International Journal of Research in Medical Sciences
Bhargav M et al. Int J Res Med Sci. 2024 May;12(5):1738-1741
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20241268

Original Research Article

Spontaneous laceration of spleen with hilar lymph node metastasis: a case of primary splenic mantle cell lymphoma in elderly

Mithlesh Bhargav[1]*, Bandana Mehrotra[2], Ashok Kapoor[3], Vaibhav Raj Gopal[4]

[1]Department of Pathology and Lab Medicine, AIIMS Gorakhpur, Uttar Pradesh, India
[2]Lab director, RML Labs Lucknow, Uttar Pradesh, India
[3]Department of Histopathology, RML Labs Lucknow, Uttar Pradesh, India
[4]Department of General Surgery, RMLIMS, Lucknow, Uttar Pradesh, India

Received: 12 March 2024
Revised: 04 April 2024
Accepted: 06 April 2024

***Correspondence:**
Dr. Mithlesh Bhargav,
E-mail: me.bhargav1119@gmail.com

ABSTRACT

Mantle cell lymphoma (MCL) is a type of non-Hodgkin (B-cell) lymphoma (NHL) with manifestations ranging from indolent to aggressive disease. It arises from mantle zone or primary follicle lymphocytes and is associated with translocation t (11;14) which is seen in almost all cases. Most of the cases present at stage III/IV with hepatospenomegaly, generalized lymphadenopathy, bone marrow involvement or lymphoid polyposis. Rate of relapse is high occurring in 50-60% patients and 5-year survival rates are low ~27-30%. Median overall survival is 3.5 years. Age >60 years, raised serum LDH, high mitotic count, Ki67>30%, blastoid or pleomorphic variants, TP53 mutation, gains in 3q,11q and deletions of 13q as well as 17p are important prognostic factors associated with worst outcome. Treatment involves conventional chemo-immunotherapy and stem cell transplantation (SCT). In our case the elderly patient had an atraumatic splenic rupture with no past medical history of trauma. The patient presented to the emergency department with severe abdominal pain in left upper quadrant. Thus emergency splenectomy was executed successfully, and the patient was stabilized. After receiving initial cycle of R-CHOP regime, he was lost to follow up. In this case report, we will discuss the clinical presentation, as well as current treatment guidelines for atraumatic splenic rupture.

Keywords: Malignant neoplasm, Lymphoma, Spleen, Hilar lymph nodes

INTRODUCTION

Splenomegaly causes a diagnostic dilemma in distinguishing pathologic conditions primarily involving the spleen or from those which arise as a result of hepatic or multisystemic diseases. Among the causes of isolated splenomegaly, lymphoid malignancies account for a relevant but often underdiagnosed cases. Splenic lymphomas constitute a wide and heterogeneous spectrum of diseases, whose clinical behavior spans from indolent to highly aggressive ones.[1] Most common lymphoma of spleen is diffuse large B cell lymphoma however among small sized lymphoproliferative disorders splenic marginal zone lymphoma accounts for 20% of marginal zone lymphomas and <2% of lymphoid neoplasms. Mantle cell lymphoma is a type of non-Hodgkin (B-cell type) lymphoma that can involve the lymph nodes, spleen, blood, and bone marrow.[2] This type of NHL is known to be highly aggressive with a short remission period to standard therapies and an increased risk of relapse.

CASE REPORT

A sixty-two-year-old male had a past history of vague abdominal pain and swelling in epigastic and left lumbar region for last 2 years. General and systemic examination was within normal limits, so symptomatic treatment was given. Presently patient came with hemoperitoneum due to

spontaneous atraumatic rupture of spleen, for which emergency splenectomy was done. Intraoperatively spleen weighed 1.56 kg and measured 22x17x8 cm. Subsequently, it was sent for histopathological examination.

Outer surface of spleen was smooth with lacerated area covered with blood clots, which was seen 7 cm away from the hilum. It was sliced in vitro; each slice measuring 1 cm in thickness. Cut surface showed two greyish-white areas of infarction, each measuring 3.5x2.5x1 cms and 3.5x2.5x2 cms respectively. Rest of the cut surface showed congestion. Hilum showed six small lymph nodes, largest measuring 2.0x0.4x0.4 cm. Representative sections were taken from congested areas as well as hilar lymph nodes. Microscopically, lymph nodes as well splenic parenchyma showed effacement of architecture by diffuse infiltrate of small sized atypical round to ovoid lymphoid cells (Figure 1 (A-E)). Atypical lymphocytes had coarse to clumped chromatin and single central prominent nucleoli (Figure 1 (C, F)). Few mitoses were also seen. Sections from infarcted areas showed coagulative necrosis and haemorrhage (Figure 1H). Patchy areas near hilum of spleen showed similar morphology as in lymph nodes along with congestion, haemorrhage and lymphoid hyperplasia (Figure 1 (D-I)). Thus a diagnosis of lympho-proliferative neoplasm was rendered and further typing based on immunohistochemistry was advised.

On immunohistochemical (IHC) examination the tumour cells were positive for Pax5, LCA and CD 20. Adjoining reactive areas in node were displaying patchy CD3 positivity (Figure 2 (A-C)). A second panel for small sized B cell type lymphoproliferative disorders was applied which yielded positive reaction for CD5, Cyclin D1, Bcl2, lambda and kappa (Figure 2 (D-F). On the other hand tumour cells were negative for CD 23 and CD 10 ruling out small lymphocytic lymphoma, follicular lymphoma, marginal zone lymphoma and lymphoplasmacytic lymphoma (Figure 2 (G, H)). The ki 67 index was 40-45% (Figure 2I). Later SOX11 was also added which was negative in our case. The patient was finally diagnosed as a case of splenic mantle cell lymphoma (SMCL).

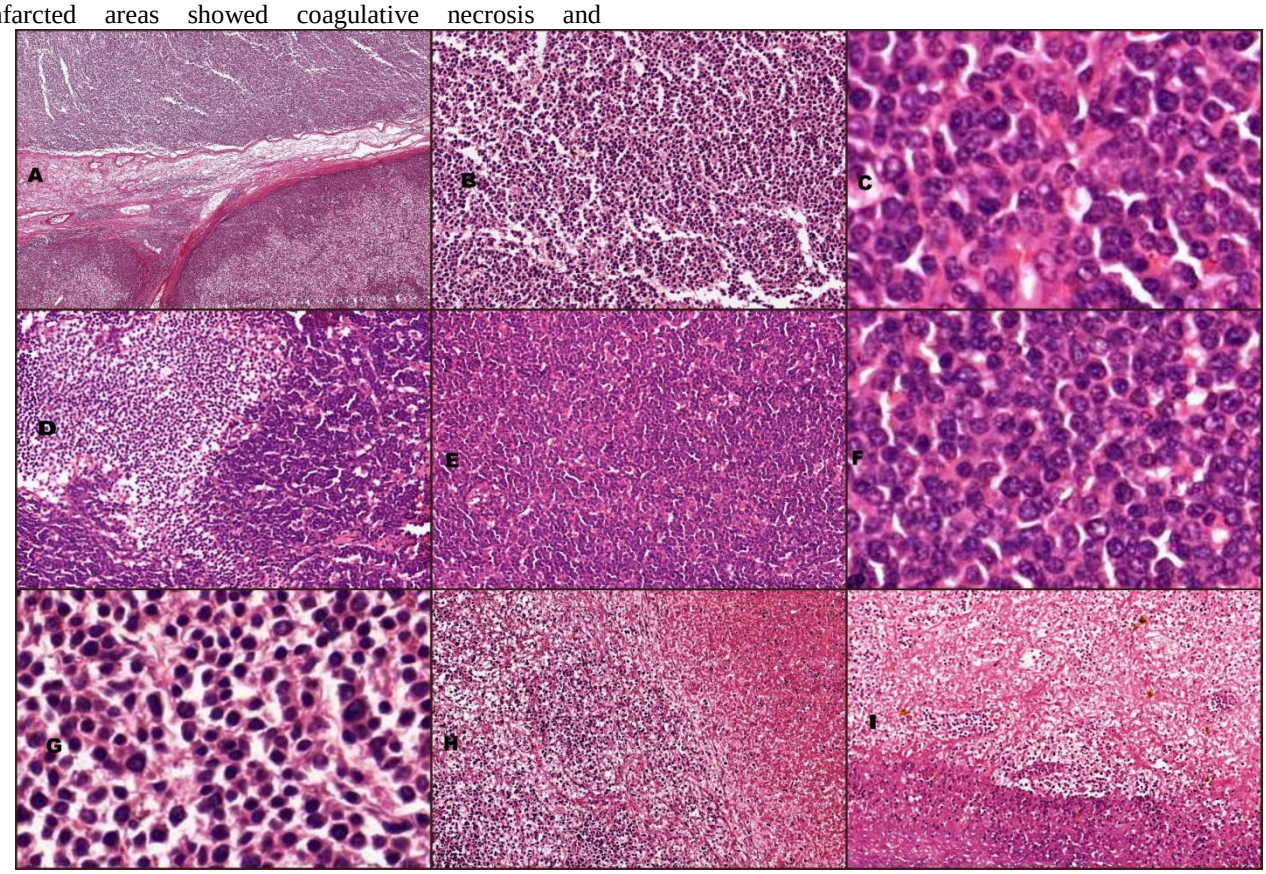

Figure 1: (A) The section from hilum of spleen shows part of unremarkable splenic parenchyma along with lymph node effaced with monotonous population of small atypical lymphoid cells (H and E, 4X). (B) The section from hilar lymph node shows small sheets of monomorphic cells (H and E, 10X). (C) The section from splenic hilum shows round to oval cells with coarse chromatin, prominent nucleoli and scant amounts of eosinophilic cytoplasm (H and E, 40X). (D) The section displays part of lymph node with a germinal centre (H and E, 10X). (E) The section from splenic parenchyma shows similar histomorphology as described in the lymph node (H and E, 10X). (F) The section from splenic parenchyma shows similar histomorphology as described in the lymph node (H and E, 40X). (G) The section from node shows reactive lymphoid hyperplasia displaying mature lymphoid cells (H and E, 40X). (H) Areas of hemorrhagic necrosis adjacent to tumour. (H and E, 10X). (I) Areas of necrosis and necrotic debris within tumour. (H and E, 10X).

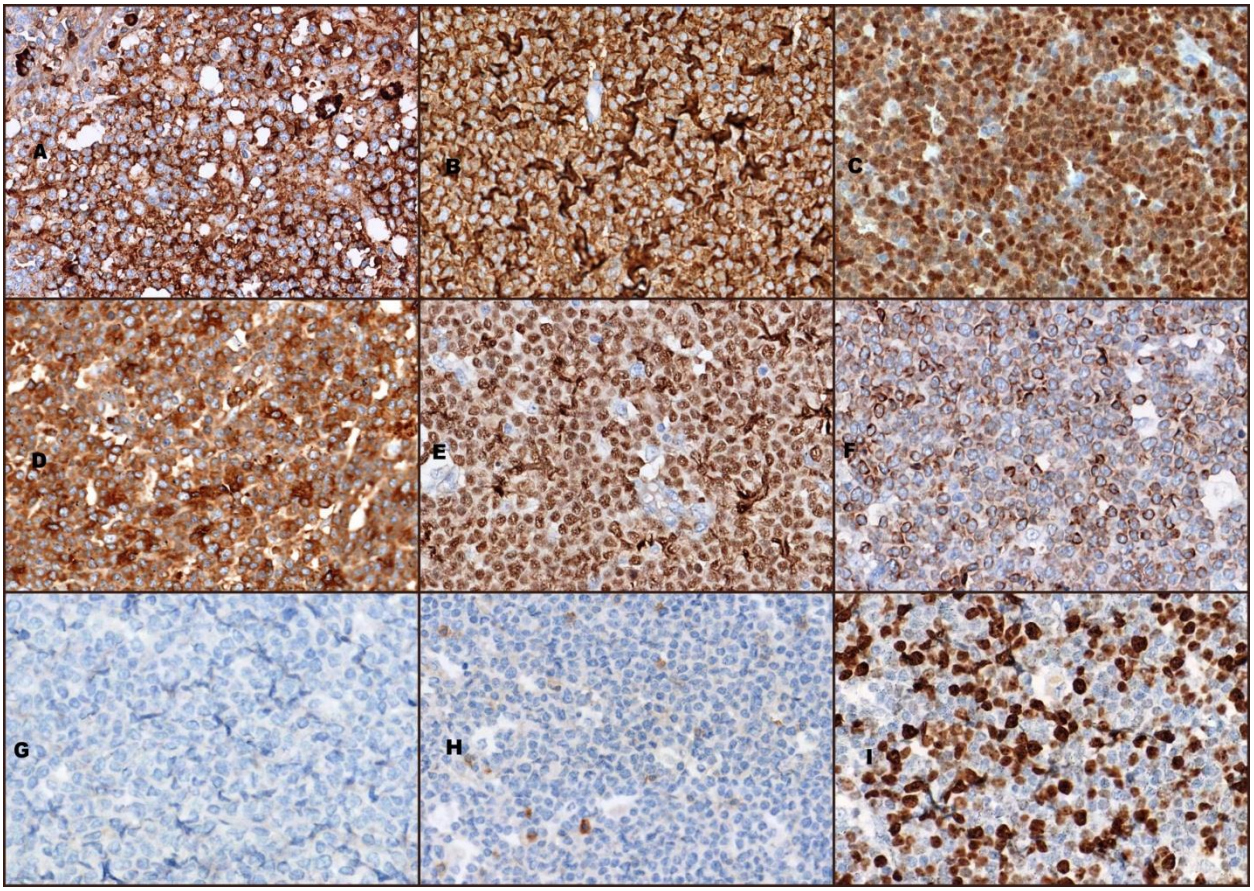

Figure 2: (A) Section shows Pax 5 positive tumour cells (IHC, 20X). (B) Section shows LCA positive tumour cells (IHC, 20X). (C) Section shows CD 20 positive tumour cells (IHC, 20X). (D) Section shows BCL-2 positive tumour cells (IHC, 20X). (E) Section shows cyclin D1 positive tumour cells (IHC, 20X). (F) Section shows CD-5 positive tumour cells (IHC, 20X). (G) Section shows CD-10 negative tumour cells (IHC, 20X). (H) Section shows CD-23 negative tumour cells (IHC, 20X). (I) Section shows ki67 ~40% in tumour cells (IHC, 20X).

DISCUSSION

Mantle cell lymphoma comprises 3-8% of adult onset NHL. This type of NHL is predominantly found in western countries and affects men more often than women (M:F 2:1). The median age of diagnosis with the disease is around 60 years of age. MCL is characterized by a translocation of the Bcl-1 gene t (11; 14) (q13; q32), which is responsible for upregulating cyclin D1 expression. It is a type of non-Hodgkin (B-cell type) lymphoma that can involve the lymph nodes, spleen, blood, and bone marrow.[3-6] In most of cases, lymph nodes are the primary site of neoplasm. Extra-nodal sites predominantly include bone marrow (69%-79%) and spleen (47%) followed by gastrointestinal tract, liver, head and neck. This type of NHL is known to be highly aggressive with a short remission period and an increased risk of relapse. The two major subgroups of MCL are divided into Nodal and Leukemic non-nodal MCL. Nodal MCL is the most common and aggressive type with overexpression of SOX11. It has high degree of genomic instability while leukemic non-nodal MCL commonly presents with leukocytosis and splenomegaly. In this subtype there is no expression of SOX11 and patients have higher genome stability. This case also showed negative reaction for SOX11 however cyclin D1 was strongly positive.

Patients may present with lymphocytosis along with anemia and thrombocytopenias. Quarter of the patients present with fever, malaise, weight loss, and night sweats. The patient can also have hepatosplenomegaly or lymphadenopathy. In cases of asymptomatic mild to moderate splenomegaly surgical removal or transcatheter ablation of splenic parenchyma is done.[7] In the case of splenomegaly with spontaneous rupture, clinical signs of shock and confusion are evaluated and immediate hemodynamic stability is achieved.

While spontaneous splenic rupture in patients with MCL is rare, this unusual complication should be considered in patients presenting with vague abdominal pain. In MCL, an enlarged spleen accounts for nearly 40% of cases.[8,9] In aggressive forms of MCL, highly proliferative neoplasm can increase tensile forces from within the spleen. It has been theorized, that the rate of splenic expansion outpaces the rate of splenic capsule compensation, which creates high sheering tensile forces of the splenic tissue leading to rupture. Another potential possibility leading to splenic

rupture is compression of abdominal musculature on splenic tissue during physical activities. The risk of splenic rupture increases drastically with age. This is mostly due to the changes in the behaviors of hematologic cell types and anatomical abnormalities leading to splenic vulnerability.[9]

Immunophenotypically, MCL is positive for CD5, CD19, CD20, CD22, PAX5, CD79a and Cyclin D1. MCL also expresses cell surface immunoglobulins IgM and IgD. The overexpression of SOX11 has been recognized as a specific marker for diagnosis of MCL. Although there is no standard care or treatment for MCL, the management of patients with MCL is based on the patient's age, immune status and fitness. Therapies include incorporating rituximab and cytarabine, alongwith autologous hematopoietic stem cell transplantation (ASCT) if eligible. Young fit patients are managed with ASCT with combination therapy of rituximab, while older patients are managed with rituximab only. Complications of MCL include anemia, thrombocytopenia, and neutropenia. In rare cases, patient can present with splenomegaly with an increase of risk spontaneous splenic rupture. Above patient was hemodynamically stablished and was put on aggressive course of chemotherapeutic regime, but was lost to follow up after initial cycle of chemotherapy.

CONCLUSION

Understanding the structural and physiologic consequences of an enlarging spleen within the abdominal cavity is important so that timely screening and accordingy treatment decisions can be undertaken. Using cost-effective methods like ultrasonography patients with MCL can be screened for splenomegaly. In patients with MCL, the early identification of splenomegaly is vital for the patient's safety and survival. While this type of NHL has a low survival rate and an increased risk of relapse, the usage of simple diagnostic tools can help prevent further complications associated with atraumatic splenic rupture.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Bhargav M, Mehrotra B, Kapoor A, Gopal VR. Spontaneous laceration of spleen with hilar lymph node metastasis: a case of primary splenic mantle cell lymphoma in elderly. Int J Res Med Sci 2024;12:1738-41.