

Case Report

Solitary Langerhans cell histiocytosis of liver with sclerosing cholangitis in an adult female

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ABSTRACT

Liver is affected as a late complication of high risk cases of Langerhans cell histiocytosis. Sclerosing cholangitis is a rare pattern associated with Langerhans cell histiocytosis of liver, which is even rarer in the adult population and has high mortality. The treatment is difficult and may require liver transplantation. We report a unique case of a 40-year-old female who developed sclerosing cholangitis associated with Langerhans cell histiocytosis without any evidence of involvement of other systems. Our patient required only surgery, and had been followed up for two years without recurrence. We could not find any other case of solitary liver involvement of Langerhans cell histiocytosis in literature published so far.

Keywords: Adult LCH, Liver LCH, Sclerosing cholangitis

INTRODUCTION

Langerhans cell histiocytosis (LCH) is primarily a disease of childhood. LCH affecting adults is rare. Liver is affected, usually, only as a late complication of high risk disease. Sclerosing cholangitis is a rare pattern associated with Langerhans cell histiocytosis of liver, which is even rarer in the adult population and has high mortality. Such cases are managed by systemic therapy and /or liver transplantation. We present an adult case of LCH affecting liver, with sclerosing cholangitis, without evidence of any other organ involvement and treated by surgery alone.^{1,2}

CASE REPORT

40-year-old female presented with c/o recurrent epigastric and right hypochondrial pain, associated with low grade fever, jaundice and loss of weight. There was no history of pruritus, edema or abdominal distension. She did not have any chronic illnesses. She had similar complaints 2

years back for which she consulted another hospital. Her investigations then, were normal. Liver biopsy was inconclusive.

On examination, epigastric tenderness was there. There was no hepatomegaly. Spleen tip was palpable. There was no ascites.

Her total WBC count was 16000/dl; alkaline phosphatase was 282U/L (33–96 U/L), serum amylase-1640 U/L (20–96 U/L), serum lipase-2290 U/L (3–43 U/L).

USG suggested recurrent pyogenic cholangitis with cholangitic abscess. MRCP showed dilated CBD with smooth tapering and irregular dilatation of intrahepatic biliary radicles in both lobes with strictures and cholangitic abscess and suggested a diagnosis of sclerosing cholangitis. CECT abdomen showed hepatomegaly with central and peripheral intrahepatic biliary radicle dilatation, dilated CBD with thickened walls and smooth tapering at head of pancreas. A few

non-enhancing hypo dense areas adjacent to terminal parts of intrahepatic ducts on left side were noted and cholangitic abscess was suggested.

She was posted for cholecystectomy. Intraoperatively, left lobe of liver appeared fibrotic with areas of bile stasis. So, left hepatectomy, cholecystectomy and hepaticojejunostomy were done.

Grossly, left hepatectomy measured 7x6.5x5 cm and showed ill-defined fibrotic areas with thickened bile ducts. Another ill-defined area of necrosis was also seen. Gall bladder was normal. Microscopically, bile ducts with concentric fibrosis with characteristic “onion skin appearance” were noted (Figure 1).

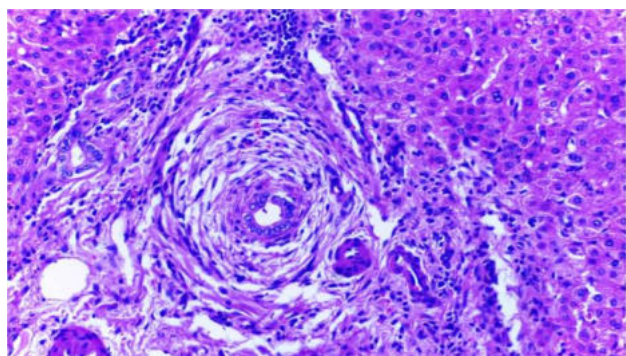


Figure 1: Concentric fibrosis around bile ducts with characteristic onion skin appearance.

There was mild to moderate lymphocytic infiltrate within the portal triad. Ductular reaction was present. Another focus showed area of necrosis, surrounded by plenty of eosinophils and cells with moderate amount of cytoplasm, oval elongated nucleus with longitudinal grooves-suggestive of Langerhans cells (Figure 2).

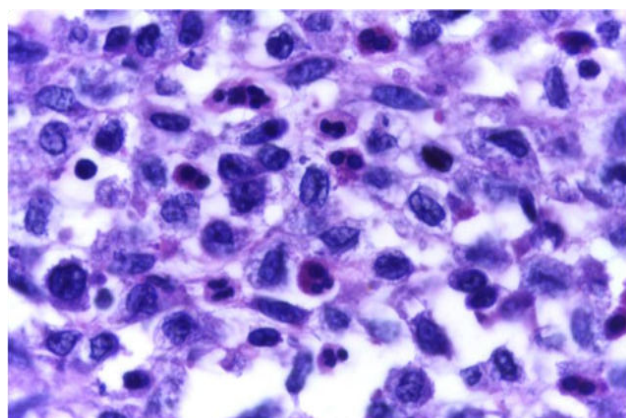


Figure 2: Cells with elongated nucleus and longitudinal Grooves (Langerhans Cells) admixed with numerous eosinophils.

These cells were positive for CD1a (Figure 3) and S100. There was no granuloma. So, the final diagnosis was Langerhans cell histiocytosis of liver with secondary sclerosing cholangitis.

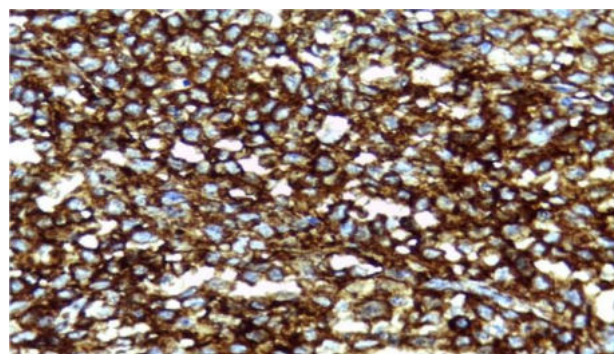


Figure 3: Strong membranous CD1a positivity in the lesional cells.

Following the diagnosis, she was extensively investigated for involvement of other systems. Her peripheral smear and bone marrow examinations were normal. CT or USG did not show evidence of other visceral organ or bone involvement. She did not have any skin lesions or polyuria. So, present case is unique, as it is a solitary liver involvement of an adult by Langerhans cell histiocytosis with secondary sclerosing cholangitis. Our patient was treated with surgery alone. She has been under follow up for the past 2 years and has not developed any new lesions so far

DISCUSSION

Liver involvement in Langerhans cell histiocytosis (LCH) in adults is poorly recognized and frequently overlooked.¹ The incidence of LCH in adults is roughly 1-2 cases per million.² The diagnosis of LCH is established by histopathologic examination. A definitive diagnosis requires CD1a positivity by immunohistochemistry in the lesional cells.¹ When LCH infiltrates the liver, it shows a propensity for the biliary ducts, leading to the development of cholestasis. M. Abdallah et al describes two distinct forms of LCH liver involvement: 1) nodular Langerhans' cell infiltration of the liver, 2) LCH liver involvement, centered on bile ducts with little or no histiocytic infiltration, which is associated with chronic fibrosis progressing to sclerosing cholangitis (SC).³ According to the description given by Ibrahim Hatemi et al, the most common affected age was above 40 years. Majority were females.⁴ Sclerosing cholangitis can be a late manifestation in most but a few patients presented with features of SC. Laboratory findings of cholestasis in a patient with LCH may reflect the damage of large or medium sized bile ducts.⁴ LCH causing SC in adults is rare, but has high mortality. Treatment of multisystem LCH is systemic therapy, i.e., steroids with methotrexate and/or vinblastine.⁵ According to a number of studies, involvement of high-risk organs including the liver, spleen, lungs and hematopoietic system poses high mortality (30-50%).^{2,5} The 3-year survival rate with liver involvement is 51.8%, compared with that of 96.7% without liver involvement.² Late LCH liver involvement with sclerosing cholangitis, cirrhosis, or liver insufficiency is difficult to treat. In such cases, liver

transplantation is the only treatment option.⁵ The long-term efficacy of liver transplantation remains unknown.⁴

CONCLUSION

In conclusion, our case is unique in that, it is an adult case of Langerhans cell histiocytosis involving only the liver with secondary sclerosing cholangitis treated by surgery alone. Patient should be followed up for early detection of involvement of other systems or recurrence.

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Ethical approval: Not Required

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