Research Paper Medical Science

PLASMABLASTIC LYMPHOMA AS ASCITES- ENDOSCOPE FURTHER

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ABSTRACT

Gastrointestinal Plasmablasticlymphoma(GI - PBL) is common in immunocompromised patients and has grave prognosis. We are reporting two cases of GI-PBL with low Serum Ascites Albumin gradient(SAAG) ascites in immunocompetent individuals. The diagnosis in the first case is made on endoscopic biopsy from deep duodenal nodule and in second case by colonoscopic biopsy of nodular growth in transverse colon followed by immunohistochemistry (IHC). In the presence of pleocytic, low protein low SAAG ascites emphasizes the need for deeper endoscopic intubation into jejunum or terminal ileum when routine endoscopy or colonoscopy is normal for clinching the diagnosis.

KEYWORDS Plasmablastic Lymphoma, Endoscopy, Colonoscopy, Ascites

INTRODUCTION:

Lymphoma of Gastrointestinal tract(GIT) constitutes 15-20% of all small intestine neoplasms. GIT-Non Hodgkins Lymphoma(NHL) is most common in Ileum (60%-65%) followed by jejunum(20%-25%), duodenum (6%-8%) and other sites.[1] Plasmablastic lymphoma of gastrointestinal tract (GI-PBL) is a rare variant of B-cell NHL with an aggressive clinical course and shorter overall survival of 1-2 years. GI-PBL is common with human immunodeficiency virus (HIV) infection and immunocompromisedstatus.[2] GI- PBL accounted for no more than 250 published cases in over 10 years[3]. In 20-30% of patients with poor outcome were noted to have serous effusions (Pleural, Pericardial and peritoneal)[4]. Unique GI-PBL CD 45 negative immunohistochemical (IHC) profile differentiates from CD45 positive NHL.Clinicians should diagnose GI-PBL, treat early and potentially salvage these patients[2].

CASE REPORTS:

Case 1: A 66 year old male presented with progressive, painless distension of abdomen for 3 months duration followed by swelling of both lower limbs. He had anorexia, weight loss of 6 kilograms and low grade fever. He had no jaundice, bleeding diathesis or altered sensorium. He was non alcoholic with no comorbidities. Bilateral pitting pedal edema and ascites without organomegaly were noted.Other systemic examination was unremarkable. Routine blood investigations revealed low total protein (2.3gm/dl) and albumin(1.4 gm/dl)without proteinuria and rest of routine investigations being normal. Hepatitis B& C markers and HIV wasnegative. Peritoneal fluid aspirate revealed pleocytosis with predominant mesothelial cells, high LDH, low protein, low SAAG (0.8) and ADA being negative. He underwent endoscopy, which was normal till second part of duodenum and on deeper intubation into D3, there was a nodule of 1cmx0.5cm with normal overlying mucosa(Fig 1,2). Biopsies were taken and sent for pathological examination. Colonoscopy with terminal ileoscopy was normal. Contrast enhanced computed tomography revealed multiple retroperitoneal lymph nodes, jejunal wall thickening and omental deposits. The plan was made for endoscopic ultrasound guided fine needle aspiration but withheld as cell block study revealed multiple lymphoid cells with large nuclei and prominent nucleoli suggestive of lymphoid neoplasm. D3 nodule histopathology revealed mucosal round cell neoplasm and IHC showed positivity for LCA,CD79a,MUM1 with high Ki67 index and kappa overexpression favoring high grade GI-PBL.(Fig3,4,5,6)

CASE 2:

A 33 year old male presented with distension of abdomen since 3 months with constitutional symptoms of low grade fever and weight loss. He was evaluated elsewhere and was started on antitubercular drugs since two months based on peritoneal aspirate analysis which showed pleocytosis with lymphocytic predominance and low SAAG. CECT abdomen showed transverse colon growth(Fig7). There were no other mass lesions or lymphadenopathy. Colonoscopy revealed large polypoidal growth in the transverse colon. Scope couldn't pass beyond obstruction .Multiple biopsies were taken from growth. Histopathology showed evidence of poorly differentiated tumour and IHC showed LCA, CD138 and MUM1 positivity, CD20 negativity with high Ki67 index and kappa overexpression favoring high grade GI-PBL.

DISCUSSION:

GI-PBL has predilection to oral mucosa and is predominantly seen in HIV positive patient. After index case in 1997, owing to development of immunohistochemistry, GI-PBL is noted in GI tract, lymph nodes, and skin even in immunocompetent individuals. The actual incidence of PBL not associated with HIV infection has not yet been determined. In the largest literature review of 228 patients with PBL, only 39% were HIV negative [5].

The publications from 1998 to 2013 had only 14 cases of GI-PBL. The median age of the reported cases is 57 years. The most common symptoms at presentation were abdominal pain (57%), weight loss (50%), anorexia (36%), and melena (36%). The B symptoms of fever and night sweats were present in 29% and 7% respectively. Other symptoms included abdominal distention, diarrhea, vomiting, and rectal bleeding.[2] Our cases were immunocompetent adults with first patient being 66 years old and second was relatively young with age of 33 years and had symptoms of weight loss, anorexia and low grade fever with ascites as the presenting complaint which is a rare presentation of GI- PBL.

Among the total lymphoma patients with serous effusions recorded by Johnston et al, 72% were males and 29% were females, suggesting male predominance. Serous effusions are more common in T cell associated lymphoma compared to B cell lymphomas.[4]Friis et.al., reported a case of EBV-positive lymphoma in an AIDS patient presenting with pleural effusion and ascites and was diagnosed as a plasmablastic lymphoma (PBL).[6]

All the published reports with serous effusions have predominance of pleural effusion or rarely added with peritoneal effusion. In our cases, it was isolated presentation of Peritoneal effusion which has not been described previously in GI-PBL except for a case report by A Bahari et al, where a 17 year old girl presented with diarrhoea and ascites. On investigating further, she was found to have duodenal and ileal polyps[7]. This in addition to our first case substantiates the need for deeper endoscopic intubation into duodenum/jejunum (or into ileum during colonoscopy) which will aid us in diagnosis especially when peritoneal aspirate is low SAAG and pleocytic.

Plasmablastic lymphoma (PBL) is classified by the World Health Organization as a type of mature B-cell lymphoma that expresses plasma cell antigens (CD38, CD138, MUM1) but not common B-cell antigens (CD20, CD19, PAX5). In our cases there was strong positivity for MUM1, negativity for CD20 favoring GI-PBL and differentiating it from morphologically similar poorly differentiated carcinoma, DLBCL, Burkitt lymphoma, plasmacytoma and EBV associated DLBCL of the elderly[2]

Because PBL does not express the more common lymphoid and/or B-cell markers, it is easy to mistake them for a poorly differentiated carcinoma or sarcoma. Thus, diagnosis of PBL is challenging, particularly when it arises in extraoral locations and in immunocompetent patients[8]

PBL is considered an aggressive lymphoma with a median overall survival of 14 months[3]. The aggressiveness of the condition is emphasized by our first case as he succumbed to death even before chemotherapy could be started.

CONCLUSIONS:

Immunohistochemistry helps in the diagnosis of GI-PBL. In low SAAG pleocytic ascites, deeper intubation during endoscopy or colonoscopy might help in diagnosis and avoiding diagnostic laparoscopy or endoscopic ultrasound examinations.

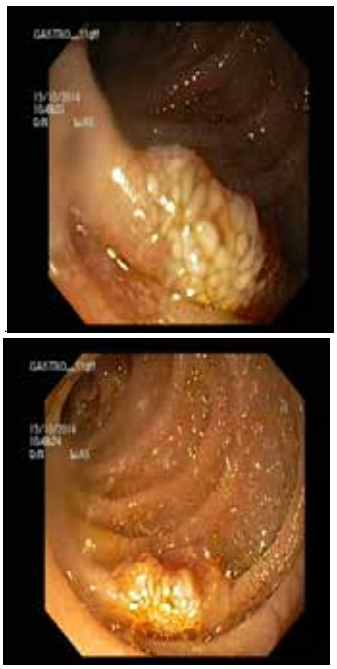

Fig 1:Nodule with normal overlying mucosa ,measuring 1x0.5cm in 3rd part of Duodenum

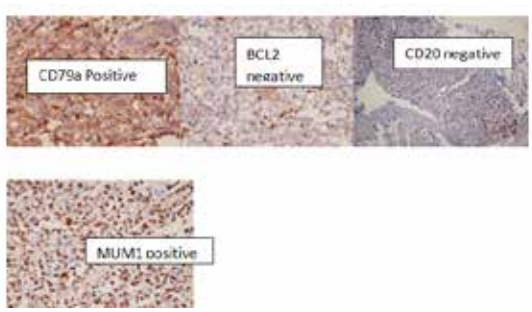

Fig 2: Immunohistochemistry : The immunoprofilefavours high grade plasmablastic Non Hodgkins Lymphoma

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