornes JS, Morson BC. Primary malignant lymphoid tumours of the intestinal tract. Report of 37 cases with a study of factors influencing prognosis. *British Journal of Surgery*. 1961 Jul;49(213):80-9.
 5. Frampas E. Lymphomas: Basic points that radiologists should know. *Diagnostic and interventional imaging*. 2013 Feb 1;94(2):131-44.
 6. Vetro, C., Romano, A., Amico, I., Conticello, C., Motta, G., Figuera, A., ... & Di Raimondo, F. (2014). Endoscopic features of gastro-intestinal lymphomas: from diagnosis to follow-up. *World Journal of Gastroenterology: WJG*, 20(36):12993-13005.
 7. Pickhardt PJ, Levy AD, Rohrmann Jr CA, Abbondanzo SL, Kende AI. Non-Hodgkin's lymphoma of the appendix: clinical and CT findings with pathologic correlation. *American Journal of Roentgenology*. 2002 May;178(5):1123-7.
 8. Ruoff C, Hanna L, Zhi W, Shahzad G, Gotlieb V, Saif MW. Cancers of the appendix: review of the literatures. *ISRN oncology*. 2011;2011.
 9. Lee J, Kim WS, Kim K, Ahn JS, Jung CW, Lim HY, Kang WK, Park K, Ko YH, Kim YH, Park C. Prospective clinical study of surgical resection followed by CHOP in localized intestinal diffuse large B cell lymphoma. *Leukemia research*. 2007 Mar 1;31(3):359-64.