



# A Rare Case of Mantle Cell Lymphoma Initially Presenting with Hemoperitoneum

Ola Al-Jobory\*; Anees Muhammed; Yasir Al-Hilli; Basheer Mohammed; Asm Islam

Department Internal Medicine, Texas Tech University of Health and Sciences, USA.

## \*Corresponding Author(s): Ola Al-Jobory

Department Internal Medicine, Texas Tech University of Health and Sciences, USA.

Email: ola.al-jobory@ttuhsc.edu

Received: Feb 16, 2022

Accepted: Mar 09, 2022

Published Online: Mar 14, 2022

Journal: Journal of Clinical Images

Publisher: MedDocs Publishers LLC

Online edition: <http://meddocsonline.org/>

Copyright: © Al-Jobory O (2022). *This Article is distributed under the terms of Creative Commons Attribution 4.0 International License*

**Keyword:** MCL; Hodgkin Lymphoma; Non-Hodgkin Lymphoma; Cancer.

**Abbreviation:** MCL: Mantle cell lymphoma; HL: Hodgkin Lymphoma; NHL: Non-Hodgkin Lymphoma.

## Introduction

B-cell non-Hodgkin lymphoma is a cancer of the immune system. MCL is one of the 70 different subtypes of NHL, which is abnormal B cells grow out of control and may crowd out healthy B cells in the lymph nodes, bone marrow, and other organs. It is a relatively uncommon form of non-Hodgkin lymphoma presents in 2% of cases only, with incidence of 1/200000 patient a year [2], more predominant in male than female around age 65.

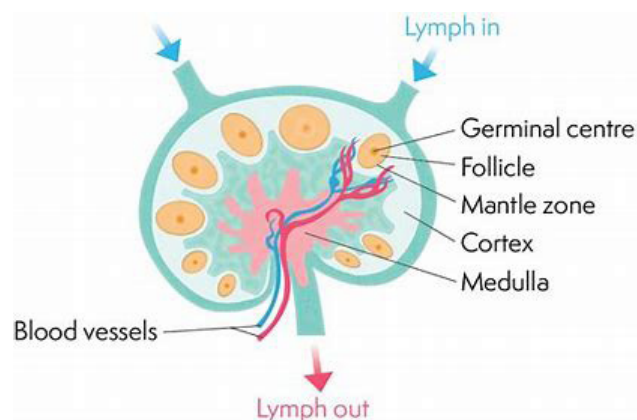
The name “mantle cell lymphoma” comes from the fact that the abnormal B cells originate in the mantle zone (the outer edge) of the lymph node (Figure 1).

## Abstract

Lymphoma is the general name for many related subtypes of cancer that arise from a type of white blood cell called a “lymphocyte.” Lymphoma is divided into two major categories: Hodgkin Lymphoma (HL) and Non-Hodgkin Lymphoma (NHL).

NHL is progressive clonal expansion of B cells or T cells and/or NK cells arising from an accumulation of lesions affecting proto-oncogenes or tumor suppressor genes, resulting in cell immortalization leading to accumulation in the lymph nodes [1].

MCL is a subtype of NHL presents usually with B symptoms but can have atypical presentation, treatment with Chemotherapy might be helpful but still has poor prognosis.



**Figure 1:** Mantle cell lymphoma.

**Cite this article:** Al-Jobory O, Muhammed A, Al-Hilli Y, Mohammed B, Islam A. A Rare Case of Mantle Cell Lymphoma Initially Presenting with Hemoperitoneum. J Clin Images. 2022; 5(1): 1121.



## Prognosis

MCL has poor prognosis despite the treatment, the median survival time is approximately 3 years (range, 2-5 y); the 10-year survival rate is only 5-10% [3]. MCL usually presents with

- Swollen lymph nodes
- An enlarged spleen causing abdominal discomfort
- anemia and fatigue
- GI disturbances-nausea, vomiting, and loss of appetite
- "B" symptoms: fever, night sweats, and significant weight loss

Atypical presenting symptoms includes abdominal distention associated with abdominal discomfort. We are reporting a case of 65 years old gentleman presenting with initial symptoms of abdominal distension and was diagnosed with Mantle cell lymphoma.

## Case

65 years old gentleman with no past medical history presented to the hospital with abdominal discomfort with distention. Patient denied alcohol use; he also denied hep C or B previously.

He stated that he lost 25 pounds intentionally as he changed his diet to healthy diet but he denied any vomiting, he had mild nausea but no sweating, no diarrhea and no bleeding or melena.

Examination revealed the abdomen was distended but not tender no guarding. Bowel sounds were present.

Lab results showed: Upon presentation, his WBC been 19,000, anion gap metabolic acidosis, AKI with creatinine 3.2, hemoglobin dropped to 7, Peripheral smear revealed smudge cell and spherocytosis. Reticulocyte count was 3, platelets 69, lipase and amylase within normal limit but AST is elevated.

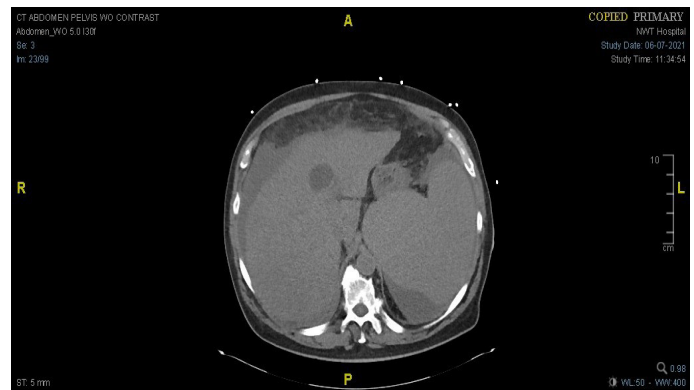
CT abdomen\* showed nodular liver consistent with cirrhosis and splenomegaly, portal hypertension and moderate size ascites with Retroperitoneal Lymphadenopathy. Paracentesis was done and 3.5 L of bloody fluid was drained and sent for analysis, which came back WBC 4600, RBC of 1,450,000, pH of 8000, albumin of 2.4 and triglyceride of 255. Hepatitis panel was negative. USG of the abdomen: cirrhotic morphology with multiple cyst largest measuring 3.8 x 2.6 cm and hepatomegaly (**Figure 2**).

CT biopsy of the abdomen of large retroperitoneal lymph node also done.

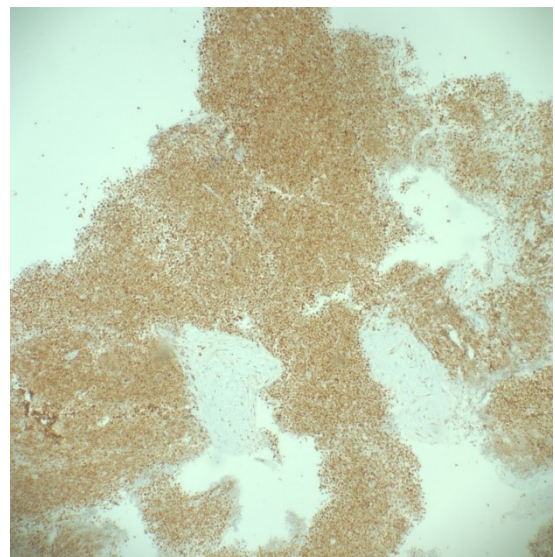
During that time, the patient breathing deteriorated, and he had more abdominal distention and they had to do another tap and they took 3 L of bloody fluid. CEA was 8.6, alpha-fetoprotein 3.7; CEA 19-9 is 105. Lymph node biopsy confirmed mantle cell lymphoma, CD5 positive. Pleural and ascitic fluid flow cytometry also positive for CD5\*\* without CD 23, positive B-cell lymphoproliferative disorder. Immunohistochemical stains positive for cyclin D1, FISH\*\*\* studies positive for t(11;14), Ki-67 index markedly elevated 95%, p53 showed wild-type staining pattern, sox 11 staining could not be performed due to low amount of sample (**Figure 3 & 4**).

He was established with oncologist, follow-up as outpatient for chemotherapy. PET/CT showed mild to moderate FDG uptake above and below the diaphragm including left cervical and supraclavicular region, bilateral internal mammary chains, bilateral cardiophrenic, retrocrural region, upper abdomen, retroperitoneum, diffuse splenic hypermetabolism. A focus of intense uptake in descending colon and anorectal junction could be related to lymphoma.

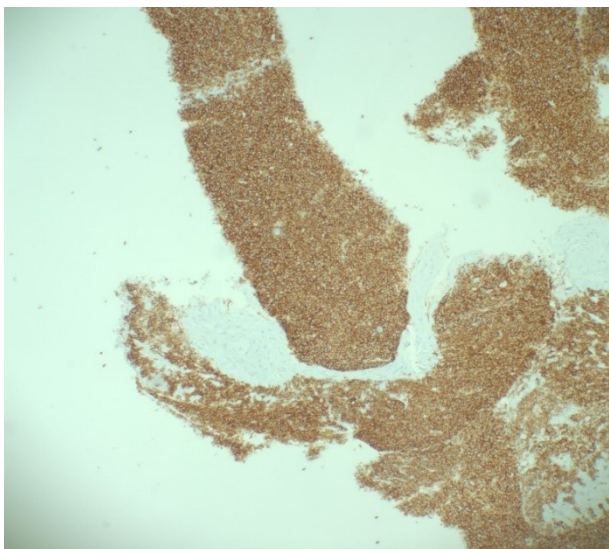
Patient readmitted 5 days later with shortness of breath due to worsening pleural effusion, drained with chest tube and discharged home. Readmitted again after first dose of chemotherapy (R mini- CHOP) with shortness of breath and abdominal pain, Patient developed leukopenia and thrombocytopenia with CT chest showing bilateral ground glass alveolar and reticular interstitial infiltrates most likely secondary to infection due to immunosuppression. Also had splenic infarct, distended gallbladder and elevated lipase, worsening transaminitis. Considering patient's stage IV lymphoma, current possible severe pneumonia, pancreatitis and splenic infarct, and transaminitis, patient and family chose for comfort and hospice care.



**Figure 2:** CT abdomen.



**Figure 3:** CD5.



**Figure 4:** Cyclin D.

### Discussion

Spontaneous hemoperitoneum is an uncommon cause of acute abdominal pain. When it occurs, it could be life threatening. Causes of spontaneous hemoperitoneum can be classified as:

**Hepatic:** Benign and malignant tumors of liver; adenoma, hemangioma, focal nodular hyperplasia, angiosarcoma, hepatocellular carcinoma, pregnancy complicated by eclampsia. May need result in spontaneous hepatic rupture, HELLP syndrome, hepatic adenomatosis, infiltrative disease of liver like amyloidosis.

Metastatic disease liver from colon, lung, renal cell carcinoma, testicular, choriocarcinoma.

**Splenic:** Spontaneous splenic bleeds usually associated with splenic infections like CMV, malaria, EBV. Infiltrative diseases, lymphoma, leukemia, angiosarcoma or cyst.

**Vascular:** Arterial vs venous related to multiple conditions.

#### Coagulation disorders

**Gynecologic:** Pregnancy, cyst rupture etc. [4].

The pathogenesis behind MCL is mutation lead to overexpression of Cyclin D1, which is family of protein expressed in all adult human tissues with the exception of cells derived from bone marrow stem cell lines (both lymphoid and myeloid).

Cyclin D1 is a regulatory subunit of cyclin-dependent kinases CDK4 and CDK6. The protein dimerizes with CDK4/6 to regulate the G1/S phase transition and entry into the S-phase [5]. The chromosomal translocation t(11;14) resulting in aberrant expression of cyclin D1. Secondary genetic events increase the oncogenic potential of cyclin D1 and frequently inactivate DNA damage response pathways. In combination these changes drive cell-cycle progression and give rise to pronounced genetic instability [6].

This is a 65-year-old male who presents with significant leukocytosis with absolute lymphocyte count more than 26,000, massive hemoperitoneum leading to hypotension causing anion gap metabolic acidosis and prerenal AKI, and right-sided

pleural effusion, improved with conservative supportive management. CT abdomen/pelvis, MRI and GI nuclear scan did not localize any bleeding spot.

Our case is unique in its presentation because hemoperitoneum secondary to mantle cell lymphoma has never been reported before. As we go through details of mantle cell lymphoma and GI involvement, no case reports or series mention direct hemoperitoneum caused by mantle cell lymphoma. There are multiple case reports in which MCL presented with upper or lower GI bleed [7], spontaneous ruptured spleen [8], but none have been reported with spontaneous hemoperitoneum.

The mechanism is not clearly known but most likely due to vascular occlusion in hypercoagulable state leading to spontaneous bleeding from leaking capillaries or infiltration of the tumor cell in the vessels wall or ruptures lymph node, which is hard to distinguish.

MCL presents with myriad of symptoms ranging from lymphadenopathy, fever, night sweats, hepatosplenomegaly, GI involvement. It can literally involve GI tract from oral cavity to anorectal junction.

Almost every case has GI involvement as shown by a prospective study on upper and lower GI biopsies [9] is extensive including stomach, duodenum, and colon. Multiple areas can be involved in one person, with extensive colonic involvement presenting as polyposis [10].

As mentioned above in case presentation, PET/CT in our patient showed focal intense lesions at descending colon and anorectal junction, considering the nature of mantle cell lymphoma involvement of GI tract, this could be possible extra nodal site of MCL.

Bone marrow involvement also reported in more than 60% patient, autoimmune phenomena including paraneoplastic syndromes. Usually presented with anemia and thrombocytopenia, with leukocytosis having absolute lymphocyte count more than 4000 reported in 10-20% cases [11]. Our patient had absolute lymphocyte count more than 26,000 at some point.

Median age of presentation is 63, more common males, most commonly presents with generalized lymphadenopathy at stage III or IV. Over 50% of patients have B type symptoms of which weight loss is most common. Massive splenomegaly is a frequent feature with cytopenia, hepatomegaly occurs more, as compared to other non-Hodgkin lymphomas [11]. like it did in our patient. This patient also had cirrhosis with portal hypertension on presentation with negative viral panel. No extensive work-up to rule out rare causes of cirrhosis was done, although hepatitis panel was negative. Patient did not have any previous history of systemic illness, pruritus, jaundice, fatigue, weight loss, joint pains. Cirrhosis was thought to be related to non-Hodgkin lymphoma, there are case reports with non-Hodgkin lymphoma presenting as acute or acute on chronic liver failure. Very likely our patient had cirrhosis secondary to MCL [12,13,14].

### Conclusion

MCL is one of the lymphoma subtypes with poor prognosis that can present with different presentations including very rare case of hemoperitoneum.

---

**References**

1. Navarro A, Beà S, Jares P, Campo E. Molecular Pathogenesis of Mantle Cell Lymphoma. *Hematol Oncol Clin North Am.* 2020; 34: 795-807.
2. Jain P, Wang M. Mantle cell lymphoma: Update on the diagnosis, pathogenesis, prognostication, and management. *Am J Hematol.* 2019; 94: 710-725.
3. Vose JM. Mantle cell lymphoma: Update on diagnosis, risk-stratification, and clinical management. *Am J Hematol.* 2017; 92: 806-813.
4. Lucey BC, Varghese JC, Soto JA. Spontaneous hemoperitoneum: causes and significance. *Curr Probl Diagn Radiol.* 2005; 34: 182-195.
5. Cohen JB, Zain JM, Kahl BS. Current Approaches to Mantle Cell Lymphoma: Diagnosis, Prognosis, and Therapies. *Am Soc Clin Oncol Educ Book.* 2017; 37: 512-525.
6. Qie S, Diehl JA. Cyclin D1, cancer progression, and opportunities in cancer treatment. *J Mol Med (Berl).* 2016; 94: 1313-1326.
7. Ahmed R, Kumar K, Makker J, Niazi M, Balar B. Recurrent Mantle Cell Lymphoma Presenting as Gastrointestinal Bleeding. *Case Rep Gastroenterol.* 2018; 12: 379-384.
8. Gennai A, Basili G, Lorenzetti L, Giuseppe Crocetti, Mario Filidei, et al. Rottura spontanea di milza in linfoma non Hodgkin: caso clinico [Spontaneous rupture of the spleen in non-hodgkin lymphoma: A case report]. *Chir Ital.* 2008; 60: 739-744.
9. Salar A, Juanpere N, Bellosillo B, Eva Domingo-Domenech, Blanca Espinet, et al. Gastrointestinal involvement in mantle cell lymphoma: a prospective clinic, endoscopic, and pathologic study. *Am J Surg Pathol.* 2006; 30: 1274-1280.
10. Martins C, Teixeira C, Gamito É, Oliveira AP. Mantle cell lymphoma presenting as multiple lymphomatous polyposis of the gastrointestinal tract. *Rev Bras Hematol Hemoter.* 2017; 39: 73-76.
11. Shivdasani RA, Hess JL, Skarin AT, Pinkus GS. Intermediate lymphocytic lymphoma: Clinical and pathologic features of a recently characterized subtype of non-Hodgkin's lymphoma. *J Clin Oncol.* 1993; 11: 802-811.
12. Siba Y, Obiokoye K, Ferstenberg R, Robilotti J, Culpepper-Morgan J. Case report of acute-on-chronic liver failure secondary to diffuse large B-cell lymphoma. *World J Gastroenterol.* 2014; 20: 16774-16778.
13. Saló J, Nomdedeu B, Bruguera M, J Ordi, P Ginès, et al. Acute liver failure due to non-Hodgkin's lymphoma. *Am J Gastroenterol.* 1993; 88: 774-776.
14. Lettieri CJ, Berg BW. Clinical features of non-Hodgkins lymphoma presenting with acute liver failure: a report of five cases and review of published experience. *Am J Gastroenterol.* 2003; 98: 1641-1646.