Title

A Rare Gastrointestinal Spectacle - Primary Gastric Small Lymphocytic Lymphoma

Introduction

Gastrointestinal (GI) lymphomas account for 30–40% of extra nodal lymphomas, while primary GI lymphomas are rare (1%-4% of all GI malignancies). "Primary GI lymphoma" denotes tumors mainly localized in a specific GI organ, with or without regional involvement. The stomach is most frequently affected (60% of cases). Predominant subtypes include diffuse large B-cell lymphoma and extra nodal marginal zone lymphoma of mucosa-associated lymphoid tissue. Conversely, primary gastric small lymphocytic lymphoma (SLL) is extremely rare, with only a few documented cases.

Case Presentation

The patient is an 85-year-old male with paroxysmal atrial fibrillation on apixaban, Crohn's disease on mesalamine, and a history of gastroesophageal stricture dilation 12 years back, presented with food regurgitation and dysphagia with solid and liquid food but without odynophagia. Computed tomography (CT) of the chest showed a dilated fluid-filled esophagus. He underwent an endoscopy which revealed a tortuous presby esophagus without food impaction, diffuse gastritis, and a mass-like lesion in the mid/distal body. Endoscopic biopsies from the mass revealed small-sized CD20-positive B-cells. Endoscopic (transesophageal) ultrasound (EUS) imaging was done which revealed a 2.8 x 2.2 cm mixed echogenic lesion that appeared to arise from the submucosa within the distal gastric body (greater curvature). EUS fine needle aspiration biopsies of the gastric wall were sent for pathology, which revealed low-grade B-cell lymphoma, CD 20 positive and CD5 positive. Differential diagnoses included small lymphocytic lymphoma, extra nodal marginal zone lymphoma (malt lymphoma), and mantle cell lymphoma. LEF1 was diffusely positive and SOX11 was negative. This favors the diagnosis of SLL/CLL.

Discussion

SLL / CLL is a neoplasm of monomorphic small lymphocytes with SLL restricted to non-leukemic cases sharing the tissue morphology and immunophenotype of CLL. SLL is biologically related to CLL and lymphoplasmacytic lymphoma (LPL) or immunocytoma (Waldenstrom’s macroglobulinemia). The most common primary gastric lymphoma subtypes are mucosa-associated lymphoid tissue lymphoma and diffuse large B-cell lymphoma, followed by mantle cell lymphoma and follicular lymphoma. Primary gastric lymphoma other than these subtypes is extremely rare. SLL rarely involves the gastrointestinal tract, although it seems to have a predilection for the appendix. Mori et

al. reported an appendiceal SLL, treated only by ileo-cecal resection and the patient was disease-free for 36 months. Pasquale et al. reported an incidental finding of an appendiceal SLL during hernia repair. Handa and colleagues reported SLL in the small intestine leading to intussusception as a complication of CLL.5 In the English literature, there is a report of a case of localized gastric LPL with Waldenstrom's macroglobulinemia, but no report of primary gastric SLL. Our case had no evidence of CLL at diagnosis or during the follow-up period. The patient was unique in having stage IIE gastric SLL presenting with perforation. The same clonal origin of the gastric and regional nodal tumors is supported by the B-cell clonal study.