INTRODUCTION

Primary extranodal non-Hodgkin's lymphomas (NHL) arise from tissue other than lymph nodes and even from sites which normally contain no lymphoid tissue. At least one-fourth of the lymphomas are probably of extranodal origin. Gastrointestinal tract is the most common site of extra nodal non-Hodgkin's lymphomas (NHLs), accounting for 15-20% of all NHL cases. Stomach is the most common site of extranodal gastrointestinal lymphoma. Although secondary involvement of the pancreas is seen often in cases of gastrointestinal lymphoma but primary pancreatic lymphoma (PPL) is an extremely rare disease that can mimic pancreatic carcinoma. Fewer than 2% of extra-nodal malignant lymphomas and 0.5% of all pancreatic masses constitute PPL. The incidence is expected to be higher in patients of human immunodeficiency virus (HIV) but occurrence of PPL in acquired immune deficiency syndrome (AIDS) is limited to case reports.

Primary liver lymphoma is also rare, corresponding to less than 1% of all lymphomas, 0.4% of extranodal NHL and approximately 0.01% to 0.06% of the non-Hodgkin lymphomas.

Case Report

A 46 year old male patient presented with complains of generalized weakness and right upper abdominal pain for 1 month of duration. The patient was grossly emaciated with jaundice and right hypochondrial tenderness. No other abnormalities were detected by clinical examination.

Routine laboratory investigation showing mild anaemia, raised ESR, raised bilirubin, liver enzymes (ALT/AST) and ALP and markedly raised LDH (strongly pointing towards malignancy). The patient was seropositive for HIV 1 antibodies with CD4 count of 120 and was started on antiretroviral therapy with tenofovir, lamivudine and efavirenz. Patient was tested negative for hepatitis –B surface antigen (HBsAg), hepatitis- C antibody (HCVAb) and Epstein-Barr Virus (EBV) antibody. Alpha fetoprotein, CEA, CA 19-9 levels were within normal range. CECT abdomen showed hepatomegaly with multiple hypodense lesions with largest one of 2.3cm in diameter and pancreatic head shows ill defined hypodense lesion of 1.7x1.8cm without any lymph node enlargement. MRI abdomen revealed multifocal nodular lesion in liver with bulky head of pancreas with cystic lesion which suspect HCC.

However on FNAC from SOL of liver and pancreas showed immature lymphoid cells in the background of few mature lymphocytes and lymphoglandular bodies, suggestive of hematoid lymphoid malignancy. A CT guided biopsy showed high grade lymphoid cells on histopathological examination (HPE). On immunohistochemical (IHC) staining cells were positive for CD20 (B cell marker), CD45 (leucocyte common antigen), negative for CD3 (T cell marker). Hence the diagnosis of primary NHL involving liver and pancreas was made.

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DISCUSSION

Malignancy of lymphoid cells arise from cells of immune system at different stage of differentiation. Some of them present as leukemia and others as lymphomas. Non-Hodkin lymphomas (NHL) were separated from Hogdkin lymphoma by recognition of Sternberg-Reed cells early in th 20th century.

NHLs are more frequent in the elderly with male predominance. Lymphomas are occur in increased frequency in patients with AIDS, usually with CD4+ T cell< 200/micro l. At least 4% of all AIDS patients develop lymphoma during the course of illness.6

NHL is a rare cause of pancreatic masses. Freeman et al. reported that only 0.6% of extranodal malignant lymphoma coming from the pancreas.7 Primary hepatic lymphoma commonly presents at fifth decade with male predominance. The most common presenting symptom is abdominal pain with hepatomegaly and jaundice is occasional physical findings. There is high prevalence of HCV or HBV infection in PLL patients. 40% - 60% of PHL seen in HCV infected patients.8 Our patient, 46years old male presented with right upper abdominal pain with generalized weakness. Patient was having jaundice and right hypochondrial tendernes. He was found to be HIV seropositive with CD4+ T cell count 120/micro l, but he was negative for both HbsAg and HCV Ab.

Though tumor can involve any part of pancreas, head of the pancreas is the most common site of PPL (>80%).9 Primary liver lymphoma may present as single or multiple masses or even as a diffuse infiltration.

In our case there is multiple hypodense lesions with largest one of 2.3cm in liver and an ill defined hypodense lesion of 1.7x1.8 cm in the head of pancreas.

Lactate dehydrogenase level can be elevated in in NHL as well as adenocarcinoma. The serum CA19-9 level in patients with PPL is usually normal.10 In our case, the elevated LDH level (425 IU/L), and normal CA19-9 levels ruled out pancreatic adenocarcinoma. CT-guided FNAC from SOL of liver and pancreas also showed immature lymphoid cells and lymphoglandular bodies which suggest hematolymphoid malignancy and was confirmed by immunohistochemical staining for CD20 (B cell marker), CD45 (leucocyte common antigen).

CONCLUSION

PPL is a rare neoplasm that mimics pancreatic adenocarcinoma but treatment and prognosis is totally different. PHL is also again extremely rare disease, usually related to HBV or HCV infection but can occur in immunocompromised patient (AIDS). Patient may present with non-specific clinical feature. But with strong suspicion especially in immunocompromised patient if invasive investigation done we can reach to proper diagnosis and also increase the survival of patient as the disease is treatable and with new drugs such as rituximab.

References

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