Primary gastric T-cell lymphoma: A case report

ABSTRACT

Primary gastric T-cell lymphoma is a rarely encountered lesion causing difficulties in endoscopic biopsy diagnosis. We present a case of gastric T-cell lymphoma with its clinical presentation, pathological, and laboratory findings, and discuss the differential diagnoses, and the biology of these lymphomas. The patient was a 55 year old male presenting with abdominal pain and nausea. He had two negative gastric endoscopic biopsies but was operated based on the clinical, and radiological suspicion of gastric cancer. Pathological examination revealed a diffuse infiltration of the stomach wall with neoplastic lymphoid cells. Immunohistochemical studies revealed leukocyte common antigen (LCA), polyclonal T-cell marker (CD3), and focal human natural killer cell-like marker (NK-like) positivity of the tumor cells. The patient had regional lymph node involvement, therefore was staged as IIE (Ann Arbor). The patient died six months after the initial diagnosis. The case illustrates the difficulty in diagnosing these tumors by endoscopic superficial biopsies. [Turk J Cancer 2004;34(1):35-37]

KEY WORDS:
Stomach, T-cell lymphoma, gastric lymphoma

INTRODUCTION

The incidence of non-Hodgkin lymphoma (NHL) arising from extranodal tissue has been reported to be increasing in Western countries (1). The most common sites of primary extranodal NHL were gastrointestinal tract and head & neck region (2-5). The issue of the most common locations of NHL in gastrointestinal tract is controversial (2,3). Primary gastric lymphoma is a relatively uncommon malignancy, representing 1-7% of all malignant neoplasms of stomach (2,6). Most cases of gastric lymphoma are of B-cell origin, while primary gastric T-cell lymphomas are rarely reported (2,7).

In this paper, we present a case of primary gastric T-cell lymphoma due to its rare occurrence and the difficulties in diagnosing these cases endoscopically.

CASE REPORT

A 55-year old man presented with epigastric pain and nausea. Endoscopic examination revealed a 4x4x3 cm ulcer located on the posterior aspect of the large curvature. Clinically a malignant ulcer was suspected. Initial biopsies were reported as “chronic superficial gastritis”. The patient reported a previous endoscopic examination a year ago which was biopsied, and had the same benign diagnosis. The biopsy was repeated and histopathologic examination revealed chronic active gastritis and ulceration with no
evidence for Helicobacter Pylori (HP) infection. Despite the benign diagnoses, based on the clinical suspicion the patient was operated. A partial gastrectomy was performed.

On macroscopic examination a large ulcerating, focally vegetating mass measuring 12x6x3 cm encompassing the antrum, distal lesser curvature, and spreading to the most of the larger curvature was found. On cross sectioning, the stomach wall was also found to be infiltrated with the tumor (Figure 1).

On histologic examination, a diffuse lymphocytic infiltrate intermingled with eosinophils, plasma cells, and few neutrophils were seen. The infiltrate had an angiocentric pattern (Figure 2) and focal perineural spread was also seen. The tumor cells were pleomorphic, bearing large pale nuclei with irregular contours, prominent nucleoli, and sparse cytoplasm. The tumor cells were observed throughout the stomach wall, spreading to the serosa. Four of the lesser curvature lymph nodes were also involved. Immunohistochemical examination of the tumor cells revealed that they were LCA, CD3, NK-like (CD57) positive; CD15 (leuM1), CD30 (BerH2), CD79a (B cell marker), bc12 (antiapoptotic protein), anaplastic lymphoma kinase (ALK), and Epstein-Barr virus marker (LMP1) negative. Based on clinical, morphologic, and immunohistochemical findings a diagnosis of “primary gastric T-cell lymphoma” was rendered. On radiological studies, lymph node enlargement was noted at the hepatic hilus and peripancreatic region. Bone marrow biopsy was negative for lymphoma involvement. Based on these findings the patient was staged as IIE (Ann Arbor). The patient was treated with one cycle of ProMACE-CytaBOM (Prednisolone, adriamycine, cyclophosphamide, etoposide, cytarabine, bleomycine, oncovine, metotrexate) The patient refused further treatment and expired at 6 months after the initial diagnosis.

DISCUSSION

Primary non-MALT lymphomas of the gastrointestinal system are rare, yet the epidemiology of this heterogeneous disease has been poorly described (1). In Turkey, according to Dinçol (5) small intestinal lymphoma (48.8%) occurs more frequently than gastric lymphoma (39.9%), on the other hand according to Sarpe (3) gastric lymphoma (43%) is more prevalent than intestinal lymphoma (30%).

In the intestines, enteropathy associated T cell lymphomas (EATL), large granular cell lymphoma and oral and colonic ulcerations with monoclonal T-cell infiltrates have been described (2,6,8,9). Primary T cell lymphomas of the stomach are rare and their clinicopathologic description, and classifications are not well described. Lymphomas of the stomach comprise 1-7% of the malignant neoplasms of the stomach (2). Most of these lymphomas are of B cell origin (6,8,9). Primary T-cell lymphoma of the stomach was first described in 1985 by Weiss et al. Etiology and incidence of these lymphomas are not well known. In the cases reported from Japan, 7% of the stomach lymphomas are of T-cell origin (2). Initially these lymphomas were thought to be of viral origin, thus the presence of Human T-cell leukemia virus (HTLV-1) and Epstein-Barr virus (EBV) in the tumor samples were studied. No increased incidence of gastric T-cell lymphoma in areas endemic for HTLV-1 was noted. However, a significant number of primary gastric T-cell lymphomas are HTLV-1 positive (2,3,6-9). In our case, HTLV-1 was not studied. To our knowledge, in Turkey HTLV-1 has not been reported so far.

Fig 1. Infiltrative tumor under intact mucosa (H&E, x40) and high power (H&E, x400)

Fig 2. Angiocentric pattern of the tumor (H&E, x200)
EBV was found in several B-cell lymphomas of the stomach while very few of the T cell lymphoma cases were EBV positive. In our case EBV is both serologically and immunohistochemically (LMP1) negative. The issue of EBV presence in primary gastric T-cell lymphomas is not settled despite several reports (2,7,10,11).

The role of HP in the development of gastric lymphomas of the MALT type has been well established. However, any role for HP in the development of gastric T-cell lymphomas is not known (6,10,12). In our case HP was negative both by urease test and immunohistochemically. There are few reports on the endoscopic presentation of gastric lymphomas (6). In our case, endoscopically a gastric carcinoma was suspected. It may be difficult to diagnose gastric T-cell lymphomas based on superficial biopsies of the stomach (6,11). The depth of the biopsy will determine the diagnostic accuracy of the procedure.

Shepherd et al. (12) investigated 250 GIS lymphomas. Among these cases 28 were T-cell lymphomas (26 small bowel, 2 stomach). Most of these tumors were characterized by a dense eosinophilic infiltrate interspersed in between the tumor cells. This is probably due to the release of cytokines such as IL-3, IL-5, and GM-CSF by the tumor cells. In our case, presence of eosinophils and plasma cells among the tumor cells is striking.

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The immunohistochemical profiles of gastric lymphomas are also controversial. There have been reports of epithelial membrane antigen (EMA) and cytokeratin positivity in T-cell lymphomas, especially the anaplastic large cell lymphomas (13). Most T-cell lymphomas of the stomach are of cytotoxic T/NK cell origin. Our case expresses NK-like (CD57) marker, and demonstrates angiocentricity supporting the hypothesis that these tumors are of cytotoxic T/NK cell origin. In our case, cytokeratin and EMA were negative while LCA was positive. Hodgkin lymphoma, follicular lymphoma, and anaplastic large cell lymphoma diagnoses were excluded based on the morphologic features, and the negativity for CD15, CD30, ALK, and CD79a. The patient was screened carefully and no other organ or tissue involvement was found except for the regional lymph nodes. Gastric T-cell lymphomas reportedly have worse prognoses than their B-cell counterparts (6,8,10). Especially p53 positivity has been associated with a worse outcome (5,10). In our case p53 was also found to be positive.

In this case, the patient was treated with ProMACE-CytaBOM protocol owing to gastric T-cell lymphomas have worse prognosis than gastric B-cell lymphomas (5,12). In summary, we presented a case of gastric T-cell lymphoma due to its rare occurrence, and to make diagnosticians aware of the difficulties in diagnosing these cases by endoscopy and superficial biopsies.

References