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A Rare Case Report of Duodenal Marginal Zone B-cell Lymphoma Related to Immunoproliferative Small Intestinal Disease and Associated Lymphoma (IPSID)

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Authors' contributions

This work was carried out in collaboration between all authors. Authors ZA, MN, MWHA and AG designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors MA, BO and MMS managed the analyses of the study. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Marginal zone B-cell lymphoma is a unique lymphoma of the gastrointestinal tract most commonly in developing countries with poor socioeconomic conditions, poor hygiene, malnutrition, and a high degree of intestinal infections where IPSID is common.
We experienced a rare case of extranodal MZBCL in duodenum (sub type of MALT) in a 14-year-old girl who complained of epigastric pain.
Abdominal Computed Tomography (CT) with Intravenous (IV) contrast showed mixed mass infiltrate the intestinal wall with mesenteric lymphadenopathy.

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Upper gastrointestinal tract endoscopy had showed obstruction in the 2nd segment of duodenum, biopsies were taken.
Pathology report according to findings refers to immunoproliferative small intestinal disease and associated lymphoma (IPSID) and by immunostains, cells features are compatible with extranodal marginal zone B cell lymphoma which is sub type of MALT lymphoma.

Keywords: Duodenum; IPSID; marginal zone B-cell lymphoma; gastrointestinal tract lymphoma.

1. CASE REPORT

A 14-year-old Caucasian female presented to gastroenterology clinic complaining of nausea, vomiting, weight loss and abdominal pain from 5 months ago. In the last 3 weeks she developed fever and chills. She didn't have any constipation or diarrhea.

Her history was unremarkable except her two sisters were died one of them was diagnosed with brain mass and the other one with abdominal mass.

On examination, the patient was febrile and had moderate pallor. The abdomen was nondistended and no surgical scars were found but, by palpation there was mild tenderness and a firm nonmobile epigastric mass was detected.

Initial evaluation revealed a: Hemoglobin (Hb) 6.6 g/dl, Albumin 2.8 mg/dl, Total Protein (TPr) 4.1 mg/dl, WBC 19,65 10^3/mm^3, HCT 24,65%, RBC 4,6 10^6/mm^3, MCV 75,3 um^3, MCH 22,37 pg, PLT 137 10^3/mm^3, C-RP 69,1 fhmg/l, serum antibodies to tissue transglutaminase was negative and all other laboratory tests were within normal limits.

Abdominal ultrasonography showed normal abdominal organs except intestinal mass.

Abdominal Computed Tomography (CT) with Intravenous (IV) contrast demonstrated mixed mass infiltrate the intestinal wall with mesenteric lymphadenopathy and without vascular infiltration (Fig. 1).

Upper gastrointestinal tract endoscopy revealed intrinsic obstruction in 2nd segment of duodenum as infiltrative lesion. Biopsies were taken.

Pathology report revealed normal duodenal mucosa with a moderate degree of villous shortening and broadening (MARSH 3b), associated with increased intra-epithelial lymphocytes, with moderate chronic lymphoplasmacytic inflammatory infiltrate of lamina propria and normal muscularis mucosa and brunner glands, this findings refer to immunoproliferative small intestinal disease and associated lymphoma (IPSID), which is a subtype of small intestinal MALT lymphoma (Fig. 2).

Immunophenotypically the cells were positive for CD20 (Fig. 3), CD79a, BCL-2 and Ki 67 labeling index was 5%-10%. But, CD3 (Fig. 4), BCL-6 and CD5 were negative.

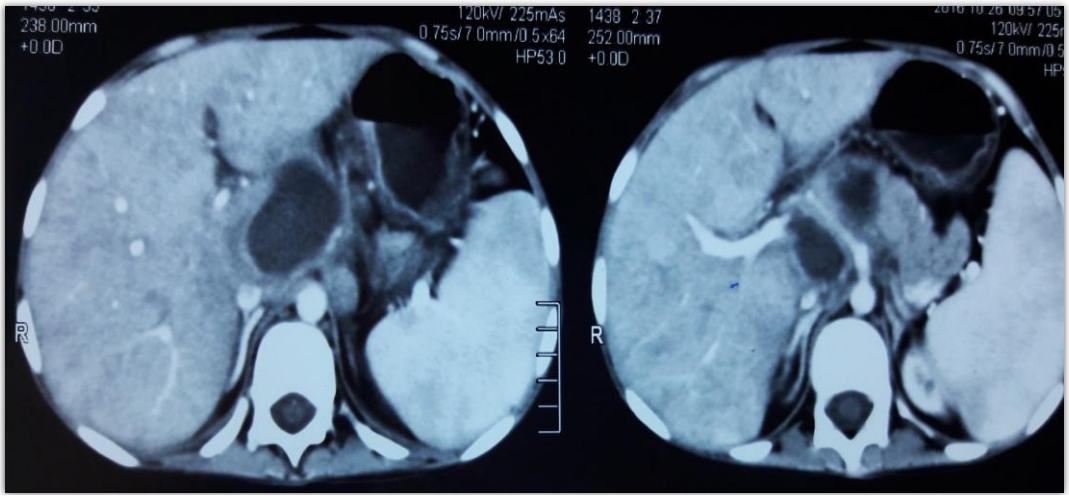

Fig. 1. (Axial plane): Abdominal (CT) with IV contrast revealed a mixed mass with mesenteric lymphadenopathy

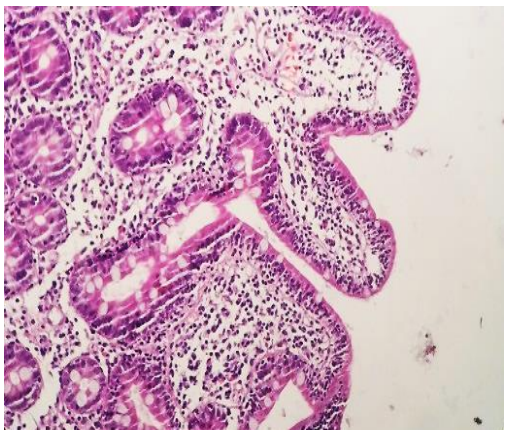

Fig. 2. (H&E), Duodenum, villous shortening, increased intra-epithelial lymphocytes, moderate chronic lymphoplasmacytic inflammatory infiltrate of lamina propria

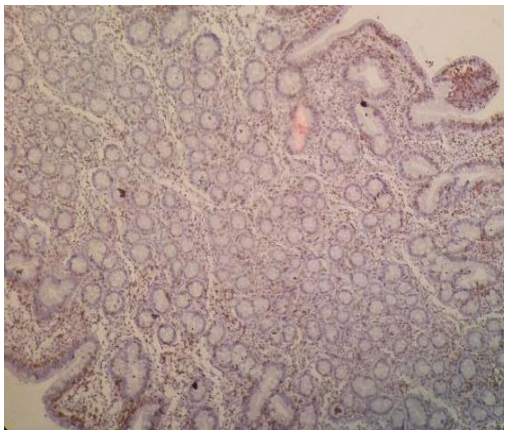

Fig. 3. (CD20), Duodenum, lymphocytic infiltration to the epithelial cells

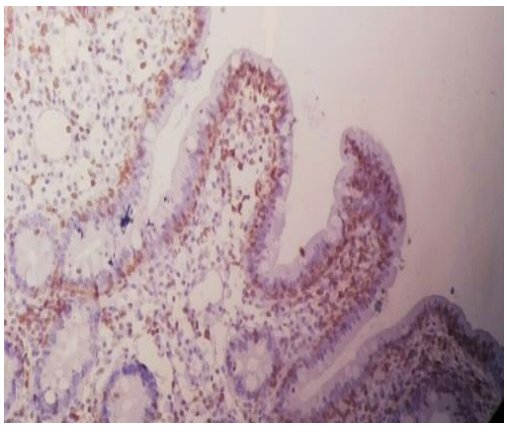

Fig. 4. (CD3), Duodenum, lymphocytic infiltration to the epithelial cells

Over all these features are compatible with extranodal marginal zone B cell lymphoma of MALT lymphoma.

Patient received chemotherapy (R-CHOP) and developed ascites. Ascites fluid analysis revealed a: SAAG 1,8 g/dl, Glucose 181 mg/dl, T. Protein 52 g/dl, Albumin 1 g/dl, LDH 116, RBC 50 cell/ul and WBC 900 cell/ul (90% lymphocyte).

After 3 days she developed sever breathlessness and was admitted to ICU and died with pulmonary edema after 12 hours.

2. DISCUSSION

Extranodal marginal zone lymphomas occur outside lymph nodes (eg, in the gastrointestinal tract, thyroid, orbit, leptomeninges, spinal cord, or skin) [1].

MALT (mucosa-associated lymphoid tissue) lymphoma (MALToma) is the term traditionally used for extranodal marginal zone lymphoma of MALT [1,2,3].

Extranodal marginal zone lymphomas commonly follow immune system dysregulation from sustained stimulation with chronic infections or autoimmune disorders [1,2,3].

Susceptibility is thus influenced by both genetic and environmental factors. Marginal zone lymphoma of MALT (MALT lymphoma) is the most common indolent subtype and represents 7% of all non-Hodgkin lymphomas [1,2,3].

The stomach is the most common extranodal site (incidence rate=3.8), followed by eye\adnexa (IR=1.4), lung, skin, and salivary glands (IR=0.9-1) [1,2,3].

Geographic variation of the gastrointestinal lymphomas is marked. For example, in the Middle East and South Africa, the incidence of intestinal B-cell lymphoma is relatively more common (in the range of ≈35%) because of the high incidence of IPSID. In the United States and Europe, celiac disease is a common cause of T-cell lymphoma [4,5,6,7,8].

IPSID (also known as Mediterranean lymphoma) is considered to be a subtype of extranodal marginal zone B- cell lymphoma, it is a primary intestinal disease of teenagers or young adults and is found most commonly in developing countries with poor socioeconomic conditions, poor hygiene, malnutrition, and a high degree of intestinal infections, although the World Health Organization recognizes it as a separate entity [4,5,6,7,8].

Patients often have a short history of diarrhea, malabsorption, weight loss and chronic abdominal pain [4,5,6] as our patient suvered.

By investigations we detect duodenal mass and take biopsies.

IPSID by biopsies of the bowel and regional lymph nodes is characterized pathologically by a heavy lymphoplasmacytic infiltration, this lymphoplasmacytic infiltration is originally reactive in nature and that it represents a response to a continuous antigenic stimulus of possible infectious nature [9,10].

In our case, the histological findings and Immunophenotyp reveled extranodal MZBCL.

We start treatment with chemotherapy (R-CHOP) and then she develop complication as ascites.

After 3 days she developed sever breathlessness and was admitted to ICU and died with pulmonary edema after 12 hours.

3. CONCLUSION

We have presented a case of extranodal MZBCL involving the duodenum which is rare as a location of extranodal MZBCL that present by nonspecific upper gastrointestinal complaints such as abdominal pain, nausea and vomiting related to IPSID.

This case will add new point to medical literature as a new location to MZBCL and poor prognosis.

CONSENT

Written informed consent was obtained from the parents for publication of this case report.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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