CASE REPORT

Extranodal marginal zone lymphoma of MALT involving the lungs, the stomach, and the colon

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Summary  
Extranodal marginal zone lymphoma of MALT, previously known as MALT lymphoma, is a low grade B-cell Non-Hodgkin’s lymphoma (NHL). Its most frequent locations are the gastrointestinal tract and the lungs while that of the colon is rare. Involvement of multiple mucosal sites is not a frequent finding but it does occur. We describe a case of a 70-year-old man who presented with extranodal marginal zone lymphoma of MALT involving three different sites: the lungs, the stomach, and the colon.

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1. Case report

A 70-year-old man, heavy smoker, presented to another institution with abdominal pain and melena with no associated constitutional, respiratory, or any other symptoms. Colonoscopy revealed multiple colonic polyps involving the transverse and sigmoid colon. Polypectomy of two of the sigmoid polyps was done along with multiple colonic biopsies. Pathology showed extranodal marginal zone lymphoma of MALT and nonspecific chronic colitis. Upper endoscopy showed a large deep ulcer on top of a polypoid mass at the lesser curvature of the stomach. Biopsies taken were consistent with extranodal marginal zone lymphoma of MALT, in addition to chronic severe active gastritis with helicobacter pylori seen on the surface epithelium. Chest radiography showed bilateral nodular infiltrates. Computed tomography of the chest revealed multiple bilateral patchy opacities without enlarged hilar or mediastinal lymph nodes (Fig. 1). Computed tomography of the abdomen and pelvis showed no abnormality.

Patient was started on helicobacter pylori eradication regimen and was referred to our institution for further work-up. At this point our main differential concerning the lung lesions was a primary lung cancer, in view of the patient’s long history of smoking and the radiographic appearance of the lesions. CT guided fine needle aspiration of one of the lesions in the left upper lobe of the lung showed extranodal marginal zone lymphoma of MALT. Bone marrow biopsy was negative for lymphoma. Patient was started on cyclophosphamide and rituximab. After three cycles of chemotherapy, a follow up chest radiography revealed clearing of the

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Fig. 1 CT scan of the chest with IV contrast showing multiple bilateral patchy infiltrates.

Fig. 2 CT scan of the chest shows persistence of minimal infiltrates in the right upper lobe.

bilateral infiltrates, and the computed tomography of the chest, abdomen, and pelvis showed persistence of only few ill defined patchy infiltrates in the right upper and middle lobes (Fig. 2).

2. Discussion

MALT lymphoma is a low grade B-cell NHL. It was first described by Isaacson and Wright in 1983 [1]. But only in 1994 it was recognized as a distinct entity of lymphoma in the Revised European—American Lymphoma (REAL) classification [2]. The REAL classification and later the WHO included the MALT lymphoma in the group of marginal zone B lymphomas [3]. MALT was described when the observation was made that the histology of certain extranodal lymphomas was similar to that of mucosa associated lymphoid tissue rather than to that of peripheral lymph nodes. MALT is defined by the presence of "lymphoid epithelial" lesion formed of monomorphic lymphocytic infiltrates surrounding the epithelial structures with invasion of these structures by atypical lymphocytes, and rare blastic cells may be seen [4]. MALT lymphoma cells typically surround reactive B-cell follicles in the distribution of the marginal zone and tend to involve this zone when they disseminate to lymph nodes and spleen. The MALT lymphoma B-cells have same cytological features and immunophenotype (CD20+, CD21+, CD35+, IgM+, IgD−) as that of marginal zone B-cells [4].

These tumors often arise within tissues involved by chronic inflammatory disorders of autoimmune or infectious etiology [4]. Examples include development of extranodal marginal zone lymphoma of MALT in the stomach after chronic antigenic stimulation by helicobacter pylori, in the thyroid secondary to Hashimoto’s thyroiditis, and in the salivary gland in association with sialadenitis in the setting of Sjogren’s syndrome. This inflammation will result in the accumulation of auto-reactive lymphoid tissue which over time becomes genetically unstable with the acquisition of chromosomal abnormalities leading to transformation into extranodal marginal zone lymphoma of MALT [5].

Clinically, about half of the patients with pulmonary involvement are asymptomatic at the time of diagnosis, and are incidentally identified on the basis of radiographic abnormalities. When present, symptoms are nonspecific and include cough, mild dyspnea, chest pain, and occasionally hemoptysis. Standard radiography findings are variable including single or multiple nodules, ill defined infiltrates, or consolidation. Computed tomography findings are bilateral and multiple alveolar opacities in 70% of the cases and reticulo-nodular infiltrates in less than 10% of the cases [6].

Extranodal marginal zone lymphoma of MALT is an indolent disease that remains localized for prolonged periods, even without treatment [7]. A variety of treatments including surgery, radiotherapy, and chemotherapy have been used with overall survival rates ranging from 80 to 90% at 5 years [7]. The regimen of cyclophosphamide and rituximab used in our case was chosen since cyclophosphamide and other alkylating agents have been used effectively in the management of extranodal marginal zone lymphoma of MALT [7]. Also, rituximab was chosen on the basis of preliminary data that it may have significant clinical activity [7].

The stomach is by far the most common localization of extranodal marginal zone lymphoma of MALT [8]. Involvement of multiple mucosal sites is well described but not common. Some cases with simultaneous gastric and intestinal involvement have been reported [7]. Thieblemont et al. reported that out of 158 patients with MALT lymphomas only 18 (11%) had multiple organ localization, 7 (3.5%) of which had stomach and intestinal tract involvement, and 5 (3%) of which had GI tract and lung involvement [5]. Grazziadei et al. reported a case of MALT lymphoma involving the lungs, the stomach, the lingual tonsils, and the bone marrow at the time of diagnosis [9]. The International Extranodal Lymphoma Study Group reported that out of 180 patients with non-gastric lymphoma only 24 (13%) had involvement of multiple mucosal sites [10].

In conclusion, extranodal marginal zone lymphoma of MALT tends to involve one site but multiple sites involvement
MALT lymphoma of the lungs, stomach, and colon does occur with sometimes unexpected combinations. This observation suggests the importance of following the guidelines in the work-up of patients presenting with extranodal marginal zone lymphoma of MALT in order to rule out other mucosal sites involvement, especially that the other mucosal site involved may be clinically silent. Furthermore, the differential diagnosis of pulmonary infiltrates in a patient known to have extranodal marginal zone lymphoma of MALT at any site should include extranodal marginal zone lymphoma of MALT involving the lungs.

References