Turkish Journal of Cancer
Volume 38, No. 2, 2008

Images in hematology-oncology

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Sigmoid colon tumor in a 31-years old female

CLINICAL HISTORY

Endoscopic biopsy and surgical specimen of a 31-years old female was sent for consultation to our institute. The referring diagnosis of the endoscopic biopsy from sigmoid colon was indifferentiated malignant neoplasm. The sigmoid colon resection material was diagnosed as reactive changes with no evidence of malignancy.

PATHOLOGY

The microscopic examination of endoscopic specimen showed ulcerated tissues infiltrated with neoplastic cells which displayed large hyperchoromatic nucleus and small granular cytoplasm with a frequent mitotic and apoptotic figures. There were focal areas of necrosis. Immunohistochemical studies revealed that these neoplastic cells were positive for CD2, CD3, CD7 and Granzyme B, and focally expressed CD8 and EBER (by in situ hybridization). They were negative for CD4, CD5, CD20, CD30, CD56, CD79a, ALK and MUM-1 (Figure 1).

In the surgical specimen there were large areas of ulceration in the colonic mucosa and inflammatory reaction throughout mesenteric adipose tissues. There was focal necrosis and small foci of viable neoplastic cells were present in the intervening areas. These neoplastic cells were pleomorphic with irregular nuclear membrane, hyperchromatic nuclei and inconspicuous nucleoli. The neoplastic infiltrate had angiocentric growth pattern (Figure 2).

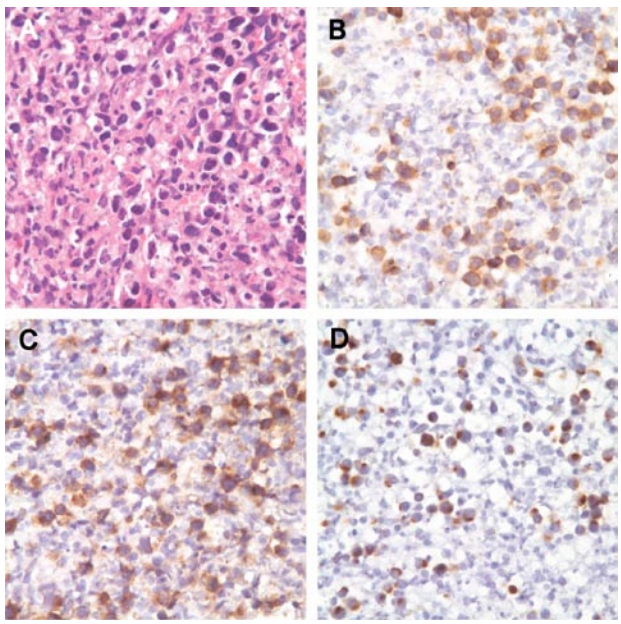

Fig 1 (A,B,C,D). (A): Neoplastic infiltrate mixed with inflammatory cells in endoscopic biopsy sections. Tumor cells with large hyperchromatic nucleus expressed CD3 (B), CD7 (C) and Granzyme B (D)

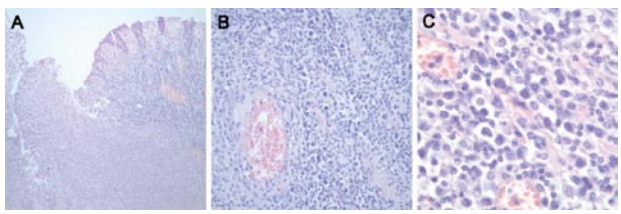

Fig 2 (A,B,C). (A): Ulceration in the colonic mucosa and mixed inflammatory-neoplastic infiltrate in surgical colectomy specimen, (B,C): Medium-sized to large neoplastic cells in submucosa

What is your diagnosis?

PATHOLOGIC DIAGNOSIS

Peripheral T-cell lymphoma, high grade, cytotoxic/suppressor T-cell phenotype.

DISCUSSION

Primary lymphoma of gastrointestinal tract most commonly involves the stomach then followed by the small intestine and colorectum. They are generally B-cell phenotype. Primary T/natural killer (NK) cell lymphoma of the colon is extremely rare. The classification of T-cell and NK-cell neoplasms proposed by the WHO classification emphasises a multiparameter approach, integrating morphologic, immunophenotypic, genetic, and clinical features. Clinical features play particular importance in the subclassification of these tumors, in part due to the lack of specificity of other parameters. Mature T-cell (post-thymic) and NK cells share similar immunophenotype and functional properties. Because of that reason neoplasms of these cells are classified under the same subtype in the WHO histological classification.

Extranodal NK/T-cell lymphoma (ENTCL), nasal type, is a predominantly extranodal lymphoma characterized by a broad morphological spectrum, infiltrate is often angiocentric, with prominent necrosis and vascular destruction. It is designated NK/T-cell lymphoma because while most cases appear to be NK-cell neoplasm (EBV+ CD56+), rare cases show an EBV+ CD56- cytotoxic T-cell phenotype. Enteropathy-type T-cell lymphoma (ETCL) is a tumor of intraepithelial T-lymphocytes, showing varying degrees of transformation but usually presenting as a tumor composed of large lymphoid cells. This tumor occurs most commonly in the jejunum or ileum and is usually associated with coeliac disease.

Both ENTCL, nasal type and ETCL in GI tract form ulcerating mucosal masses that invade the wall of the intestine. These cases can cause diagnostic challenge to physicians and pathologists. Early symptoms and colonoscopic findings may be similar to those of inflammatory bowel disease. Neoplastic cells show variable cytological appearances, including a prominent mixture of inflammatory cells, such as normal appearing small lymphoid cells, plasma cells, and less often eosinophils and histiocytes. Therefore, it may be difficult to distinguish the disease from an inflammatory or infectious process as in our case.

The morphologic and immunophenotyping studies show a peripheral T cell lymphoma most consistent with either ETNCL, nasal type or ETCL. Due to the lack of clinical information the case could not be further classified. A history of coeliac disease would support ETCL. Although the case is negative for CD56, rare cases of CD56 negative (CD3+ CD56+/-) ENTCL, nasal type have been published in the literature. Focal EBER (by in situ hybridization) positivity and colonic localization favor ENTCL, nasal type. Several authors have suggested that CD56 expression on tumor cells plays an important role in early dissemination. The disease often has a rapid progressive course and poor prognosis.

References

1. Jaffe ES, Harris NL, Stein H, et al., editors. WHO Classification of Tumors: Tumors of Haematopoietic and Lymphoid Tissues. Lyon: IARC Press, 2001.
2. Loehr WJ, Mujahed Z, Zahn FD, et al. Primary lymphoma of the gastrointestinal tract: A review of 100 cases. Ann Surg 1969;170:232-8.
3. Shepherd NA, Hall PA, Coates PJ, et al. Primary lymphoma of the colon and rectum. A histological and immunohistochemical analysis of 45 cases with clinicopathological correlations. Histopathology 1988;12:235-52.
4. Okada M, Maeda K, Suzumiya J, et al. Primary colorectal T-cell lymphoma. J Gastroenterol 2003;38:376-84.
5. Kim HS, Lee DK, Baik SK, et al. Primary CD56+ T/NK cell lymphoma of the colon. J Gastroenterol 2002;37:939-46.