Journal of Cancer Clinics

Research Article
Published: 22 Jun, 2018

Obstructive Jaundice: A Rare Presentation of Non-Hodgkin Lymphoma in a Child

Apostolos Pourtsidis[1], Smaragda Papachristidou[1], Marina Servitzoglou[1], Dimitrios Doganis[1], Konstantina Kapetaniou[1], Maria Nikita[1], Evgenia Magkou[1], Dimitrios Kipraios[2], Astero Malama[3], Panagiota Giamerelou[4] and Margarita Baka[1]*

[1]Department of Oncology, P & A Kyriakou Children's Hospital, Greece

[2]Department of Gastroenterology, Agios Savvas Hospital, Greece

[3]Department of Radiology, Aghia Sophia Children's Hospital, Greece

[4]Department of Pathology, P & A Kyriakou Children's Hospital, Greece

Abstract

Obstructive jaundice is an unusual manifestation of a Non-Hodgkin lymphoma in a child. Although Burkitt lymphoma is the most common histological type of Non-Hodgkin lymphomas of the abdomen, diffuse Large B-cell Lymphoma can occur as well. We report a case of a 14-year old girl who presented with obstructive jaundice due to non-Hodgkin Lymphoma encasing the common bile duct. Imaging studies aided to diagnosis. Histological examination of the specimen, obtained by laparoscopic surgery, revealed diffuse Large B-cell Lymphoma. Patient received chemotherapy according to FAB LMB 96 protocol. Bilirubin rates normalized after the initial course of chemotherapy. She concluded her treatment and remains free of disease. In conclusion, obstructive jaundice is a rare type of presentation of diffuse Large B-cell Lymphomas but with high clinical suspicion, proper imaging studies and a proper biopsy, diagnosis can be obtained.

Introduction

Non-Hodgkin Lymphomas (NHL) represent the fourth most common childhood cancer in Europe [1]. However, Diffuse Large B-Cell Lymphoma (DLBCL) is a rare type of non-Hodgkin lymphoma in children. Although the abdomen is the most common primary site of NHL, involvement of the stomach, pancreas or common bile duct is not common [2]. The initial presentation of Non-Hodgkin Lymphoma (NHL) varies among cases. Even though jaundice is a frequent manifestation of NHL, it rarely occurs as a presenting symptom [3,4]. Moreover, DLBCL most commonly presents in the mediastinum, making an abdominal primary site extremely unusual. We present a case of non-Hodgkin lymphoma, most specifically DLBCL, with obstructive jaundice as the only initial symptom.

Materials and Methods

A 14-year old girl was admitted to our hospital with jaundice. She presented with paler feces and darker urine, which had initiated ten days prior to her admission. She did not experience any abdominal pain. The physical examination revealed no remarkable signs, apart from jaundice. Her complete blood count test was within normal range (WBC: 8900/μl, Neut: 68.6%, Lymph: 21.5%; Hb: 13.0g/dl; Hct: 38.6% and PLT: 246000/μl). Blood chemistry revealed mildly elevated liver enzymes (SGOT: 43 U/l-SGPT: 108 U/l), significantly elevated γ-GT (234 U/l) and conjugated hyperbilirubinemia (total bilirubin: 10.3 mg/dl - conjugated bilirubin: 9.2 mg/dl). ESR, LDH and uric acid were within normal range.

Abdominal ultrasound revealed dilatation of the Common Bile Duct (CBD) of a spindle form, as seen in a cyst. In order to plan the surgical procedure, a Magnetic Resonance Cholangiopancreatography (MRCP) was performed. The distal part of the common bile duct was dilated (diameter= 5mm) although it was not visible in its proximal part. The consequent Magnetic Resonance Imaging (MRI) scan revealed indistinct (vague) abnormal tissue, 35 × 28 × 19 mm, between the stomach and the head of pancreas, infiltrating the head and body of pancreas. Furthermore, the surrounding fat the tissue was also vague. This tissue encased the common bile duct in its proximal part. Another abnormal tissue was described at the right adrenal gland, making it vaguely seen. Both of these tissues were mildly enhanced with intravenous contrast and seemed to

OPEN ACCESS

Correspondence:
Apostolos Pourtsidis, Department of Oncology, P & A Kyriakou Children's Hospital, Levadias 8, PC: 11527, Athens, Greece, Tel: 30 213200924; E-mail: tolispou@gmail.com

Received Date: *28 Feb 2018*
Accepted Date: *26 Apr 2018*
Published Date: *22 Jun 2018*

Citation:
Pourtsidis A, Papachristidou S, Servitzoglou M, Doganis D, Kapetaniou K, Nikita M, et al. Obstructive Jaundice: A Rare Presentation of Non-Hodgkin Lymphoma in a Child. J Cancer Clin. 2018; 1(1): 1004.

Copyright *© 2018 Apostolos Pourtsidis. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.*

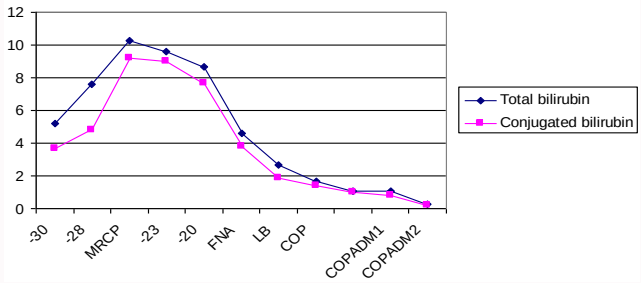

Figure 1: Course of bilirubin values during the diagnostic procedures and initiation of chemotherapy.

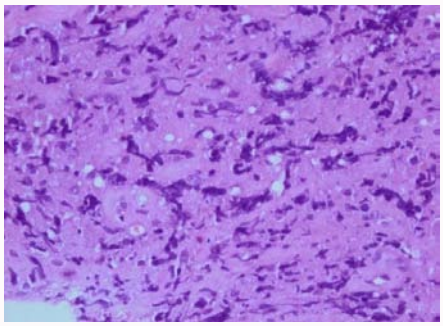

Figure 2: Hematoxylin eosin staining (×400).

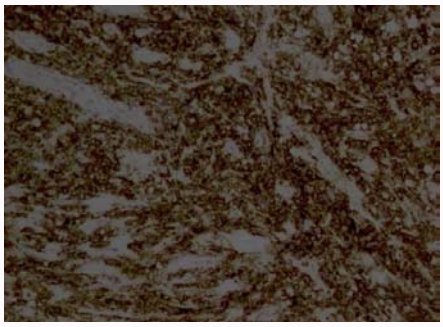

Figure 3: CD19 (x200).

originate from the pancreas.

A repeat ultrasound was similar, but indicated that, in fact, there was only one large abnormal tissue extending among stomach, pancreas and right kidney, and probably infiltrating the right adrenal gland and pancreas. However, this mass did have neither specific characteristics nor the form of a tumor. An abdominal Computer Tomography (CT) scan with bowel contrast agent was performed, which showed thickening of the pyloric antrum wall, which was encased by the abnormal tissue that spread to the previously described areas. Furthermore, there was strong indication that the tissue did not infiltrate the pancreas. An enlarged lymph node was shown in the area between inferior vena cava and portal vein. According to the radiology report, lymphoma was the most probable diagnosis. Bone marrow aspirate was normal.

A biopsy was obtained via Endoscopic Ultrasound-Guided Fine-Needle Aspiration (EUS-guided FNA). The findings were indicative of non-Hodgkin lymphoma. Unfortunately, the samples were unable to confirm a firm diagnosis. The procedure was unsuccessfully repeated. Finally, Laparoscopic Biopsy (LB) was performed, which provided a

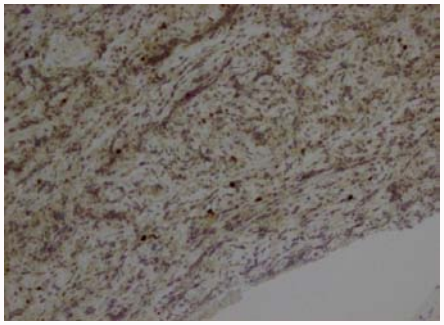

Figure 4: MUM-1 (x200).

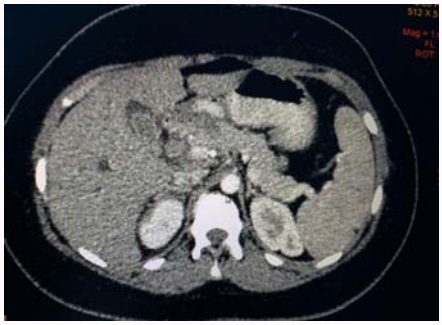

Figure 5: CT scan.

good enough sample to confirm the diagnosis. The pathology report revealed Diffuse Large B-cell Lymphoma. In immunohistochemistry, the tissue cells were CD79 (100%), CD19 (90%), bcl2 (10%), bcl6 (40%), CD5 (<=5%), CD43 (30%), κ/λ=1, IgM (100%), IgG (100%), MUM (90%), BOB-1 (20%), PAX-5 (20%), MIB-1 (>90%, but not 100%) positive. Furthermore, a cluster of T-cells (CD3+) was reported in less than 5%. The tissue tested negative for CD10, CD34 (Qbed-10), CD30, CD56, Cyclin-D1, CD99, Desmin, MyO-D1, EMA, PHOX-2B, INI-1, S100p, SYN, TdT, WT1 (6FH2), CD57 and Chromogranin. Disease was classified as stage II (Lugano classification).

Results

Treatment was initiated, according to FAB LMB 96 protocol. She was classified as group B (non-resected stage II tumor) and received chemotherapy as in arm B4 without randomization. Bilirubin rates had already decreased following the first biopsy and normalized after the first chemotherapy course (Figure 1). The first reevaluation ultrasound indicated adequate response to COP. The second reevaluation ultrasound following COPADM1 showed complete remission. She continued her treatment without significant problems. At third reevaluation point, following CYM1, both ultrasound and MRI verified complete remission. However, due to the most specific results that CT scan initially provided for our patient, the investigation was repeated at end-of-treatment and confirmed lymphoma remission. She concluded her treatment uneventfully (Figure 2).

She is still under intensive evaluation with monthly ultrasound and liver function tests. She remains in remission 8 months after treatment conclusion.

Discussion

Diffuse Large B-Cell Lymphoma (DLBCL) is a neoplasm that originates from transformed mature B cells [5]. Current studies suggest that DLBCL represent approximately 10-12% of all types

of NHL in childhood. It is rarely diagnosed in children less than 4 years old but the incidence rises with increasing age. Contrary to NHL, which present most commonly in abdomen, the most usual presenting site of DLBCL is the mediastinum [6]. In addition, DLBCL most commonly presents as a localized disease, making ascites or pleural effusion a rare finding [7]. LDH is seldom elevated. Bone Marrow (BM) or Nervous System (CNS) involvement is not frequent (Figures 3 and 4).

Although DLBCL is a distinct neoplasm from Burkitt lymphoma, treatment based on LMB regimen is commonly used worldwide. More specifically, FAB-LMB 96 protocol seems to provide an event-free survival rate of approximately 92 ± 7 % for children with DLBCL (without CNS or BM involvement) [8].

The main challenge for our patient was to confirm the diagnosis. Despite being an excellent diagnostic tool, ultrasound failed to determine the diagnosis in our case. However, MRCP indicated that common bile duct obstruction was due to external pressure. Furthermore, CT scan proved to be superior to MRI as a primary imaging tool (Figure 5). This was mainly attributed to better bowel imaging because of orally given contrast.

Apart from imaging difficulties, obtaining a biopsy also proved challenging. EUS-guided FNA is considered a noble and excellent method. Although it failed to confirm the diagnosis in our case due to inadequate sample size and lower quality, it is considered a promising method in order to obtain a biopsy. Our patient's diagnosis was finally obtained via laparoscopic biopsy. Thus it is clear that minimally invasive techniques have a clear advantage in obtaining a biopsy in such cases.

In conclusion, DLBCL is a rare type of abdominal lymphoma. However, with high clinical suspicion, proper imaging and persistent efforts for a proper biopsy, diagnosis can be obtained.

References

1. Watanabe Y, Ito T, Horibe K, Harada T, Ando H, Seo T et al. Obstructive jaundice, an unusual initial manifestation of intraabdominal non-hodgkin's lymphoma in children: Complications of percutaneous transhepatic cholangial drainage. J Pediatr Surg. 1997; 32(4): 650-3.
2. Yang T, Pan J, Zou Y. Obstructive Jaundice caused by Non-Hodgkin Lymphoma in common bile duct. J Pediatr. 2015; 167(5):1168.
3. Kurosawa H, Matsunaga T, Shimaoka H, Sato Y, Kuwashima S, Sugita K et al. Burkitt lymphoma associated with large gastric folds, pancreatic involvement, and biliary tract obstruction. J Pediatr Hematol Oncol. 2002; 24(4):310-2.
4. Ugur H, Tacyildiz N, Yavuz G, Unal E, Sayili A, Emir S et al. obstructive jaundice: An unusual initial manifestation of intra-abdominal non-hodgkin lymphoma in a child. Pediatr Hematol Oncol. 2005; 23:87-90.
5. Bakhit M, Mc Carty TR, Park S, Njei B, Cho M, Karagozian R et al. vanishing bile duct syndrome in hodgkin's lymphoma: a case report and literature review. World J Gastroenterol 2017; 23(2): 366-72.
6. Minard-Colin V, Brugières L, Reiter A, Cairo MS, Gross TG, Woessmann W et al. Non-hodgkin lymphoma in children and adolescents: Progress through effective collaboration, current knowledge, and challenges ahead. J Clin Oncol. 2015;33(27):2963-74.
7. Reiter A, Klapper W. Recent advances in the understanding and management of diffuse large B-cell lymphoma in children. Br J Haematol. 2008;142:329-47.
8. Oschlies I, Klapper W, Zimmermann M, Krams M, Wacker HH, Burkhardt B, et al. Diffuse large B-cell lymphoma in pediatric patients belongs predominantly to the germinal-center type B-cell lymphomas: A clinicopathologic analysis of cases included in the German BFM (Berlin-Frankfurt-Munster) multicenter trial. Blood. 2006; 107(10);4047-52.