A Primary Diffuse Large B-Cell Lymphoma of Appendix

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Abstract

Primary appendiceal lymphomas are exceedingly rare neoplasms mostly ignored until histological examination of appendices. We report a case of primary appendiceal lymphomas in a 22–year-old man. Appendectomy was performed under diagnosis of acute appendicitis. Macroscopically a fleshy intramural mass in middle third portion of appendix was observed. Microscopic examination revealed a diffuse large B-cell lymphoma inducing acute appendicitis. Further medical workups of the patient was unremarkable and the disease was restricted to appendix. After initial simple appendectomy, the patient underwent adjuvant chemotherapy and was healthy after two years. This case emphasized great value of detailed histological examination of all appendices.

Keywords: Appendix; Lymphoma; Appendicitis

Introduction

Primary appendiceal lymphomas are extremely rare neoplasms that first time were reported by warren in 1899.¹ Appendiceal lymphomas constitute 0.015% of all gastrointestinal (GI) lymphomas.¹ ³ ⁴ A few cases of appendiceal lymphomas are diffuse large B-cell lymphoma (DLBCL).² ³ ⁷ We reported on a case of primary appendiceal DLBCL manifested as acute appendicitis in a 22-year-old man.

Case Report

We received an appendectomy specimen of a 22-year-old man in our department under diagnosis of acute appendicitis. The appendix measured 9.5 cm in length and 4 cm in greatest diameter and on opening, a round fleshy whitish–cream, intramural mass measuring 3.5 cm in diameter was observed that was partially obliterating appendiceal lumen. The appendix underwent routine tissue processing. By histological examination of haematoxylin-eosin stained slides, a neoplasm composed of atypical proliferated lymphoid series extended from mucosa to deep muscularis propria was noted (Figure 1A, 1B, 1C). The tumoral cells predominantly characterized by large vesicular nuclei, at times with prominent nucleoli. In addition, focal mucosal ulceration and neutrophilic infiltration were seen. By immunohistochemistry, these cells were positive for pan B-cell marker CD20, 35% for Ki-67 and non-reactive for CD45RO, CD30, bcl2 and bcl6. By further workup, no lymphadenopathy, organomegaly or gastrointestinal abnormality was found. These clinicopathological findings confirmed the diagnosis of primary appendiceal DLBCL simulating acute appendicitis. After initial appendectomy and a histological proven diagnosis, adjuvant chemotherapy was administered. The patient had no compliant or recurrence after two years of follow up.

Discussion

Malignant lymphomas compromise 1-4% malignancies of GI tract.⁸ Current studies showed only 0.015% of GI lymphomas to be primary appendiceal lymphomas.¹ Appendiceal lymphoma usually present in the
second and the third decades of life. These usually manifest as acute appendicitis and often are diagnosed postoperatively by histopathology. Rarely, their initial presentations are intussusceptions, lower GI bleeding, palpable abdominal mass and abscess. Grossly, the appendix may be diffusely enlarged and thickened, at time obliterated by a fleshy intramural tumor. The diameter of lymphomatous appendix ranged from 2.5 to 4.5 cm and their length from 7 to 17 cm. It is important that inflamed appendix without neoplasm rarely to be more than 1.5 cm in diameter.

Histopathologically, Burkitt’s lymphoma, mantle cell lymphoma, T-cell lymphoma, DLBCL and maltoma may primarily involve appendix. Here we described clinicopathologic features of primary appendiceal DLBCL in a 22-year-old man that presented as acute appendicitis. By review of literature, five scattered isolated cases of DLBCL were found that rarely manifested as acute appendicitis. According to previous reports and our recent observation, primary DLBCL of appendix is a very rare underlying disease associated with acute appendicitis that ignored until histopathologic examination of appendices. This case emphasized great value of detailed and careful histopathologic
examination of all appendices with clinical suggestion of acute appendicitis.

**Conflict of interest:** None declared.

**References**


