Primary intestinal diffuse large B cell lymphoma presenting as multiple lymphomatous polyposis: A case report

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Abstract

Multiple lymphomatous polyposis (MLP) is a rare extranodal manifestation of lymphoma. In most cases, multiple lymphomatous polyposis is associated with mantle cell lymphoma and in rare instances it is associated with diffuse large B cell lymphoma (DLBCL). We report a case of a 30-year-old male who presented as a case of acute intestinal obstruction, with weight loss, suspected to be intestinal tuberculosis initially. Laparotomy was done and affected segment of small intestine was resected and sent for histopathological examination. Grossly there were multiple small sessile polyps affecting a segment of small intestine. Histopathological and immunohistochemical studies on biopsy specimens from small intestine confirmed gastrointestinal Non- Hodgkin lymphoma, of diffuse large B cell type. Though mantle cell lymphoma is the main cause of multiple lymphomatous polyposis of gastrointestinal system, but diffuse large B cell lymphoma should also be considered, as prognosis of the two variants may differ considerably.

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INTRODUCTION

Primary gastrointestinal lymphomas most commonly present as a single lesion. In 4.3 % of cases gastrointestinal system lymphomas occurred as 2 to 6 separate nodules at one anatomic location and in only 2.6 % were described as multiple lymphomatous polyposis involving colon and small intestines¹. Multiple lymphomatous polyposis is an extranodal involvement of lymphoma, which is characterized by multiple polypoid tumors which affect long segment of the gastrointestinal system, as described for the first time by Cornes in 1961². Multiple lymphomatous polyposis is thought to represent

mantle cell lymphoma of the gastrointestinal tract and the MLP cases associated with Diffuse large B cell lymphoma are very rare³. We describe the clinical, histopathological and immunohistochemical features of a case of primary intestinal diffuse large B cell lymphoma, presenting as multiple lymphomatous polyposis, a rare combination.

CASE REPORT

A 30-year-old male patient presented to hospital with chief complaints of nausea and vomiting and had a history of recent weight loss. After evaluation of the patient with radiological examinations, the diagnosis of acute intestinal obstruction was made, and the patient underwent an emergent operation. Ileal resection was performed during the operation and the resected segment of ileum was sent to our department for evaluation. Grossly, multiple small sessile polyps ranging in diameter from 0.5 - 1.5 cms were seen on the cut section of the resected segment of intestine, along with few small ulcerated areas. A very little patchy, healthy-appearing mucosal area was also noticed in the resected segment of the intestine (Fig 1). Microscopic examination of the haematoxylin and eosin (H and E) stained sections

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showed a tumor infiltrating the intestinal tissue and exhibiting a diffuse pattern. The tumor tissue was found to be comprised of pleomorphic, atypical lymphoid cells with eosinophilic cytoplasm, vesicular nuclei, irregular nuclear membrane and prominent nucleoli. There were many mitotic figures seen in the sections. Also seen were bizarre looking tumour gaint cells (fig 2a, fig 2b). A B-cell phenotype immunoreaction was observed with CD30, leucocyte common antigen (LCA) and CD79a in tumour cells (fig 3a, fig 3b, fig 3c). No immunoreaction was observed in the tumor tissue with pancytokeratin, epithelial membrane antigen (EMA), CD10 and CD20 (fig 3d, fig 3e). The Ki 67 proliferative index was approximately 80% (fig 3f). The diagnosis of diffuse large B-cell lymphoma (DLBCL) was made.

DISCUSSION

Primary gastrointestinal system lymphomas are rarely encountered lesions. These lymphomas are usually observed as ulcerative, superficial, polypoid, or diffuse lesions. The areas frequently involved by gastrointestinal system lymphomas are mainly the stomach, followed by the duodenum, the jejunum, ileum and colon⁴. In our case the affected areas are in the ileum. The lesions of gastrointestinal multiple lymphomatous polyposis are represented by multiple sessile or pedunculate polyps, of a few millimeters to several centimeters in size⁵. As in our case, MLP involves single segment of the intestine,

and features smooth and sessile multiple polypoid structures with diameters ranging between 0.5cms and 1.5 cms. Both radiologic and endoscopic appearances in these cases mimic inflammatory bowel diseases, particularly Crohn's disease. Besides these benign lesions, similar macroscopic features are observed in malignancies such as mantle cell lymphoma, mucosa-associated lymphoid tissue (MALT) lymphoma, follicular lymphoma, B-cell chronic lymphocytic leukemia, and adult T cell lymphoma, and those malignancies should be considered in the differential diagnosis as well³. Diffuse large B-cell lymphoma (DLBCL), the largest subgroup of non-Hodgkin lymphomas, is the most frequent type displaying the extranodal involvement. The most common extranodal involvement locations reported for DLBCL cases are the stomach, bone, skin, small intestine, lung, heart, liver, and the genitourinary system. DLBCL usually forms solitary lesions, whenever they are present in gastrointestinal system and rarely displays a MLP pattern⁶. We discussed herein a case of primary intestinal DLBCL, and its rare presentation as multiple lymphomatous polyposis. It is emphasized that, although very rare, this entity must be ruled out in the differential diagnosis of patients presenting with the signs and symptoms of Crohn's disease as well as mantle cell lymphoma, prognosis and modes of treatment of which vary considerably from DLBCL.



Figure 1: Segment of ileum showing flattening of mucosa, with presence of multiple, small, sessile polyps

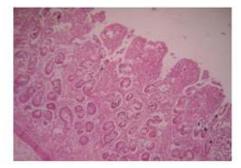


Figure 2(a): Histological section showing tumour cells infiltrating the layers of the intestinal segment. (100×, HandE stain)

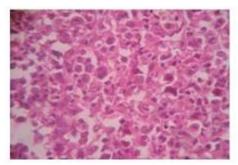


Figure 2(b): Histological section showing atypical lymphocytes with eosinophilic cytoplasm (400x, HandE stain)

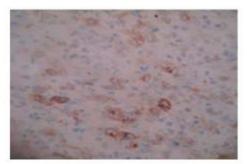


Figure 3 (a): CD 30



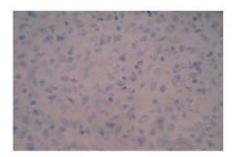


Figure 3(b): CD 79a

Figure 3(c): LCA

Figure 3(d): EMA

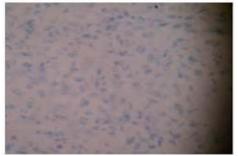


Figure 3(e): CD 10



Figure 3(f): Ki 67

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