CASE REPORT

PREGNANT LADY WITH UNDIAGNOSED HODGKIN'S DISEASE PRESENTING AS SECONDARY SCLEROSING CHOLANGITIS

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Cholestatic jaundice as a paraneoplastic feature of secondary sclerosing cholangitis is rare in Hodgkin's lymphoma. A 25 years old lady, 34 weeks primigravida was referred from the Emergency Department to the Medical Unit Khyber Teaching Hospital-MTI, Peshawar with four weeks of fever, progressive jaundice, pruritus, night sweats and weight loss. LFTs showed cholestatic picture, ERCP showed scanty intrahepatic giving beading and autumn tree appearance typical of sclerosing cholangitis. CBD was normal. Doppler U/S of hepatic and portal vein reported normal. She was started on steroids, ursodeoxycholic acid and antibiotics 3rd generation cephalosporins to which she did not respond well. This prompted a Liver biopsy which showed Hodgkin's disease having mixed cellularity. She was shifted to specialized oncology unit for further management where she died of irreversible liver damage. This is a rare case of secondary sclerosing cholangitis in Hodgkin's lymphoma of liver and the first case reported to our Hospital.

Keywords: Paraneoplastic; Hodgkin's lymphoma; Jaundice; Cholangitis
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INTRODUCTION

Secondary sclerosing cholangitis is a rare disease that has many underlying pathological causes which if timely addressed may have a favorable outcome.[1] The morphological and cholangiographic picture of secondary sclerosing cholangitis may mimic that of primary sclerosing cholangitis.[2] Any obstructive or infiltrative pathology affecting the biliary tree over long term may cause secondary sclerosing cholangitis. Choledocholithiasis, surgery, vascular insults, trauma, congenital fibrocystic diseases, primary immunodeficiency, toxins, chemicals, and certain drugs are some additional causes for its development.[3]

CASE REPORT

We report the case of a 25-year old lady 34 weeks primigravida presented to medical unit on call in Khyber teaching hospital Peshawar with 4 weeks history of fever, progressive jaundice, pruritus and 1-week history of abdominal distension. The fever was low grade most of the time with a periodicity of 8–10 days and an afebrile period of 7 days in between. There were associated constitutional symptoms of occasional night sweats, body aches, loss of appetite, nausea, and generalized body weakness. She lost around 5 kg of body weight since her illness started. The jaundice was progressive with mild generalized pruritus. Past and family history were unremarkable. Drugs history was also unremarkable except she used 2nd and 3rd generation antibiotics prescribed by her GP but with no improvement. On examination she was ill-looking, pale, deeply jaundiced, frail lady, with stable vitals and fully oriented running a temperature of 99.5 °F. Small bruises at injection sites were noted. There was no lymphadenopathy and peripheral stigmata of chronic liver disease. Abdominal was full in both flanks with shifting dullness elicitable. There was Painless hepatosplenomegaly with each 4-finger breadth enlarged below costal margins. The gravid uterus correlating with the 34 weeks gestation with obvious fetal heart sounds were present.
Labs revealed microcytic hypochromic anemia with Hb of 5.6 g/dl, platelet count 120,000 /cmm, TLC 15,400 /cmm, Polymorphs 80%, lymphocytes 18%, monocytes 3%, eosinophils 1%, ESR 75 mm/1st hour, LDH 1047 U/l, CPK 15 U/l. LFTs showed S. bilirubin 7.82 mg/dl, direct bilirubin 4.5 mg /dl, indirect bilirubin 3.0 mg/dl, ALP 1262 U/l, ALT 45 U/l, gamma GT 316 U/l. Serum Calcium 8.8 mg/dl, albumin 1.9 g/dl, S. uric acid 6.2 mg/dl, INR 1.5, APTT 7 sec prolonged, D-dimers 5179 ng/ml, HBsAg, Anti HCV and Anti HIV negative. ANA was negative, ASMA (anti smooth muscle antibody) negative, AMA (anti mitochondrial) negative. Renal profile, S. electrolytes, ABGs and Urine RE unremarkable. U/S abdomen revealed hepatosplenomegaly with multiple hypoechoic micronodules in liver parenchyma, moderate ascites and lymphadenopathy in porta hepatis and pre-aortic region. CT abdomen findings were consistent with U/S abdomen with hypodense micronodules scattered in liver parenchyma and sub-centimeter lymph nodes in porta hepatis and pre-aortic region (Figure-1). CXR was unremarkable. Ascitic fluid R/E revealed exudative ascites with no malignant cells seen. Bone marrow biopsy was reported unremarkable.
ERCP showed scanty intrahepatic giving beading and autumn tree appearance typical of sclerosing cholangitis. CBD was normal. Doppler U/S of

hepatic and portal vein reported normal. After delivery U/S-guided liver biopsy from one of the multifocal nodules revealed a tissue diagnosis of Hodgkin's lymphoma, mixed cellularity subtype. She received 3rd generation cephalosporins, IV steroids, ursodeoxycholic acid, IV fluids, and hematinics, with no improvement. We shifted her to oncology unit for specific management where she unfortunately died of irreversible liver damage.

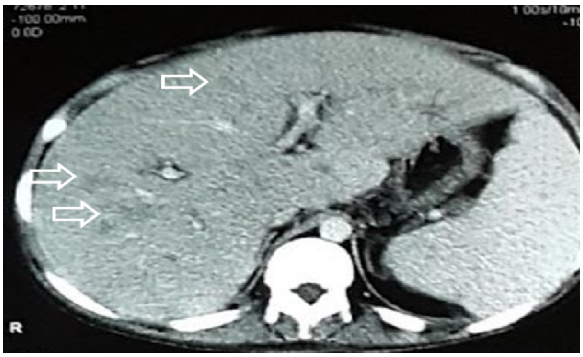

Figure-1: Showing Axial CT scan slice of abdomen showing scattered hypodense nodular lesions in the liver parenchyma (arrows) and splenomegaly (*).

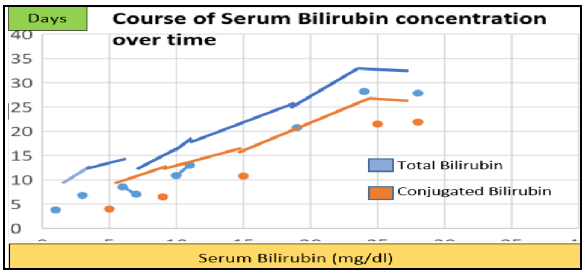

Figure-2: Chart showing the rising trend of both serum total bilirubin and conjugated bilirubin over time in our patient.

DISCUSSION

Liver damage is relatively common in patients affected by Hodgkin's disease. Cholestatic jaundice as a paraneoplastic feature of secondary sclerosing cholangitis is rare in Hodgkin's lymphoma. Cholestatic jaundice in Hodgkin's lymphoma although rare has many causes including biliary obstruction by portal lymphadenopathy, viral hepatitis, drugs toxicity, and vanishing bile duct syndrome (VBDS). VBDS is an uncommon though a serious cause of cholestasis in Hodgkin's lymphoma (HL).[4] Secondary sclerosing cholangitis as a paraneoplastic manifestation of HL has a good prognosis than VBDS and can be steroid responsive.[5] VBDS is nearly fatal except some cases have shown reversibility after liver transplantation.[6] Primary sclerosing cholangitis (PSC) as opposed to SSC is a chronic progressive cholestatic liver disease of unknown etiology and with no specific treatment.[7]

This report emphasizes that any infiltrative liver parenchymal disease including both primary Hodgkin's lymphoma of the liver and secondary liver involvement by Hodgkin's lymphoma that causes prolonged cholestasis; damage the intrahepatic biliary channels by different mechanisms, leading to secondary sclerosing cholangitis. The cholangiographic picture of the secondary sclerosing cholangitis may mimic primary sclerosing cholangitis. Therefore, keeping in view, the clinical background of the patient the diagnosis of primary sclerosing cholangitis can only be made after carefully ruling out all the known causes of secondary sclerosing cholangitis. The overall prognosis and outcome depends on many factors including the rapid onset of unremitting jaundice within weeks as in our patient, which indirectly reflects the extent of damage to intrahepatic biliary channels by lymphoma cells. In some cases of secondary sclerosing cholangitis where the cause is timely cured, further progression may be arrested but at the stage where extensive irreversible damage to intrahepatic biliary channels occurs as in our patient, liver transplantation may be the only way of improving survival regardless of the extrahepatic spread of Hodgkin's lymphoma. This is a rare case of secondary sclerosing cholangitis in Hodgkin's lymphoma of liver and the first case reported to our Hospital.

REFERENCES

1. Abdalian R, Heathcote EJ. Sclerosing cholangitis: A focus on secondary causes. Hepatology 2006;44(5):1063–74.
2. Krishnamurthy GT, Krishnamurthy S. Nuclear Hepatology: A textbook of Hepatobiliary diseases. Berlin: Springer; 2000.
3. Kariv R, Konikoff F. Sclerosing cholangitis--primary, secondary and more. Isr Med Assoc J 2002;4(12):1141–2.
4. Jaffe ES. Malignant lymphomas: Pathology of hepatic involvement. Semin liver Dis 1987;7(3):257–68.
5. Abedi SH, Ghassami M, Molaei M, Mohsenifar Z, Mohammad Alizadeh AH. Secondary Sclerosing Cholangitis and Hodgkin's lymphoma 2015;8:83–7.
6. Hubscher S, Buckels JA, Elias E, McMaster P, Neuberger JM. Reversible vanishing bile duct syndrome after liver transplantation: Report of 6 cases. Transplant Proc 1991;23(1 Pt 2):1415–6.
7. LaRusso N, Shneider BL, Black D, Gores GJ, James SP, Doo E, *et al*. Primary sclerosing cholangitis: Summary of a workshop. Hepatology 2006;44(3):746–64.

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