A Case of Indolent T cell Lymphoproliferative Disease of GI tract

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Introduction: Indolent T cell lymphoproliferative disease of the gastrointestinal tract is a benign lymphoid neoplasm provisionally recognized by the World Health Organization classification of lymphoid neoplasms in 2016 as a distinct disease entity. Indolent GI T-LPD generally show non-invasive pattern and protracted clinical course with long indolent phase.

Case report : A 80-year-old male presented to the outpatient for colonoscopy for the purpose of health screening. The 4.5cm * 2cm sized polypoid lesion in the sigmoid colon was observed 15cm above from anal verge. After admission, the polypoid lesion was excised with endoscopic submucosal dissection method(Figure 1). The excised lesion showed thickening in mucosa level and monomorphic lymphoid infiltration in lamina propria level with a non-destructive appearance(Figure 2A). The lymphoid cells were positive for CD3, CD4, CD5, CD8 and negative for CD20, CD56, CD79a. KI-67 index was low positive(Figure 2B,C). Molecular pathology analysis revealed a monoclonal T-cell receptor Beta, Delta, and Gamma gene rearrangement. From the results, it was possible to diagnose indolent T cell lymphoproliferative disease. In follow up colonoscopy after 4 months, no recurrence was found at the previous resection site. However, another whitish nodular lesion on rectum was observed at 5cm above from AV. In immunohistochemistry analysis, the lymphoid follicles from the lesion was positive for CD3, CD20 and CD79a. He is alive without any symptom, and is undergoing further examination.

Discussion: It is important to distinguish Indolent GI T-LPD with GI T-cell lymphoma because the therapeutic course for GI T-cell lymphoma should be more aggressive. In most cases the chemotherapy showed no obvious benefit for indolent T cell lymphoproliferative patients. In Immunohistological analysis, Indolent GI T-LPD usually shows T-cell receptor rearrangement. Since misdiagnosis of the GI tract disease can lead to delay in proper therapeutic approach, TCR clonality analysis and molecular genetic test should be applied for differential diagnosis.

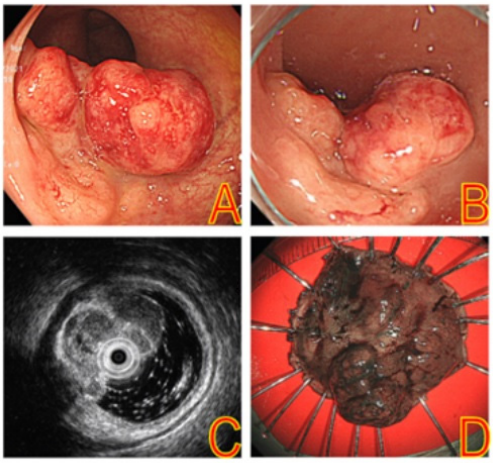

Figure 1. (A), (B) 4.5 cm x 2cm sized polypoid subepithelial lesion with lobulation was observed 15cm above AV during colonoscopy (C) EUS showed hypoechoic lesion throughout mucosa and submucosa layers. (D) the resected tissue through ESD showed thickened mucosa lobuation.

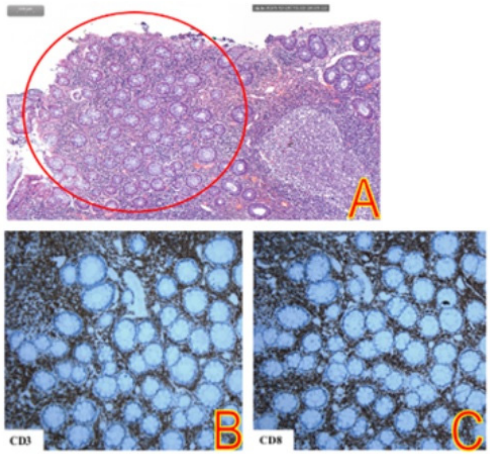

Figure 2. (A) small sized monomorphic lymphoid infiltrations were noted in between glands of lamina propria. (B), (C) immunohistochemical stain revealed CD3 and CD8 positive test result for the lesion