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Cell & Cellular Life Sciences Journal
ISSN: 2578-4811

Protracted and Tedious-Indolent T Lymphoproliferative Disorder of Gastrointestinal Tract

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Editorial

Volume 8 Issue 1

Received Date: March 01, 2023

Published Date: March 10, 2023

DOI: 10.23880/cclsj-16000177

Editorial

Indolent T lymphoproliferative disorder of gastrointestinal tract is an exceptional disorder associated with infiltration of monoclonal T cells within mucosa of gastrointestinal tract. Generally, a non-destructive infiltrate constituted of miniature, monotonous lymphoid cells demonstrating an immuno-phenotype of CD4+ / CD8- or CD4- / CD8+ appears to infiltrate gastrointestinal mucosa. The exceptionally discerned, indolent T lymphoproliferative disorder of gastrointestinal tract commonly arises within middle aged male subjects, frequently manifests with diarrhoea and exemplifies an indolent clinical course. Although small intestine and colon are commonly incriminated, entire gastrointestinal tract may depict the lesions and no segment of the gut is spared. Indolent T lymphoproliferative disorder represents with a chronic clinical course associated with frequent episodes of disease relapse, inadequate response to cogent chemotherapy and extended survival.

The infrequent, indolent T lymphoproliferative disorder of gastrointestinal tract is predominantly discerned within fifth decade to sixth decade with demonstrable age of disease emergence within 23 years to 79 years. A male predominance is observed with male to female proportion at 2.4:1. The condition is devoid of specific contributory factors [1,2].

Of obscure aetiology, indolent T lymphoproliferative disorder of gastrointestinal tract preponderantly incriminates small intestine and colon. Nevertheless, no site of gastrointestinal tract is exempt from disease emergence. Exceptionally, lesions are accompanied by regional or abdominal lymph node enlargement or incrimination of bone marrow.

Commonly, indolent T lymphoproliferative disorder of gastrointestinal tract represents with clinical symptoms as abdominal pain and diarrhoea. Additionally, symptoms such as vomiting, weight loss or associated features of malabsorption may be discerned [1,2]. Clinically, indolent T lymphoproliferative disorder of gastrointestinal tract can simulate diverse inflammatory bowel diseases as ulcerative colitis or Crohn's disease. Few subjects may demonstrate a history of Crohn's disease. Besides, indolent T lymphoproliferative disorder is a chronic condition associated with frequent disease relapses. Generally, the disorder is persistent and accompanied by prolonged clinical course with extended survival. Occasionally, the disorder may progress to aggressive T cell lymphoma, a neoplasm delineating adverse prognostic outcomes [1,2].

Upon microscopy, lesions demonstrate an intense lymphoid infiltrate confined to lamina propria or superimposed mucosal surface of gastrointestinal tract. Occasionally, the lymphoid infiltrate extends to subjacent submucosa. Lymphoid infiltrate is essentially non-destructive wherein adjacent mucosal glands appear displaced and distorted [3,4]. The lymphoid infiltrate is comprised of monomorphic, miniature lymphoid cells incorporated with minimal, pale staining cytoplasm, mildly irregular nuclear contour, mature nuclear chromatin and inconspicuous nucleoli. Indolent T lymphoproliferative disorder incriminating the oral cavity may manifest as ulcers accompanied with subjacent infiltrate of lymphoid cells. Characteristically, lesions are devoid of necrosis, ulceration or foci of angiocentric lymphoid infiltrate and angiodestruction [3,4] (Figures 1,2 & Table 1).

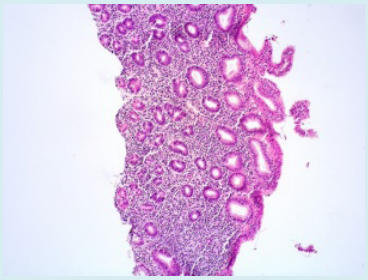

Figure 1: Indolent T lymphoproliferative disorder of gastrointestinal tract demonstrating a mucosal infiltrate of lymphoid cells incorporated with minimal cytoplasm, irregular nuclear contour, coarse nuclear chromatin and indiscernible nucleoli. Foci of mucosal ulceration, necrosis, angio-destruction or angio-centricity are absent.

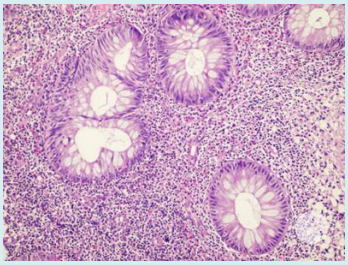

Figure 2: Indolent T lymphoproliferative disorder of gastrointestinal tract delineating mucosal infiltration of monoclonal lymphoid cells permeated with scanty cytoplasm, irregular nuclear outline, mature nuclear chromatin and inconspicuous nucleoli. Foci of mucosal ulceration or necrosis are absent.

Gastrointestinal disease	Presenting clinical symptoms	Recurrent genetic alterations
Enteropathy associated T cell lymphoma	Abdominal pain, anorexia, adenopathy, fatigue, infection	Activating mutations of JAK1 and STAT3
Inflammatory bowel disease •ulcerative colitis •Crohn's disease	Abdominal pain, cramps, diarrhoea, haematochezia, fever, fatigue, anorexia, unintended weight loss	Germline mutations of IL10RA, IL10RB, integrin genes
Indolent T lymphoproliferative disorder of GI tract	Abdominal pain, diarrhoea, dyspepsia, food intolerance, vomiting	STAT3-JAK2 fusions in CD4+ lesions
NK cell enteropathy	Constipation, diarrhoea, GI bleeding, vague abdominal pain, vomiting	JAK3 K563_CD565del
Monomorphic epitheliotropic intestinal T cell lymphoma	Abdominal pain, bowel obstruction, diarrhoea, intestinal bleeding, perforation, weight loss	STAT5B, N642H, loss of function mutations of SETD2

Table 1: Differential Diagnosis of Indolent T Lymphoproliferative Disorder of Gastrointestinal Tract [3,4].

Indolent T lymphoproliferative disorder of gastrointestinal tract appears immune reactive to CD2, CD3, CD5, CD7, T cell intracellular antigen 1(TIA1) and T cell receptor beta F1 (TCR-BF1). Besides, lesions exhibiting an immuno-phenotype CD4+ / CD8- or CD4- / CD8+ may be equivocally delineated. Exceptionally, an immuno-phenotype of CD4+ / CD8+ or CD4- / CD8- may appear. Decimation of CD7 is infrequently exemplified. Indolent T lymphoproliferative disorder of gastrointestinal tract is associated with minimal Ki67 proliferative index of ~5%. Indolent T lymphoproliferative disorder of gastrointestinal tract is immune non-reactive to CD30, CD56, granzyme, T cell receptor gamma delta (TCRγδ) or Epstein Barr virus (EBV) [5,6].

Upon flow cytometry, indolent T lymphoproliferative disorder of gastrointestinal tract simulates cellular immuno-phenotype of mature T lymphocytes. Exceptionally, aberrant decimation of CD7 is observed. Indolent T lymphoproliferative disorder of gastrointestinal tract delineates clonal genetic rearrangements of T cell receptor (TCR) as discerned by polymerase chain reaction (PCR) assay. Repetitive chromosomal translocation t (9; 17) (p24.1; q21.2) STAT3/JAK2 genetic fusion is encountered, especially within instances immune reactive to CD4+ [5,6]. Genetic alterations as reoccurring genetic mutations and novel rearrangements incriminating genes configuring JAK/STAT pathway are frequently observed [5,6].

Indolent T lymphoproliferative disorder of gastrointestinal tract requires segregation from neoplasms such as intestinal T cell lymphomas as denominated by enteropathy associated T cell lymphoma (EATL) and monomorphic epitheliotropic intestinal T cell lymphoma (MEITL), celiac sprue and inflammatory bowel disease (IBD) as ulcerative colitis or Crohn's disease [5,6]. Surgical tissue sampling of site incriminated within gastrointestinal tract is optimal for discerning the condition [5,6].

Indolent T lymphoproliferative disorder of gastrointestinal tract demonstrates an unsatisfactory therapeutic response to conventional chemotherapy and immunotherapy. Currently, a suitable treatment regimen employed to manage the condition remains undefined. Factors contributing to pertinent prognostic outcomes remain undocumented [5,6].

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