B-cell lymphoma of the appendix: A case report and review of literature

Abstract

Appendicular tumors are rare, and lymphoma of the appendix is rarer. A 50-year-old female patient presented with vague abdominal discomfort and lump in the right iliac fossa. The diagnosis of diffuse large B-cell lymphoma was made after laparotomy and histopathological examination (HPE) supported by immunohistochemistry study. For appendicular neoplasms diagnosed postoperatively, including lymphoma, a meticulous grossing and HPE cannot be over emphasized. In case of wall thickening of >2.50-3 cm detected by the computed tomogram scan, the possibility of a neoplasm or lymphoma in particular should be included as the differential diagnosis irrespective of the clinical presentation.

Key words: Appendix, intestinal tract, lump abdomen, lymphoma

INTRODUCTION

Lymphomas of the small intestine are not very uncommon. However, appendicular lymphoma is a rare entity despite the fact that appendix is rich in lymphoid cells. Neoplasms as a whole are uncommon in the appendix. Collins has shown an overall incidence of 4.6% for benign tumors and 1.35% for the malignant tumors of the appendix in large study of appendicular specimen.[1] Malignant tumors of the appendix include carcinoid tumors and adenocarcinomas. The first case of primary lymphoma of the appendix was reported by Warren in 1898. Dawson et al. had cited five criteria that must be met for the diagnosis of a primary gastrointestinal lymphoma.[2] The criteria’s are no palpable superficial lymph nodes, chest radiographic findings should not reveal adenopathy, the white blood cell count (both total and differential) should be normal, at laparotomy the alimentary lesion is predominantly involved with lymph node involvement (if any) confined to the drainage area of the involved segment of gut, and there should be no involvement of the liver and spleen. Here, we present a case of primary appendecial B-cell lymphoma and review available literature related to it.

CASE REPORT

A 50-year-old female patient presented with the complaint of vague abdominal discomfort and occasional pain in the lower abdomen of 2 months duration. The patient was not having any symptom of associated bowel and bladder disturbances. On examination, there was an ill-defined, tender and partially mobile lump on the right iliac fossa region. Computed tomogram (CT) scan of the whole abdomen suggested an inflammatory pathology in the right iliac fossa with normal cecum and terminal ileum. Appendix could not be visualized separately. CT guided fine-needle aspiration cytology (FNAC) was done. However, FNAC showed necrotic material with neutrophils, reactive lymphoid cells and few histiocytes only. No granuloma and other abnormality were seen. Furthermore, CT guided aspirate for acid fast Bacilli (AFB) did not reveal presence of AFB. Exploratory laparotomy was performed under clinical diagnosis of acute appendicitis. Intra operatively a grossly swollen, elongated appendix was found without adhesion and appendicectomy was performed along with removal of a mesenteric lymph node. Grossly, the appendix was 10 cm long with thickened wall and the maximal internal diameter was 2.5 cm [Figure 1]. Cut section showed obliterated lumen with grey-white homogenous surface. Histopathological examination (HPE) revealed diffuse involvement of the wall of appendix by moderately uniform large cells with vesicular
nuclei and prominent nucleoli and many mitotic figures [Figure 2a and b]. HPE of the accompanying mesenteric lymph node showed necrotizing lymphadenitis. Immunohistochemistry (IHC) was done for CD45, CD20, CD3, cytokeratin, chromogranin and synaptophysin. IHC study was positive for the expression of CD45 [Figure 3a] and CD20 [Figure 3b]. Hence, the final diagnosis of diffuse large B-cell lymphoma (DLBCL) of the appendix was made. Further, bone marrow examination was done, which did not reveal infiltration by the tumor. The case was subsequently referred for chemotherapy.

**DISCUSSION**

Neoplasms of the appendix are very uncommon and usually diagnosed at operation or autopsy. Benign tumors of the appendix are relatively common. Benign tumors of the appendix consist of leiomyoma, neuroma and lipoma. The incidence of appendicular lymphoma varies from 0.02% to 2.6%, respectively. Our case was one in 239 appendicular specimens (0.41%) at the Department of pathology at our center in the last 3 years. There is also a previous report of well-differentiated lymphocytic lymphoma of appendix elsewhere. Mean age of patients with appendicular lymphoma has been shown to be around 54 years (range: 42-62 years). Our patient was 50-year-old female. The modes of presentation are varying. Commonly patients present with symptoms and signs of acute appendicitis. Appendicular lymphomas are also incidental findings intraoperatively and may present with nonspecific symptoms such as abdominal pain, anorexia, nausea, vomiting, fever, and weight loss. Rarely, it may present with intussusceptions or lower gastrointestinal bleeding. In our case, also the symptoms mimicked appendicitis and radiologically it appeared as an appendicular lump. Diameter of the appendix involved by a malignant lymphoma is of concern for diagnostic purpose. The Armed Forces Institute of Pathology reported five cases of non-Hodgkin's lymphoma of the appendix discovered retrospectively on CT scan, where the maximal appendiceal diameter ranged from 2.5 to 4 cm. The differential diagnosis of diffusely enlarged appendicular mass are benign conditions such as appendicitis, mucinous epithelial neoplasms, and neuroendocrine tumors such as carcinoid tumor, and paraganglioma. In appendicitis the diameter of the appendix usually does not exceed 15 mm. Neuroendocrine tumors shows infiltrative growth pattern, and involves distal appendix rather than the circumferential involvement. Other neuroendocrine tumors are rare, but may resemble the enlargement like that of lymphoma. Mucinous epithelial neoplasms generally are associated with mucoceles, seen on CT scan as a cystic dilatation of the lumen with or without calcifications. Areas of focal nodular solid enhancement of the wall favor the presence of mucinous cystadenocarcinoma.

Out of all the reported cases of lymphoma in the intestinal tract so far, there are cases that were reported before 1968 when the old nomenclature of lymphosarcoma was used. Rest of the cases reported in the last three decades, of which nine cases were of Burkitt's lymphoma, which is the majority, two cases of mental zone lymphoma, two cases of marginal zone B-cell lymphoma, three cases of T-cell lymphoma and four cases of DLBCL. However, since cases of lymphoblastic lymphoma and DLBCL in the old literature were not further classified, the case number of DLBCL might be underestimated.

**CONCLUSION**

We have described here a rare case of primary DLBCL of the appendix. To the best of our knowledge, until presenting this report there are only five cases in the literature previously reported. For appendicular neoplasms diagnosed postoperatively, including
lymphoma, a meticulous grossing and HPE cannot be over emphasized. Irrespective of the clinical presentation, in case of wall thickening of >2.5-3 cm on the CT scan, the possibility of a neoplasm or lymphoma in particular should be included as the differential diagnosis.

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