NON-HODGKIN LYMPHOMA OF GASTRO-INTESTINAL TRACT-CLINICAL PRESENTATION, EVOLUTION AND TREATMENT.

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Introduction.Extranodal lymphomas arise anywhere outside the lymph node region: from sites with primary lymphoid organs (spleen, thymus,Waldeyer ring); from organs or tissues devoid of lymphoid tissue (brain, soft tissue); or from organs with a significant lymphoid tissue component (gastrointestinal tract). In the gastrointestinal tract, lymphoid elements occur in the lamina propria and submucosa,but primary or secondary lymphomatous neoplasms may occur in any portion of the gastrointestinal tract(1).

Secondary gastrointestinal involvement is common because of the frequent origination of lymphomas in the mesenteric or retroperitoneal nodes. The primary lymphomas of the gastrointestinal tract usually involve only one site.The most commonly involve the stomach but can involve any part of the gastrointestinal tract from the esophagus to the rectum.

Incidence and Pathogenesis.The incidence of primary gastrointestinal NHL is approximately one in 100,000 individuals per year. There is a male predilection, with a male-female ratio of 3:2. A number of risk factors other than HIV infection have been identified in the pathogenesis of gastrointestinal lymphoma: Helicobacter pylori(H pylori) infection, celiac disease, inflammatory bowel disease, immunosuppression after solid organ transplantation(1). Although there is no lymphoid tissue in the gastric mucosa, chronic H pylori infection is associated with the development of lymphoid tissue in the lamina propria. Most low-grade primary gastric lymphomas arise from this mucosa-associated lymphoid tissue (MALT) and are classified as MALT lymphomas. It has been suggested that high-grade lymphomas result from transformation of the low-grade tumor (2). Immunoproliferative small intestine disease, a special form of MALT lymphoma, is also suspected to have an infectious etiology (3). Celiac disease has been noted as a risk factor for small bowel adenocarcinomas, esophageal cancer, melanoma, and NHL (4).Celiac disease is often associated with enteropathy type T-cell lymphoma.Patients with HIV-induced immunodeficiency are at high risk for developing a B-cell phenotype intestinal lymphoma with unusual morphologic features, a high grade of malignancy, and a poor prognosis (5).

Esophagus.Esophageal lymphoma occurs secondary to cervical and mediastinal lymph node invasion or contiguous spread from gastric lymphoma. Primary esophageal lymphomas are predominantly B-cell type.The predominant appearance is that of submucosal infiltration, but may also manifest with a polypoid mass ulceration, or nodularity (6).

Perforation and fistulization may be demonstrated.Barium studies better demonstrate subtle mucosal and submucosal abnormalities and CT better defines the extent of local disease and the disease stage.

Stomach.Primary gastric lymphoma represents 1%–5% of gastric malignancies (7) and is the most common type of extranodal lymphoma, accounting for 50%–70% of all primary gastrointestinal lymphomas. It is recognized that chronic H pylori gastritis is associated with the development of low-grade MALT lymphoma.Primary gastric lymphoma originates as a low-grade MALT lymphoma,and transforms into intermediate or high-grade large cell lymphoma if not diagnosed or treated in time (8).

When it is diagnosed at an early stage has a good prognosis, and eradication of H pylori with antibiotic therapy has resulted in regression of early stage tumors. The double-contrast barium studies may reveal ulcerative, polypoid, or infiltrative patterns, which are the same as those of gastric carcinomas.Together with barium studies gastroscopie in all gastrointestinal lymphoma;demonstrats a lot of pathological aspects and offers possibility to do biopsy very important for diagnosis.The diagnosis of lymphoma may be suggested by the presence of multiple polypoid tumors with central ulceration ("bull' seye" appearance), giant cavitating lesions, or extensive infiltration with gastric fold thickening . The latter finding may be distinguished from linitis plastica on the basis of the preservation of gastric distensibility. That have been described:single or multiple ulcers of varying size; single or multiple masses with or without an ulcer, along with thickened folds; rugal thickening, commonly converging to an ulcer or a mass; mucosal nodularity of varying size, either focal or diffuse; and coarse areae gastricae(1). Low-grade MALT lymphoma has a wider spectrum of appearances than does high-grade MALT lymphoma, in which a mass-forming lesion or severe fold thickening is present (2).Preservation of the perigastric fat planes at CT is more likely to be seen in lymphoma than in adenocarcinoma, in the presence of a bulky tumor (9).The stomach remains pliable even with extensive lymphomatous infiltration, and the lumen is preserved, making gastric outlet obstruction a rather uncommon feature (10). Adenopathy is seen in both adenocarcinoma and lymphoma, but if it extends below the renal hila or the lymph nodes are bulky, lymphoma is more likely (11). Complications such as obstruction, perforation, or fistulization can occur and can be detected with CT and barium studies.

Small Bowel.Lymphoma is the most common malignancy of the small bowel (12), and its incidence related to B-cell hyperactivation in HIVpositive patients has increased. The small bowel lymphoma accounting for 20%– 30% of all primary gastrointestinal lymphomas. The distal ileum is the most common site of small bowel B cell lymphoma because of the greater amount of lymphoid tissue in this portion of the bowel.

Small bowel B-cell lymphoma may appear as a circumferential bulky mass in the intestinal wall with extension into the small bowel mesentery and regional lymph nodes.

The tumor may involve a long segment of bowel and may ulcerate and perforate into the adjacent mesentery.Aneurysmal dilatation of the lumen may be seen due to replacement of the muscularis propria and destruction of the autonomic nerve plexus by lymphoma.A focal, polypoid, homogeneous intraluminal mass without wall thickening or lymphadenopathy has been described (13).

Barium studies show single or multiple polypoid lesions,diffuse or segmental ulcerative or infiltrative change, or diffuse or focal nodularity. Peritoneal lymphomatosis from primary gastrointestinal lymphoma is rare compared with carcinomatosis.The prevalence of malabsorption and intestinal recurrence is high in enteropathy-associated Tcell lymphoma. The peripheral T-cell lymphoma is seen in the small intestine, particularly the jejunum and has a higher prevalence of multifocal involvement and bowel perforation(1).

Large Bowel.Primary lymphoma of the large bowel accounts for 0.4% of all tumors of the colon, and colorectal lymphomas constitute 6%–12% of gastrointestinal lymphomas (14). Primary lymphoma often affects the cecum and rectum than other parts of the large bowel (15). The primary colorectal lymphoma comprises low-grade B-cell lymphoma arising from MALT, mantle cell lymphoma, and T-cell lymphoma (14). Most colorectal lymphomas are NHL, usually of B-cell origin. Mantle cell lymphoma is an aggressive disease that manifests as multiple polyps (lymphomatous polyposis). Low-grade B-cell lymphoma arising from MALT has an indolent course and might also manifest as multiple polyps. Peripheral T-cell lymphoma of the colon manifests as either a diffuse or a focal segmental lesion with extensive mucosal ulceration at double-contrast barium enema examination (1). The colonic perforation frequently occurs.There are the following findings in lymphoma: polypoid masses,frequently near the ileocecal valve; circumferential infiltration,a cavitary mass excavating into the mesentery; endoexoenteric tumors; mucosal nodularity; and fold thickening (14).The focal strictures, aneurysmal dilatation, or ulcerative forms with fistula formation may be seen. Features that help differentiate lymphoma from adenocarcinoma include extension into the terminal ileum, well-defined margins with preservation of fat planes, no invasion into adjacent structures, and perforation with no desmoplastic reaction (16). Primary rectal lymphoma is a rare type of gastrointestinal lymphoma and is clinically indistinguishable from rectal carcinoma.Primary lymphoma of the appendix is very rare,it is more common to see cecal lymphoma extending to the base of the appendix .Patients typically present with acute symptoms that are suggestive of acute appendicitis.

Treatment. There is still no consensus on the optimal treatment for primary gastrointestinal lymphoma. Nowadays surgery is limited to rare cases and radiotherapy – combined or not with chemotherapy – represents an effective therapeutic option ensuring long-term, organ-salvage benefits mainly in aggressive histological subtypes. In the MALT lymphomas associated with H-pylori infection,antibiotics alone can induce lasting remissions.A global therapeutic approach has changed over the last 10 years: innovative, conservative options to reduce treatment toxicity, thus preventing systemic relapses, have made their appearance and are on the rise.

Conclusions. Gastrointestinal lymphoma is an uncommon disease with a wide variety of imaging appearances. Primary gastric lymphoma is a rare cancer of the stomach with an indeterminate prognosis.The features such as a bulky mass or diffuse infiltration with preservation of fat planes and no obstruction, multiple site involvement, and bulky lymphadenopathy are suggestive of lymphoma.

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