Case Report

Primary hepatic Hodgkin's lymphoma: A case report

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Abstract

Introduction: Hodgkin lymphoma (HL) is an uncommon hematological malignancy that primarily occurs in young adults and less frequently in elderly individuals. HL has characteristics cells derived from B lymphocytes (known Reed–Sternberg (HRS) cells). Primary hepatic Hodgkin disease is very rare presentation accounting for less than 0.4% of the cases. Due to its rare occurrence, the pathogenesis of PHL is still unclear, Clinical manifestations, laboratory findings, and imaging features are usually nonspecific, making it difficult to diagnose. **Patient Concerns:** 69 years old Saudi Female, known case of Hypertension presented to our hospital with history of fever, jaundice, and poor appetite for about 2 weeks with significant weight loss. **Diagnosis:** Laboratory findings showed cholestatic pattern with total bilirubin 107.2 mg/dl, alkaline phosphatase 2076 IU/l, AST 153 IU/l and ALT 73 IU/l. Imaging with US revealed normal liver size with diffuse increase echogenicity, MRCP showed multiple stones within the gallbladder without evidence of obstruction or CBD dilatation and pan-computed tomography (CT) revealed mildly enlarged and fatty liver. CT-guided fine needle aspiration cytology (FNAC) and biopsy from the liver were consistent with primary hepatic Hodgkins lymphoma. **Intervention:** The patient received 5 cycles of ABVD. **Outcomes:** After the completion of the 5 cycles patient showed good response to the treatment with normalization of her liver function and regression in the size of liver on CT. **Conclusion:** PHL is a rare disease. The clinical presentation is variable and radiological features are not specific. Histology is mandatory for definitive diagnosis. The optimal therapy and outcomes for PHL is still unclear. ABVD is the most frequently used chemotherapy regimen. Multidisplinary approach including surgery and radiotherapy is another option.

MeSH Keywords: Diffuse liver involvement, extranidal, Hodgkin lymphoma, needle biopsy, rare, Saudi Arabia

Introduction

Hodgkin lymphoma (HL) is an uncommon hematological malignancy that primarily occurs in young adults and less frequently in elderly individuals. HL has characteristics cells derived from B lymphocytes (known Reed–Sternberg (HRS) cells). In Saudi Arabia HL accounts for 3.6% of all cancers and it is the seventh most common cancer. Patients with HL present with painless lymphadenopathy that slowly progressive. Primary hepatic Hodgkin disease is very rare presentation accounting for less than 0.4% of the cases. Due to its rare occurrence, the pathogenesis of PHL is still unclear, Clinical manifestations, laboratory findings, and imaging features are usually nonspecific, making it difficult to diagnose.

We report a rare case of Hodgkin lymphoma in a 69-year-old female with diffuse liver involvement.

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Case Report

Patient information

We report 69 years old Saudi Female, known case of Hypertension presented to our hospital with history of fever, jaundice, and poor appetite for about 2 weeks.

The patient noted loss in her weight which was unintentionally. She denied any history of blood transfusion, raw milk ingestion,

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recent travel, herbal medicine use, previous history of tattoos nor high risk behaviors.

Clinical findings

On examination, the patient was conscious and oriented, febrile (temperature 37.8–39°C).

Her physical examination was remarkable only for jaundice with no stigmata of chronic liver disease. There was no hepatosplenomegaly or lymphadenopathy. The skin had a normal appearance and temperature. No other abnormalities were found.

Diagnostic assessment

Laboratory findings were as follows: Total bilirubin 107.2 mg/dl, alkaline phosphatase 2076 IU/l, aspartate aminotransferase (AST) 153 IU/l and alanine aminotransferase (ALT) 73 IU/l [Table 1]. Serological studies for hepatitis A virus (HAV), hepatitis B virus (HBV), hepatitis C virus (HCV), Epstein Barr virus (EBV) and HIV were negative. Tests for antinuclear antibodies, antimitochondrial antibodies and anti-smooth muscle antibodies were negative. Serum alfa fetoprotein (AFP) level, serum carcinoembryonic antigen (CEA) levels were within normal limits. However, serum lactate dehydrogenase (LDH) level was raised.

The rest of the important laboratory results were within normal limits [Table 2].

Ultrasound of the abdomen revealed normal liver size with diffuse increase echogenicity keeping with diffuse fatty infiltration with no focal lesions or masses [Figure 1]. No common bile duct (CBD) stones or dilatation. The pancreas and spleen were normal.

Magnetic resonance cholangiopancreatography (MRCP) showed multiple stones within the gallbladder without evidence of obstruction or CBD dilatation. No definite abnormality within liver, spleen, and pancreas [Figure 2].

The Patient underwent pan-computed tomography (CT) scan as she continued to have spikes of fever and revealed mildly enlarged and fatty liver. The rest of the scan was unremarkable with no evidence of lymphadenopathy [Figure 3].

CT-guided fine needle aspiration cytology (FNAC) and biopsy from the liver were carried out Reed–Sternberg (HRS) cells and stained positively for CD30, PAX5 and CD20 while CD 15 and CD 45 were negative and showed.

Bone marrow examination did not reveal lymphoma infiltration.

Table 1: Liver function tests

Lab	2 Months before admission	At admission	Normal value
ALT U/L	13	73	UP TO 41
AST U/L	21	153	UP TO 40
ALK.PHOS U/L	120	2076	82-331
Total BILIRUBIN UMOL/L	3.7	107.2	0-17.1
CONGATED UMOL/L	1	105	0-3.4
GAMMA GT U/L	60	54531	8-61

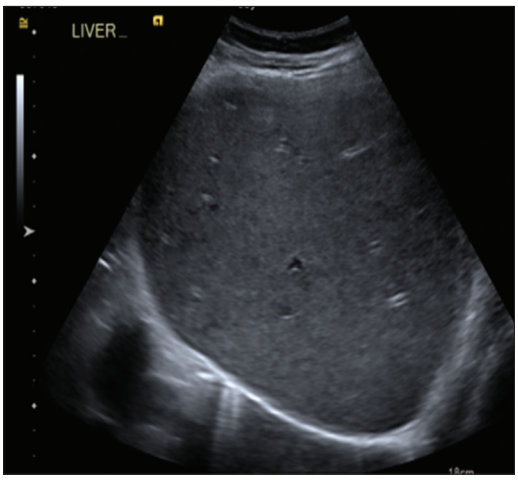

Figure 1: Ultrasound showing normal liver size with diffuse increase echogenicity keeping with diffuse fatty infiltration with no focal lesions or masses

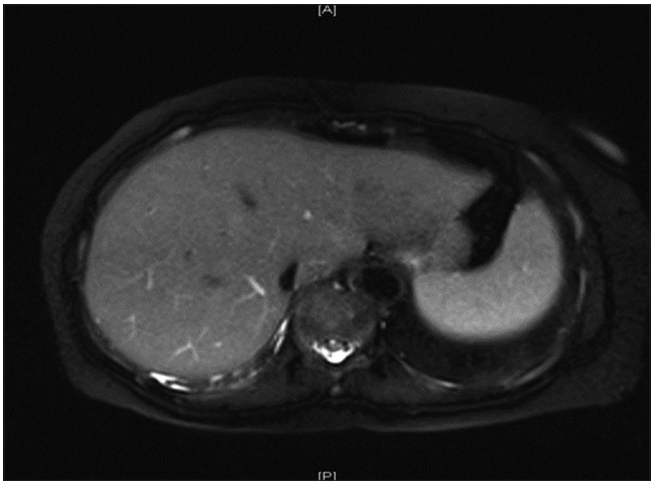

Figure 2: MRCP showing no definite abnormality within liver

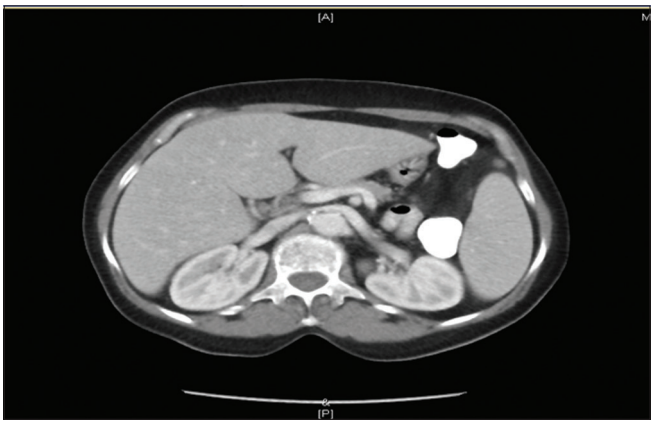

Figure 3: Computed tomography (CT) scan revealed mildly enlarged and fatty liver

Due to limitations in access to position emission tomography (PET)/computed tomography (CT) in our institution, PET/CT was not done.

These findings were consistent with primary hepatic Hodgkins lymphoma.

Therapeutic intervention

The patient received 5 cycles of ABVD.

Follow-up and outcomes

After the completion of the 5 cycles patient showed good response to the treatment with normalization of her liver function and regression in the size of liver on CT [Table 3].

Discussion

Hodgkin lymphoma (HL) is an uncommon hematological malignancy that primarily occurs in young adults and less frequently in elderly individuals. HL has characteristics cells derived from B lymphocytes (known Reed–Sternberg (HRS) cells) and are usually present within a microenvironment rich in immune effector cells.[1]

The overall incidence of HL is low, with an average incidence in European populations of ~2–3 per 100,000 individuals. However, HL is one of the most common cancers diagnosed in young adults in these populations.[2] In Saudi Arabia HL accounts for 3.6% of all cancers and it is the seventh most common cancer.[3]

Patients with HL present with painless lymphadenopathy that slowly progressive. symptoms are present in minority of patients secondary to compression (cough, chest pain, back pain and movement limitation), or B symptoms (night sweats, unexplained weight loss and fever).[4] Uncommonly, itchiness may the presenting symptom.[5]

Primary hepatic Hodgkin lymphoma (PHL) is very rare.[6] However, lymph node disease with secondary liver involvement is common and account for 5-10%.[7]

The etiopathogenesis of PHL is unknown. Multiple etiological factors have been proposed. Recent reports have described an increased incidence of PHL in patients with hepatitis C virus (HCV) infection, h HIV infection and Epstein–Barr virus (EBV).[8]

PHL commonly presents at 50 years. The clinical presentation is variable the commonest presenting symptom is abdominal pain. In one third of the patient B symptoms are the presenting feature. In less than 5% of patients present with Jaundice.[9] More importantly the presence of splenomegaly goes against a diagnosis of PHL.[10] Acute liver failure as initial presentation was reported in the literature.[11,12]

Table 2: Laboratory test

Lab	Result	Normal value
Complete blood count		
WBC (10 ×^{9}/L)	5.32	4.5-13.5
RBC (10 ×^{12}/L)	4.10	3.8-6.5
HGB G/L	120.0	11.5-180
HCT %	0.364	0.35-0.52
MCV FL	88.8	77-98
MCHC G/L	330.0	310-360
PLT	206	150-400
Inflammatory markers		
ESR MM/HR	13	0-20
CRP	7	Less 5.0
Electrolytes		
NA MMOL/L	138	136-145
K MMOL/L	4.3	3.5-5.1
UREA MMOL/L	2.0	2.76-8.07
CR UMOL/L	44	62-106
Lactic acid dehydrogenase		
LDH U/L	494	135-225
Hepatitis panel		
HBsAG Qual (S/N)	Negative	Negative
HEP A IgM Ab	Negative	Negative
HEPATITIS Bc IGM	Negative	Negative
HEP C VIRUS Ab (S/co)	Negative	Negative
Human immunodeficiency virus		
HIV 1, HIV 2 Ab	Negative	Negative
Brucella serology		
Brucella serology	Negative	Negative
Antinuclear antibody		
ANA	Negative	Negative
Antimitochondrial antibody		
Antimitochondrial Ab	Negative	Negative
Anti-smooth muscle antibody		
Anti-smooth muscle antibody	Negative	Negative
LKM antibodies		
LKM antibodies	Negative	Negative
Coagulation profile		
PT SEC	14.2	10.0-14.1
INR	1.23	0.86-1.2
APTT SEC	38.9	24.6-40.1

Table 3: Liver function tests post treatment

Lab	At admission	Post treatment	Normal value
ALT U/L	73	20	UP TO 41
AST U/L	153	25	UP TO 40
ALK.PHOS U/L	2076	117	82-331
Total bilirubin UMOL/L	107.2	3.6	0-17.1
CONGATED UMOL/L	105	1.5	0-3.4
GAMMA GT U/L	54531	152	8-61

Due to the rarity of the disease, the clinical presentation diversity and nonspecific radiological features (may manifest as a solitary lesion, multiple nodules and diffuse infiltration of the liver parenchyma), definitive diagnosis is made by liver biopsy.[13] The presence of elevated LDH in the presence of

normal AFP can point towards the diagnosis of PHL when suspected.[14]

The histological diagnosis of HL depends on finding diagnostic HRS cells and immunohistochemical staining for CD30, the B cell-associated antigen paired box protein Pax-5 (PAX5), CD15 and EBV.[1]

The optimal therapy and outcomes for PHL is still unclear, ABVD (doxorubicin, bleomycin, vinblastine and dacarbazine) is the most frequently used chemotherapy regimen, with patients receiving 2 to 6 cycles.[15]

Multidisciplinary approach including surgery and radiotherapy is another option, there are reports that liver resection followed by adjuvant chemotherapy and/or radiotherapy is associated with a good prognosis.[16]

This case encourages broaden the differential diagnosis of hepatic impairment to include PHL. Furthermore, lymphoma survivors are at increased risk of other malignancies as consequence of the chemotherapy in addition to cardiovascular disease, pulmonary disease, thyroid disease, and psychosocial issues. The diagnosis and the treatment complication will be detected earlier when the patient's care is being managed by a primary care provider.

Conclusion

PHL is a rare disease. The clinical presentation is variable and radiological features are not specific. Histology is mandatory for definitive diagnosis. The optimal therapy and outcomes for PHL is still unclear. ABVD is the most frequently used chemotherapy regimen. Multidisplinary approach including surgery and radiotherapy is another option. The diagnosis and the treatment complication will be detected earlier when the patient's care is being managed by a primary care provider.

Ethics approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Declaration of patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Conflicts of interest

There are no conflicts of interest.

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