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Severe Jaundice, due to Vanishing Bile Duct Syndrome in Hodgkin's Lymphoma, Fully Reversible after Chemotherapy

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Abstract

Liver involvement in Hodgkin's lymphoma is common and is caused by hepatic infiltration, biliary obstruction by lymphoma, hepatitis, sepsis or complications of chemotherapeutic treatment. Jaundice caused by the vanishing bile duct syndrome related to Hodgkin's lymphoma is very rare. The mechanism is poorly understood but a paraneoplastic effect seems most likely as liver biopsy and imaging study the absence of lymphoma cells in liver. We describe a 13-year-old man with vanishing bile duct syndrome and Hodgkin's lymphoma who was treated successfully with chemotherapy. The markedly elevated serum Bilirubin levels completely normalized. Our case demonstrates that although dosing of chemotherapy in this situation can be very difficult, a good clinical outcome is possible, which makes the attempt at curative treatment worthwhile.

Keywords: Hodgkin's lymphoma; jaundice; vanishing bile duct

Introduction

Hodgkin's disease is among the common diseases that affect the lymph nodes and can present as fever of unknown origin. However, its presentation with obstructive jaundice has been reported only rarely [1-4]. Although it can present some times as a febrile illness [5], a cholestatic presentation with febrile illness is rare [6]. We present a case here in which a patient developed prolonged fever with jaundice.

Case Report 1

Patient is 13 years old boy who presented with history of swelling in right neck for 1 year. Which size increases progressively fever and jaundice for two months. The fever was high grade and continuous and it was associated with sweating. His previous work up has been negative for malaria, typhoid and tuberculosis. He has been treated with antituberculosis drugs and antibiotics without any improvement. Examination revealed an ill-looking boy with a of 1000F, pulse 100/min. He was jaundiced. Liver and spleen was not palpable. No ascites or a mass was palpable. Rest of the examination was unremarkable on local neck examination 10x10 cm rubbery swelling in right neck, skin over swelling is tense, swelling was not mobile, temperature over swelling was not raised. No other lymph-

adenopathy. Laboratory investigations showed a hemoglobin 5.9g/dl, hematocrit 18.3%, MCV 57.0, a white blood cells count was 8400/cmm, and platelet count was 500,000/cmm. Electrolytes were normal. Total bilirubin was 7.0mg/dl, direct bilirubin 5.32mg/dl. Renal function normal. Hepatitis B surface antigen, anti-HCV, anti-nuclear antibody (ANA) and chest x-rays were normal. Biopsy from lymph node reported as hodkins disease nodular sclerosis. CECT (neck, chest, abdomen) scane shows extensive right cervical lymphadenopathy, no evidence of any mediastinal, axillary, retroperitonial, inguinal lymphodenopathy. Bone marrow biopsy shows no evidence of infiltration. Patients transfused three unit of blood, than haemoglobin 10.5g/dl. Started on chemotherapy (ABVD) and showed response. Clinically size of node decrease more than 50%. Jaundice subsidy (table 1) after 2 two cycles of chemotherapy.

Case Report 2

Patient was 28 years old boy who presented with history of swelling in left inguinal region, since 3 years, which increases progressive for l last 25 days, stool and dark coloured urine, loss of appetite and weight. There was no history of fever, cough, chest pain, bleeding per rectum and any alteration in bow2el habbits. On examination general condition fair, paller preaent, jaundice deep, no axillary cervical lymphadenopathy. Puls rate 120/min, blood pressure 140/90 mmHg. Chest bilateral clear, cardiovascular system normal, central nervous system normal, Per abdomen examination soft non tender, liver palpable 2 cm below right subcostal margine in mid clavicular line. local examination. Bilatreral inginal lymphnode palpable left side 8x9x7 cm, hard fixed non tender node palpable. Right small 2x2 cm none firm non tender mobile node palpable. Biopsy from left node done suggestive of non hodgkins lymphoma, poorly differentiated lymphocytic lymphoma. Liver biopsy suggestive of cholestasis in hepatocytes and in the biliary canaliculi. The portal tracts shows edema ductal proliferation and neutrophillic infiltration around the bile duct. Ultra sound abdomen shows liver normal, no mass are seen in porta hepatices region. Chest xray normal. Laboratory investigation hemoglobine 12.3 total lucocyte count 16,800, PCV 38, ESR 5. Total bilirubine 10.9mg/dl. Conjugated 6.8mg/dl, SGOT 146, SGPT 170, ALP 24. Patients started on chemotherapy with cyclophosphamide, vincristine, predneselone (COP) regime day 1 and day 8, after one cycle local radiotherapy to left inguinal area 2500Gy in 15 fraction 1.66 Gy /fraction from 14/1/1985 to1/2/85. After receiving 3 cycles and radiotherapy lymph node and bilirubine subside. Total 8 cycles of cycles of chemotherapy administrated, last on 31/7/85. No evidence of disease after last chemotherapy, patients on regular follow up and clinically NED. On follow up 2/4/13 patients develop swelling in left inguinal region 1.5x 1.5 cm. CECT chest and abdomen shows subcentric lymphadenopathy in aortocaval and paraaortic location, b/l inguinal lymphodenopathy largest of 9.1 mm. FNAC left inguinal suggestive of possibility of NHL small cell can't ruled out. Patients advised for biopsy.

Discussion

Liver involvement in Hodgkin's lymphoma is found in 5% of the patients at presentation and in 60% of autopsies. This liver involvement consists of hepatic infiltration of lymphoma, biliary obstruction by lymph node masses or infiltration of the common bile duct. Other hepatic disease secondary to drug toxicity, viral hepatitis or sepsis can also occur [7-10]. A less well known but particularly serious problem is the development of vanishing bile duct syndrome (VBDS). Including our patient, 18 cases with this complication have been reported [11-14]. As in our patient, most cases presented with neck swelling later on jaundice. The pathogenesis of VBDS in Hodgkin's lymphoma is unknown. One theory is that infiltration of lymphoma cells in the portal tracts destroys bile ducts. Another possibility is that cytokines released by lymphoma cells cause ductular destruction as a paraneoplastic phenomenon [15].

The observation that portal tracts usually contain no or only few lymphoma cells in patients with VBDS favors the latter theory [16]. The other causes of VBDS are listed in Table 2 [17] and most of them could easily be ruled out. Drug toxicity is a far more common cause of VBDS. In our patient there were no arguments for drug toxicity. The antibiotics were taken months before the jaundice appeared. Liver involvement in chronic EBV infection mostly consists of hepatitis sometimes in the presence of autoantibodies. One case shows a VBDS after an episode of hepatitis in a patient with chronic EBV infection, which appeared fully reversible after 1 year [18]. Nearly all previously reported patients died of liver failure, despite treatment-induced remission of Hodgkin's lymphoma [19-20]. Only Crosbie et al. described a single patient with successfully treated lymphoma who survived with serum bilirubin levels decreasing from 622 to 11 mmol/l. In a liver biopsy sample taken after therapy, ductular proliferation was detected [21]. In this journal Liangpunsakul et al. described a patient with a cholestatic liver disease caused by Hodgkin's lymphoma. The changes in the first sample may repre-

sent an early stage of the disease. In addition, this patient was successfully treated with chemotherapy [22]. Our case illustrates that when VBDS is diagnosed one should be aware of the possibility of Hodgkin's lymphoma as an underlying disease (Table 2) or hodkins lyphoma can cause VBDS. As in our patients liver biopsy not taken because CECT doesn't shows any abnormality, jaundice of patients completely resolve after two cycles of chemotherapy. Thus, we consider as it is case of vanishing bile duct syndromes.

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