Primary Lymphoma of Duodenum

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Introduction

Primary lymphoma of the gastrointestinal tract accounts for 5% of all gastrointestinal tract tumours and only 1% of all lymphomas. Diffuse, large cell lymphoma of B-cell origin (DLBC) is the predominant histologic type of primary extra-nodal lymphoma originating from a gastrointestinal site. Primary non-Hodgkin’s lymphoma of the duodenum is an even more rarely occurring primary tumor of the gastrointestinal (GI) tract which constitutes less than 12% of all non-Hodgkin’s lymphoma (NHL).1,2 Periampullary lymphoma or lymphomatous involvement of ampulla of Vater is even much more rare a phenomenon to be seen. It is difficult to differentiate periampullary lymphoma from epithelial carcinoma of these sites on basis of clinical judgement or on basis of radiological findings. Accurate histopathological diagnosis is required to chalk out the plan of what could be the optimal treatment strategy. The principal determinants of choice of therapy and prognosis of the patient depends upon cell type, grade, and presence of disseminated disease.3,4

Case 1

A 40 year old male patient presented to surgical unit II BBH with complaints of epigastric pain, and vomiting, containing food particles. The epigastric pain was first noticed by patient about 4 months back. Initially it was more like a feeling of heaviness after meals, later on he sometimes felt mild, dull pain with aching character, aggravated with meals, with no specific relieving factor. There was no associated history of difficulty in swallowing. The patient however had a preference of semisolid and liquid diet, which he tolerated without discomfort and vomiting.

For two months patient was also experiencing vomiting episodes, projectile in nature, watery, and non-bilious, containing semi-digested food particles. Vomiting followed about half an hour to one hour after meals, and sometimes the patient felt some relief in the pain. There was history of marked anorexia, weight loss and generalized weakness, as well as easy fatigability. He was put on supplemental parenteral nutrition, due to inadequate oral intake. Baseline investigations showed microcytic, hypochromic anemia. Rest of baseline labs, including CXR were normal. Upper GI endoscopy showed dilatation of proximal stomach, with food particles. Distal stomach was extensively involved with a large, polypoidal, and necrotic growth, appearance suggestive of NHL. Biopsy of growth was taken. However the histopathological report did not give a definitive diagnosis and showed necrotic, and inflamed gastric mucosa. CT Scan abdomen and pelvis with contrast showed a circumferential malignant growth involving the pyloric canal and proximal duodenum, which was collapsed and contrast didn’t go into small bowel. There was no involvement of the intra-abdominal nodes and no distal metastasis. A repeat ultrasound guided endoscopy showed hypertrophied mucosa, bulging into the lumen from pyloric antrum to the duodenal bulb. The conclusion was presence of a submucosal tumor, for which multiple biopsies were done. Unfortunately, biopsy report was again unfruitful, showing inflamed gastric mucosa only. So a decision was made to operate the patient with an aim of curative resection if possible, and for achieving a proper tissue sample for histopathology. Per-operative findings showed a large tissue growth extensively involving the distal stomach as well as first and second part of the duodenum. The ampulla of Vater however was not involved, and was patent. There were no celiac, or hepatic nodes, no hepatic metastasis and spleen was of normal size. Partial gastrectomy was done, along with a gastrojejunosotomy, and tube duodenostomy was done to control the duodenal stump. Post-operatively patient remained stable and had a swift recovery. Oral intake of fluids was allowed on 8th post-operative day, and patient was discharged on 10th post-operative day, when he resumed a good oral intake. The controlled duodenal fistula in form of tube duodenostomy ceased to produce output one week post op, and it was removed on 14th post-operative day. Histopathological examination showed Non-Hodgkin’s Lymphoma, Diffuse large B cell type infiltrating the muscularis propria of stomach and duodenum. Immune
histochemistry showed positive Leucocyte Common Antigen (LCA) in the tumor cells. CD 20 was positive and CD 3 was negative. Patient was referred for oncologist’s opinion and further management to NORI hospital Islamabad. They advised a BM biopsy, before starting chemo-radiotherapy. Bone marrow biopsy showed scanty tissue with no infiltrate, and grade 1 fibrosis. Patient was lost follow up after this as he had financial problems regarding the treatment offered by oncologist.

Patient re-appeared again, two months after surgery when after receiving first cycle of chemotherapy he developed jaundice. He was admitted to SU II BBH again. Workup showed that there was extensive growth in region of porta hepatis, compressing the common bile duct. Patient was provided total parental nutrition was given supportive therapy, with a plan of PTC to relieve the jaundice. But patient expired, after a long brave fight with the disease.

**Case 2**

A fifty years old male, presented through emergency room with complaints of epigastric pain for two months and vomiting after meals, containing food particles for last two weeks. He was dehydrated, and anorexic. He was resuscitated with IV fluids and workup was done as he stabilized. Baseline lab investigations were normal. X-ray chest was unremarkable. Upper gastrointestinal endoscopy showed a malignant looking structure in the second part of duodenum. Biopsy was done. USG abdomen pelvis was normal with no intra-abdominal lymphadenopathy, no hepatic metastasis, and normal spleen. Barium study showed an apple core lesion in the second part of duodenum. Biopsy was adequate for immunohistochemistry. Patient was informed about the disease, its natural history, associated risks and prognosis. He however chose to go to a spiritual healer for treatment and refused further medical care. Follow-up was lost on this patient after he went against medical advice for good.

**Discussion**

Non-Hodgkin lymphomas are a group of malignancies of the lymphoid system with many varieties which differ in prognosis. WHO classification of hematological and lymphoid tumors, broadly classifies these diseases as B-cell and T-cell neoplasms. B-cell lymphomas make up about 90% of all lymphomas. Follicular lymphoma and diffuse large B-cell lymphoma (DLBCL) are the most commonly occurring varieties. Approximately 55,000 to 60,000 new cases of non-Hodgkin lymphoma are diagnosed annually in the US, and the number has almost doubled during the past 3 decades. It’s a routine to use the Ann Arbor Staging Classification to classify the extent of disease, and the International Prognostic Index is used to define prognostic subgroups. Molecular and genetic prognostic markers, which may be used in the future to further refine and specify treatment strategies. Treatment of NHL basically depends upon the histology subtypes and extent of disease.

Follicular lymphoma (FL) is the commonest variety of NHL found in the nodes, and the least common in the gastrointestinal (GI) tract, where mucosa-associated lymphoid tissue lymphoma are the most frequently seen variety of lymphoma. According to literature, Diffuse large B-cell lymphoma (DLBCL) rarely involves the duodenum, and clinic-pathological characteristics of DLBCL involving the duodenum have not been well established. MRI features of duodenal lymphoma include marked symmetric or asymmetric concentric wall thickening, thickening of mucosal folds, proximally aneusymal small-bowel loop dilation may be seen, and luminal strictures may be present. Para-enteric mesenteric fat stranding may also be seen.

For patients with diffuse large B-cell lymphoma, treatment of Stage I and II disease involves doxorubicin-based chemotherapy (CHOP) combined with Rituximab; later on followed by radiation therapy to the involved group of lymph nodes. Those with stage III and IV disease are treated with rituximab combined with chemotherapy alone. Rituximab is an anti-CD20 monoclonal antibody that has clinically proven its efficacy in treating patients with various lymphoid malignancies, including the most aggressive forms of B-cell non-Hodgkin's lymphoma (NHL) and B-cell chronic lymphocytic leukaemia (CLL).

Relapse of disease is a major problem. Patients who initially present with these diseases often know they have a malignancy considered curable in many cases, and diagnosis of relapse can be emotionally devastating. Many patients undergo major depression. For such patients one approach is to give high-dose therapy with stem cell support. This works better for chemo-sensitive varieties of lymphomas. While the varieties who were with chemo-resistant from the beginning, subsequently should be treated with novel experimental therapies in case of relapses.
relapse of aggressive lymphoma, like DLCL is difficult in terms of cost-effectiveness as well as psychosocially. Investigators have been trying to devise certain disease-related features which can determine whether the patients suffering from lymphomas have a high or low risk of relapse, and we can now use combinations of these features to determine which patients may be safely given the classical CHOP regimen and which patients will need more intensive chemotherapy.12

Role of stem cell transplant is now gaining popularity as a treatment option in treating aggressive tumors for difficult and relapsing cases. High-dose chemotherapy with stem cell rescue is particularly effective as salvage treatment for diffuse large cell lymphoma.11 Stem cell transplant (SCT) protocols routinely require excellent response with initial chemotherapy as a requirement for entry into clinical trials. Other limitations for SCT protocols include age greater than 60 or 65 years, disabling COPD, end stage renal disease, or poor cardiac function, and CNS or bone marrow involvement.12

References