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Pediatric Primary Duodenal Non Hodgkin Lymphoma: Case Report

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Abstract

We would like to present a case of a 5-year old female who was hospitalized for melena in our pediatric ward. She underwent an upper endoscopy which showed a confluent polypoid ulcerative lesion in the duodenum. Histological view showed a non-Hodgkin's lymphoma.

This is a rare neoplasm of duodenum and must be differentiated from other type of malignancies.

Keywords: Duodenal polypoid lesion; Primary non-Hodgkin's lymphoma of the digestive tract; Primary duodenal 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 discontinuous 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 Presentation

A five-year-old girl was presented in the pediatric emergency service at Mother Teresa University Hospital Centre, with digestive hemorrhage, melena. The other complaints were 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 is active and in good humor. She had no fever. Heart sounds were normal without murmurs. Lung auscultation was uneventful. Abdomen was palpable, we have not seen hepatosplenomegaly. Extremities were normal without edema.

Arterial pressure=96/63 mm/Hg; Heart rate=88/min; respiratory rate=22/min; sat O2=97% Weight=18 kg; height=106 cm

Laboratory studies showed:

Blood count: WBC-16400/mm^3; RBC-2800000/ mm^3; Hgb-7,4 g/dl;PLT-331000/ mm^3

Biochemical examinations:Glicemia-64 mg/dl ;Urea-14 mg/dl ;Creatinine-0,69 mg/dl ;ALT-35 U/L ;AST-143 U/L Total Bilirubin-0,5;Total proteine -7.4 g/dl ; Albumine=3.3; PT=70% ; Na=135;

K=3,8; Cl=100;PCR=26 mg/L;VES=60 mm/h; Urine : normal; LDH=634 U/l ;

IgA antitransglutaminase was normal. Stool tests for H pilory antigen and parasitology were also normal; Chest X ray=normal; Markers of hepatitis 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 (**Figure 1**). On CT scan, there was a 4 cm well-defined, markedly enhancing soft tissue mass involving the duodenum, with moderate dilatation of bile duct (**Figure 2**).

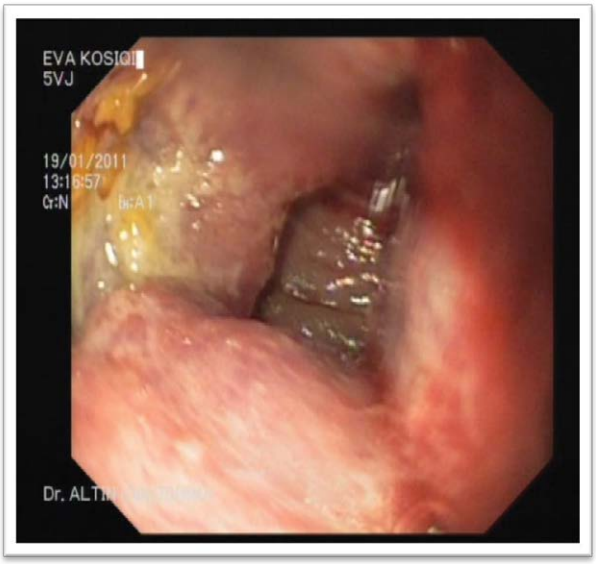

Figure 1 Polypoid ulcerous in duodenal bulb and stretching to near the Ampulla of Vater.

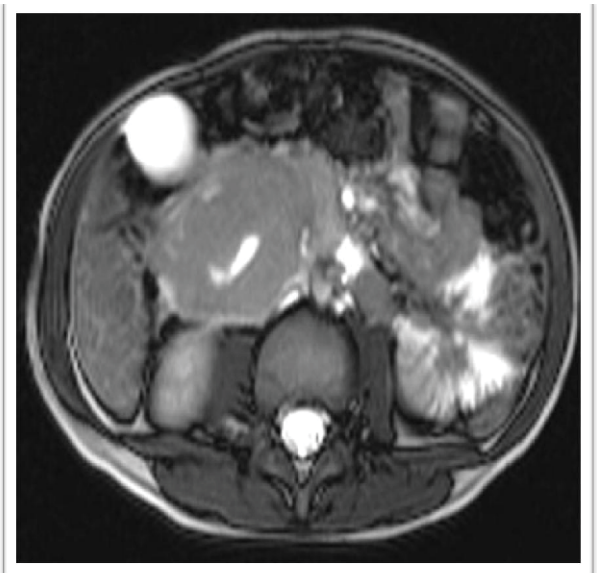

Figure 2 soft tissue mass involving the duodenum, with moderate dilatation of bile duct in CT scan.

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 with cellular pleomorphism and heterogeneity intermediate between Burkitt lymphoma and diffuse large B cell lymphoma (**Figure 3**).

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

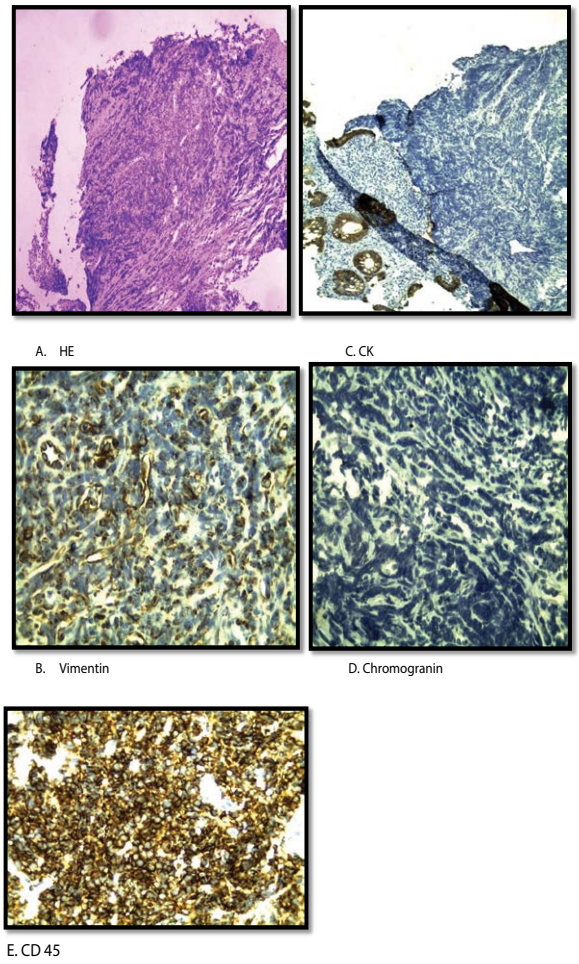

Figure 3 Stains for diagnosis between a lymphoid tumour and a neuroendocrine tumour. (HE and IHC Stains: A. HE, B. Vimentin, C. CK, D. Chromogranin, and E. CD45).

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 (Burkit'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 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 diffuse growth pattern lymphoid cells with pleomorphism. 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. EUS has gained momentum as an integral tool in the diagnosis, locoregional staging, and monitoring response of gastrointestinal lymphoma to treatment. The value of EUS and CT, however, is a matter of debate in the follow-up of patients as it is well established that histological remission precedes the normalization of wall changes in patients with lymphoma [11,12], thus precluding the necessity for endoscopic biopsy follow-up. Unlike adults, child treatment is based on chemotherapy alone in all cases. Global survival is 75% at 5 years, all stages combined. Surveillance is affected by abdominal ultrasonography. Tumor regression may be spectacular in few days.

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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