JOURNAL OF CASE REPORTS 2015;5(2):393-396

Non Hodgkin Duodenal Lymphoma in a 5 year old girl

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Abstract:

A 5 year old female child presented to pediatric outpatient department with melena and non-specific abdominal pain. She underwent an upper endoscopy which showed a confluent polypoid ulcerative lesion in the duodenum. Histopathology was consistent with non-Hodgkin's lymphoma. This is a rare neoplasm of duodenum and must be differentiated from other type of malignancies.

Key words: Duodenum, Gastroscopy, Lymphoma, Melena, Non Hodgkins Lymphoma.

Introduction

Primary lymphomas of the digestive tract are uncommon and constitute a heterogeneous group of neoplasms that occur primarily in the stomach. Primary gastric lymphoma constitutes 1.48% of all gastric cancers in children [1,2]. Because of the paucity of lymphoid tissue in the duodenum, primary duodenal involvement of the lymphoma is a rare condition. It accounts for less than 5% of all small bowel lymphomas [3]. Periampullary lymphoma of ampulla of Vater is even rarer [4].

We report the case of a 5-year-old female presenting with melena and abdominal pain with a diagnosis of duodenal non-Hodgkin's lymphoma. This study focuses on the significant possibility of non-Hodgkin in children with duodenal ulcerative mass.

Case Report

A five-year-old girl presented in the pediatric emergency service at Mother Teresa University Hospital Centre with presenting complaints of melena. Melena was associated with abdominal pain and vomiting. She has no history of other diseases or medicaments administration. She was the first child of an Albanian couple without consanguinity.

During physical examination she looked pale but was active, alert and afebrile. Vitals showed heart rate of 88/min; respiratory rate was 22/min; blood pressure of 96/62 mm Hg and oxygen saturation of 97%. Her weight was 18 kg and height 106 cm. Heart sounds were normal without murmurs. Lung auscultation was uneventful. Abdomen was tender, liver and spleen

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Received: August 12, 2015 | **Accepted:** August 27, 2015 | **Published Online:** September 15, 2015

Conflict of interest: None declared | **Source of funding:** Nil | **DOI:** http://dx.doi.org/10.17659/01.2015.0101

size were normal. Extremities were normal without edema.

Investigations showed hemoglobin of 7.4 g/dL. Biochemical examinations revealed normal level of glucose, electrolytes, liver and kidney function tests. Inflammatory markers such as PCR and VES were 26 mg/L and 60 mm/h respectively. LDH was high 634 U/L. Urine analysis was normal. IgA anti-transglutaminase was in the normal range. Stool tests for *Helicobacter pylori* antigen and parasites was negative. Chest X ray was normal and hepatitis markers were negative.

We decided to perform an upper endoscopy. During examination we detected a polypoid ulcerous, hemorrhagic, irregular-shaped 2 cm mass, in the duodenum which started from ¼ of apex of duodenal bulb and stretching to near the Ampulla of Vater [Fig.1]. On CT scan, there was a 4 cm well-defined, markedly enhancing soft tissue mass involving the duodenum (see the white arrow), with moderate dilatation of bile duct [Fig.2].

A biopsy was performed and the HE stain revealed presence of diffuse growth pattern with diffuse infiltrate of medium-sized, slightly pleomorphic cells. We performed immunohistochemical (IHC) stains only to make a differential diagnosis between a lymphoid tumor and a neuroendocrine tumor. The final diagnosis was non-Hodgkin's lymphoma [Fig.3].

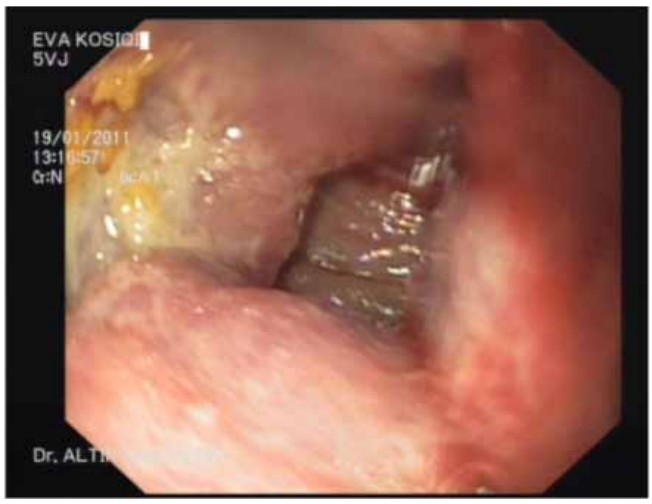

Fig.1: Endoscopic image of polypoidal mass and ulcer in duodenum.

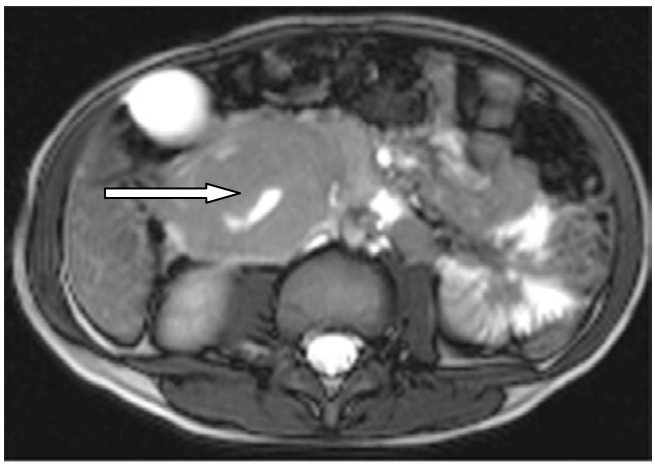

Fig.2: CT image show duodenal mass and slight dilatation of bile duct.

Discussion

Although lymphomas are considered the third most common tumor in children, primary non-Hodgkin lymphoma of the duodenum (PLD) is an uncommon primary tumor of the gastrointestinal (GI) tract. It represents only 5% to 16% of small intestinal lymphomas [5,6]. Periampullary lymphoma or lymphomatous involvement of ampulla of Vater is even rarer. Only a few reports in pediatric patients are available. PLD is rare before 2 years and has a peak frequency to 7 years. It is more common among boys than among girls (3/1) [7,8]. Diffuse, large cell lymphoma of B-cell origin (Burkitt's lymphoma) is currently recognized as representing the predominant histological type. Certain risk factors have been implicated in the pathogenesis of gastrointestinal lymphoma including *Helicobacter pylori (H. pylori)* infection, human immunodeficiency

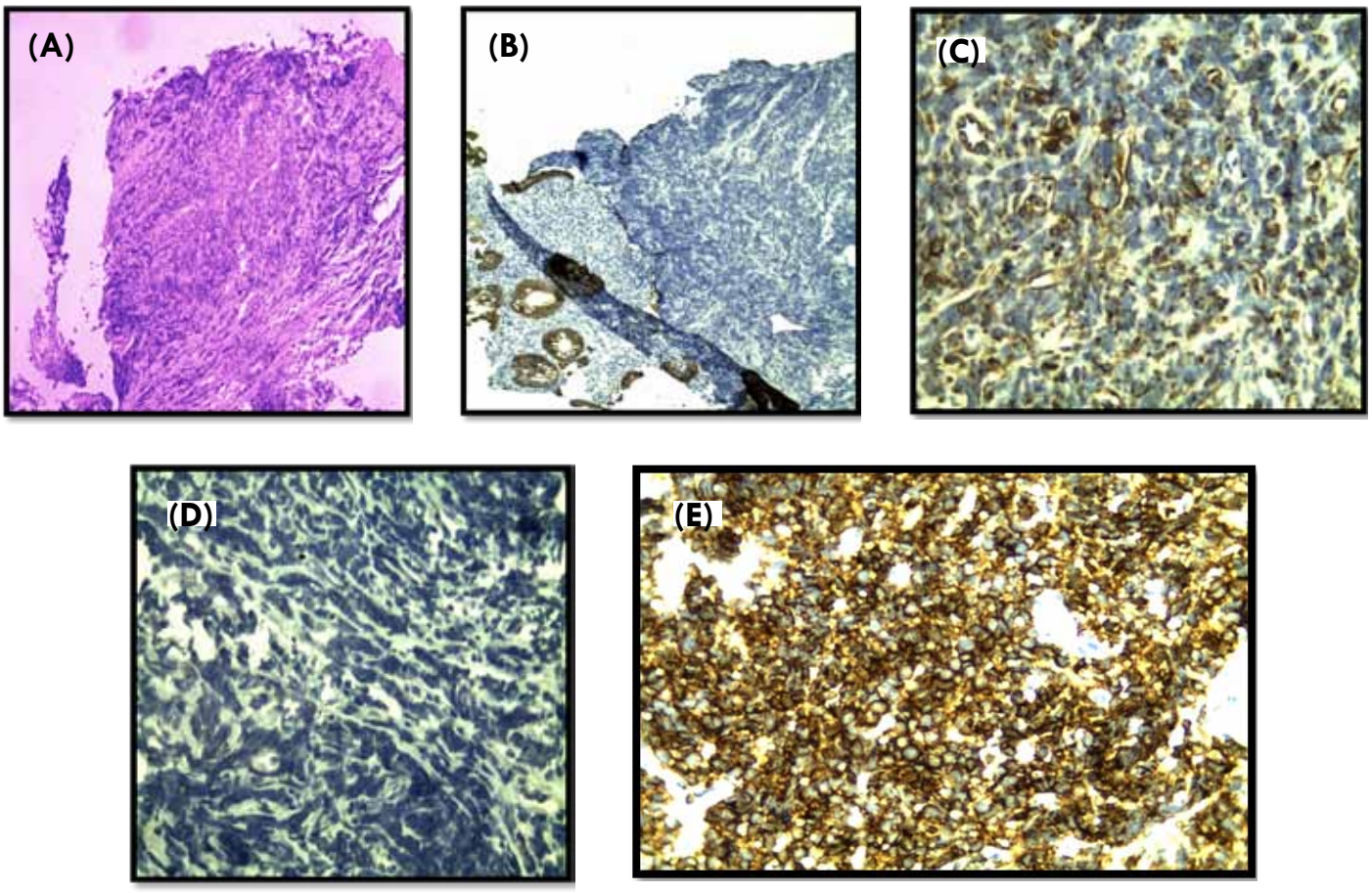

Fig.3: HE and IHC Stains: (A) HE, (B) Vimentin, (C) CK, (D) Chromogranin, (E) CD45.

virus (HIV), celiac disease, *Campylobacter jejuni (C. jejuni)*, Epstein-Barr virus (EBV), hepatitis B virus (HBV), human T-cell lymphotropic virus-1 (HTLV-1), inflammatory bowel disease and immunosuppression [9,10]. The clinical presentation of small intestinal lymphoma is non-specific and the patients have symptoms, such abdominal pain, nausea, vomiting, weight loss and rarely acute obstructive symptoms, intussusceptions, perforation or diarrhea. The macroscopic appearance of small intestine lymphoma is a mass, polyp and ulcer on endoscopy, which cannot be distinguished from other lesions. Microscopic examination reveals a diffuse growth pattern with large cells (usually 5 x normal lymphocytes) resembling immunoblasts (amphophilic cytoplasm, eccentric nuclei with one central nucleoli) or centroblasts (pale or basophilic cytoplasm, vesicular chromatin due to chromatin margination, 2-3 nucleoli, often near membrane), or rarely anaplastic, associated with neutrophils. An important aspect to be considered is the increasing sensitivity and specificity of imaging techniques like EUS and PET-CT in the diagnosis of lymphomas. Unlike adults, child treatment is based on chemotherapy alone in all cases. Global survival is 75% at 5 years, all stages combined. Surveillance is effected by abdominal ultrasonography. Tumor regression may be spectacular in few days. In our case, presentation of disease was mimicking a peptic ulcer because of melena and abdominal pain. The combination of upper digestive endoscopy, CT images and histopathological view was the cornerstone of this

rare neoplasm in children. This child underwent to chemotherapy at oncologic service and the mass was reduced successfully during the treatment.

Conclusion

Pediatric duodenal involvement of the lymphoma is a rare condition. Periampullary lymphoma or lymphomatous involvement of ampulla of Vater is even rarer. Since, periampullary lymphoma is not easy to differentiate from others tumors of these sites clinically and radiologically, endoscopic duodenal biopsy and accurate histopathological diagnosis is essential diagnostic tool to plan optimal treatment strategy. The clinical outcome may be excellent if diagnosis is made earlier, even when presenting with rare clinical manifestations.

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