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Case Report – "Common Symptom of Rare Disease"

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Abstract
The Primary Gastrointestinal Lymphoma (PGL) is a relatively rare malignancy and easily misdiagnosed due to its variable clinical symptoms of the digestive tract. Despite their rarity, primary lymphomas of the GI tract are important since their evaluation, diagnosis, management, and prognosis are distinct from that of lymphoma at other sites and other cancers of the GI tract. In this report, we describe a case of middle aged male patient presented to us with Diarrhoea, Vomiting and Abdomen pain with no significant past co-morbidities, with no history of smoking or alcohol consumption. Clinical examination, investigations and further workup of patient revealed small bowel lymphangectasia in ileal biopsy and primary intestinal lymphoma.
Keywords: *Diarrhoea, Ileal lymphangectasia, Intestinal lymphoma.*

Introduction

Primary gastrointestinal lymphoma are rare, accounting for only 1 to 4 percent of all malignancies.[1] It can arise from the stomach, small intestine or colon. The GI tract is the predominant site of extra nodal non-Hodgkin lymphomas (NHLs).[2]

Of all GI lymphomas, 75% are located in stomach, followed by small bowel and colon. They have peak incidence in the 7th decade of life, with male to female ratio of 1.5:1. Primary intestinal lymphomas can be differentiated from secondary intestinal lymphomas by the absence of superficial and mediastinal lymphadenopathies on work up, and there is no evidence of disease on both, peripheral blood smears and bone marrow biopsies. Lymphoma may well be related to secondary intestinal lymphangiectasia by means of lymphatic channel blockade and can precede the onset of lymphoma by several decades.[3]

Case History

57 year old gentleman, from Bangalore presented to us with chief complaints of chronic diarrhoea since 3 months. 6 – 8 episodes per day, watery in consistency, non- mucus and non-blood stained since 3 months, decreased appetite since 3 months and abdomen pain since 20 days of colicky type, periumbilical region and not subsided by passing stools. vomiting since 3days. 2-3episodes per day,

non bilious, non blood stained associated with nausea. There was no history of fever. No history of bleeding per rectum. He was not a known diabetic or thyroid disorder. No h/o weight loss, No history of high risk behaviour. No history of co-morbidities. No history of previous hospitalisation.

On examination, patient was conscious and oriented. Pulse rate 110b/m, Blood pressure 130/80mmHg, Respiratory rate 15breaths/minute, Temperature - 37.8 degree Celsius. No dehydration. Bilateral pitting pedal edema was present. No pallor, icterus, clubbing and lymphadenopathy.

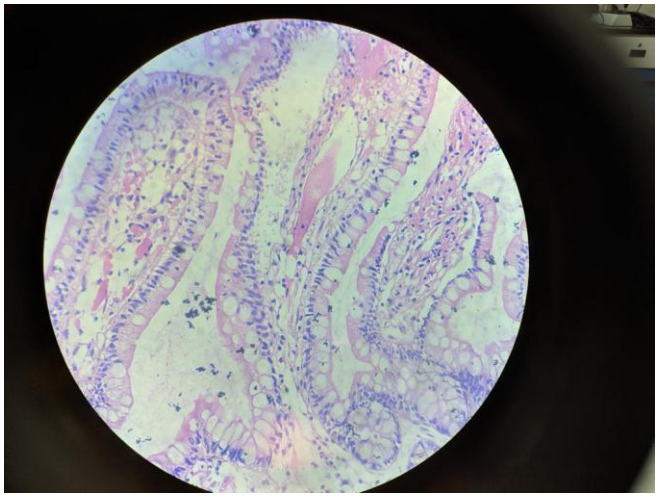
On per abdomen examination, abdomen was distended and soft. Spleen was enlarged measuring 14cm below left costal margins, firm in consistency, smooth surface, sharp margins, non-tender and non-pulsatile. Splenic notch was felt. There was no other organomegaly. No free fluid. Bowel sounds heard and rest of the systemic examination was normal.

Laboratory investigations revealed hemoglobin - 13.4 mg/dl, Total Leucocyte count – 4,900/mm3, Platelet count - 96,000/mm3. Differntial count of neutrophils 64%, lymphocytes - 26%, Eosinophils - 2%, Monocytes - 8%, Basophils - nil. Peripheral smear - Normocytic Normochromic blood picture with leucopenia and thrombocytopenia. Random blood sugar - 116mg/dl, Blood urea - 107mg/dl, Serum creatnine - 0.9mg/dl. Serum sodium-135mg/dl, Serum potassium - 4.9mg/dl, Serum Chloride - 114mg/dl. Total bilirubin - 1.8mg/dl, indirect bilirubin - 1.1 mg/dl, total protein - 6.9mg/dl, serum albumin - 2.9mg/dl, serum globulin - 4.1mg/dl, A/G ratio 0.7, AST - 45U/L, ALT - 32U/L, ALP - 28U/L. HIV I, II - negative, Serum HbsAg - negative, HCV antibodies - negative. Stool analysis for ova, cyst - negative, stool AFB stain - negative. Serum LDH – 253 U/L. Chest x-ray PA view – normal study.

USG abdomen and pelvis revealed - Massive splenomegaly with thickened small bowel loop and mild ascites. Thyroid function test: TSH - 2.35 uIU/ml, serum Free T3 - 1.89 pg/ml, serum Free T4 - 1.33 ng/dl. Serum calcium - 8.8mg/dl.

In view of chronic diarrhoea, we investigated patient further. Colonoscopy revealed no significant lesion, ileal biopsy was sent. Histopathology slide section showed structure of intestine with villi lined by goblet cells. Lamina propria, submucosa and occasional villi contain few dilated lymphatic channels lined by flattened endothelial cells, few containing eosinophilic secretions. Lamina propria is oedematous, infiltrated with few chronic inflammatory cells of lymphocytes and plasma cells. Features were consistent with lymphangectasia. (Figure A)

Figure A



Our Patient underwent CECT abdomen and thorax.

Impression was circumferential symmetrical wall thickening involving duodenum, jejunum and proximal ileum. Mild hepatomegaly. Massive splenomegaly, dilated portal and splenic vein with perisplenic collaterals. Minimal ascites and mild bilateral pleural effusion. Para aortic group of lymphnodes not enlarged. No mediastianal Lymphadenopathy, Lungs normal. Features suggestive of Primary gastro- intestinal Lymphoma.

Further, Bone marrow aspiration and biopsy was performed. Report revealed Hypercellular marrow showing myeloid and megakaryocytic hyperplasia with decreased erythropoiesis.

Discussion

Primary malignant tumours of the small intestine are very rare. Ileum is the most common site 60-65% followed by jejunum 20-25% and duodenum 6-8%.[4] the age of presentation varies with histological subtypes of lymphoma. Patients can present around 5th decade of life. Males are more commonly affected than female. The clinical presentation can be nonspecific and the patients have symptoms, such as colicky abdominal pain, nausea, vomiting, diarrhoea, weight loss and rarely acute obstructive symptom.[5]

Incorporation of FDG-PET has a significant advantage in staging with a sensitivity of 80% and a specificity of 90%.

Unfortuanately, we lost the patient during follow up period of investigations.

Before, current multimodality therapy was established, it was considered as high-grade lymphoma having a negative prognostic factor for survival, associated with lower complete remission rate and a shorter survival rate.[6]

The treatment of Primary Intestinal Lymphoma has shifted away from surgery and toward chemotherapy regimens. Surgery is now limited to cases of perforation, haemorrhage, or obstruction due to the tumour, and it is no longer the cornerstone of treatment with its mortality rate reaching up to 8%.[7]

The treatment of Primary intestinal lymphoma consists of anthracycline-based chemo immunotherapy. Chemo immunotherapy with 3 to 4 cycles of standard R-CHOP followed by "involved-field" radiotherapy could be considered as a gold standard option for localized stages (stages I and II in the Lugano classification). Advanced-stage patients (Ann Arbor stage III/IV) usually undergo 6 to 8 cycles of R-CHOP in order to obtain a complete remission rate.R-CHOP regimen consists of Rituximab (R) cyclophosphamide, doxorubicin hydrochloride (hydroxydaunorubicin), vincristine sulfate (Oncovin), and prednisone every 14 to 21 days.

Over all 5 year survival rate is between 50 – 70 % with multimodality treatment.[8]

Conclusion

Gastrointestinal lymphomas are usually not clinically specific and indistinguishable from other benign and malignant conditions. Our patient didn't have classical B symptoms or lymphadenopathy as described in various types of lymphomas. So, the clincians should have high index of suspicion of diagnosis. With the better insight into the rare diseases, patient can be diagnosed as early as possible and can improve the survival rate of the patient without delay in intiation of treatment.

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