Rare Incidental Occurrence of Mantle Cell Lymphoma with Alcoholic Liver Disease and Pancreatitis

Rateesh Sareen¹, Menka Kapil¹, G N Gupta¹, Dinesh Agrawal²

ABSTRACT

We present an extremely rare case of Mantle cell Lymphoma with concomitant alcoholic liver disease and pancreatitis in a 48 year old male patient. There was absence of abdominal lymphadenopathy. The complete blood count showed absolute lymphocytosis which on subsequent peripheral blood examination showed small to medium sized atypical cells raising suspicion of prolymphocytic leukemia. The flow cytometry and Immunohistochemistry examination confirmed Mantle cell lymphoma. The morphology of cells in mantle cell lymphoma are known to mimic prolymphocytes on peripheral blood. However, the rare concomitant occurrence of both diseases makes the case an interesting one for readers and emphasizes on the usefulness of peripheral blood examination although not completely diagnostic but raising an alarm for the diagnosis of this rare lymphoma.

Key words: Mantle cell lymphoma, Alcoholic liver disease, Prolymphocytes.

CASE REPORT

A 48 year old male was admitted with complaints of abdominal pain, weakness and loss of appetite since a month. The patient was a known case of alcoholic liver disease and chronic pancreatitis. On physical examination there was icterus, pallor and bilateral pedal edema. Personal history was insignificant. There were few small 1 to 2 cm sized posterior cervical lymph nodes also palpated. The biochemical investigation showed Sodium-136 mmol/L, Chloride- 109 mmol/L, Total bilirubin- 5.0 mg/dl, direct bilirubin- 4.4 mg/dl, Total protein- 6.2 gm/dl, Albumin- 3.1 g/dl, A:G ratio- 1.0, Gamma GT- 557 U/L, Alkaline phosphate-590 U/L, SGOT-1907 U/L, SGPT- 809 U/L, Amylase- 371 U/L, Uric acid- 6.6 mg/Dl, LDH- 3921 U/L, Lipase- 24 U/L and non reactive for Hepatitis B, A, E &HIV-1 & 2. Renal function tests were normal. X –ray chest and CT chest showed diffuse ground glass haziness in bilateral lung with interlobular septal thickening likely to be diffuse alveolar hemorrhage. Mild bilateral pleural effusion is also noted. Left paratracheal, prevascular and paratracheal lymph nodes were enlarged (Figure 1 & 2). The Complete blood count showed hemoglobin 7.4 g/dL, Red blood cell count- 2.44 x 10¹² / mm³, White blood cell count 99.38 x 10⁹/mm³, platelet count 23 x 10⁹/mm³. WBC differential showed 79% lymphocytes, 5% neutrophils, 1% monocytes and 15% atypical cells. The atypical cells were small to medium sized cells having high nucleo cytoplasmic ratio, condensed chromatin with occasional nucleoli and scanty cytoplasm. These cells could not be classified as prolymphocytes or blasts (Figure 3). Bone marrow aspiration was hypercellular with suppression of erythroid series and presence of medium to large sized atypical cells with prominent nucleoli? prolymphocytes. Megakaryocytes were reduced in number with normal morphology. A provisional diagnosis of lymphoproliferative disorder was made (Figure 4). Bone marrow biopsy was hypercellular with complete architectural effacement and infiltration by medium to large lymphocytes with chromatin condensation and mild pleomorphism was noted (Figure 5). Flow cytometry examination from peripheral blood revealed a monoclonal B-lymphoid lineage expressing CD 19 with co expression of CD 5. Bright CD 20, moderate CD 38 and bright CD 45 expression and a marker profile comprising of monomorphic small to medium sized cells. A provisional diagnosis of B cell lymphoproliferative disorder was made and cyclin D1 immunohistochemistry was advised for further confirmation. Cyclin D 1 on bone marrow biopsy showed positive nuclear staining and hence a diagnosis of Mantle cell lymphoma was confirmed (Figure 6). Mantle cell lymphoma (MCL) is a B cell neoplasm comprising of monomorphic small to medium sized lymphocytes arising from the inner mantle zone[1,2]. It constitutes 3-10% of non Hodgkin’s lymphoma and occurs in middle age to older individuals with male preponderance. It is the most common sites of involvement are spleen, bone marrow and occasionally peripheral blood. The extra nodal involvement involves Waldeyer ring and gastrointestinal tract.¹⁷
Peripheral involvement is occasional on routine morphological examination whereas it is common on flow cytometry. Patients present with marked lymphocytosis with presence of cells with irregular nuclear contours mimicking prolymphocytes. Lymph nodes are characterized by a nodular, diffuse mantle zone growth pattern. Small to medium sized centrocytes with dispersed chromatin and inconspicuous
come the suppressive effect of RB1 and P27 Kip1 leading to development of MCL. Those cases that are cyclin D1 and t(11, 14) negative but have all features of conventional MCL have high expression of cyclin D or D 3. In such case diagnosis should be made with caution. The median survival of patients is 3-5 years. High mitotic rate > 10 hpf and Ki 67 positive cells > 40% to 60% are adverse prognostic indicator.

There are cases in literature where pancreatic tumor has presented with pancreatitis or ascites. Collison tumors like pancreatic adenocarcinoma with MCL have also been reported or a pancreatic tumor with ascites as manifestation. But the incidental finding of MCL with pancreatitis in absence of abdominal lymphadenopathy makes the case an interesting one for readers.

REFERENCES

14. Sequence analysis proves clonal identity in five patients with typical and blastoid mantle lymphoma C Cameron Yr1, L Jeffrey Medeiros1, Candy C Cromwell1, Ashwini P Mehta1, Pei Lin1, Rajalakshmi Luthra1 and Lynne V Abuzzoo1 Modern Pathology (2007) 20 , 1–7 doi: 10.1038/modpathol.3800716; published online 20 October 2006.
20. Salaverria I1, Zettl A, Béa S, Moreno V, Valls J, Hartmann E, et al. Specific

Figure 5: Bone Marrow biopsy (Hematoxylin & Eosin, 40 x).

Figure 6: Bone Marrow Immunohistochimistry Cyclin D1 , 40x).

nucleoli are seen. There is paucity of centroblasts, immunoblasts and proliferation centers. Occasionally scattered single histiocytes creating a starry sky appearance are also noted. As such the lymphoma do not progress to large cell lymphoma but occasionally loss of mantle zone, dispersed chromat, marked pleomorphism and increased mitotic activity are seen in case of relapse. Morphological variants of MCL are blastoid (cells resembling lymphoblast with dispersed chromat and high mitotic rate), pleomorphic (large cells with oval to irregular nuclear contours, pale cytoplasm & prominent nucleoli), small cell variant (mimicking small lymphocytic lymphoma) and marginal zone like (cells resembling monocytoid B cell of Mantle zone). Blastoid and pleomorphic variants have aggressive course. Immunochemistry[12-16] Cells are positive for CD 5, FMC-7 and CD43 and negative for CD10 & bcl-6. CD23 is negative or weakly positive. Immunochemistry is positive for bcl-2 and cyclin D1. There is presence of t(11,14) between IGH and cyclin D1 in all cases of MCL. T he translocation results in over expression of CCND1 m RNA and protein. The deregulated expression of cyclin D1 is assumed to over-


Cite this article: Sareen R, Kapil M, Gupta GN, Agrawal D. Rare Incidental Occurrence of Mantle Cell Lymphoma with Alcoholic Liver Disease and Pancreatitis. OGH Reports. 2018;7(1):49-52.