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Langerhans Cell Histiocytosis Associated a Chronic Colangiopaty

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Abstract

Langerhans cell histiocytosis (LCH) is a rare neoplasm characterized by the presence of positive CD1a histiocytes. Predominates in the pediatric age with cutaneous lesions as the first manifestation. We present a case of 47-year-old male with history of diabetes insipidus and one year of multiple episodes of acute cholangitis, liver biopsy showed architectural distortion, portal fibrosis, portal spaces with inflammatory infiltrate composed by histocytes, lymphocites, plasma cells, foamy histiocytes and abundant eosinophils, granulomas, bile duct damage and ductular reaction with acute cholangiolitis. The patient was subjected to chemotherapy and with morphologic remission pending for transplantation. Although adult presentations of LCH is described in the literature this patient's age is unusual. As a morphological finding, we shown that the presence of granulomas with histiocytes and eosinophils in biopsies should be considered as a suspicious finding for LCH and Hodgkin lymphoma and Mastocytosis should, however, be consider in the differential diagnosis.

Keywords: Langerhans cell histiocytosis; Liver neoplasm; Adult liver biopsy; Acute cholangiolitis; Macrophage activation syndromes

Abbreviations: LCH: Langerhans Cell Histiocytosis; HE: Hematoxylin and Eosin

Introduction

Histiocytes derive from hematopoietic myeloid progenitors, which further differentiate into monocytes, macrophages and dendritic cells [1]. Originally where a group of rare disorders characterized by pathologic accumulation of cells derived from the monocyte, macrophage, and dendritic cell lineage. In 1987, the Working Group of the Histiocyte Society classified the histiocytoses as Langerhans cell related, non-Langerhans cell related, or malignant [2], however, understanding molecular mechanisms permitted a new classification integrating both clinical, pathological and molecular findings; divides the histiocytoses into 5 categories: Langerhans (L), cutaneous and mucocutaneous (C), malignant (M), Rosai Dorfman disease (R), and hemophagocytic lymph histiocytosis and macrophage activation syndrome (H)[3].

Case Report

47-year-old male with history of diabetes insipidus and one year of multiple episodes of acute cholangitis associated with abdominal pain, jaundice, high bilirubin direct levels, transaminases and alkaline phosphatase. AMA negative. The cholangioresonance reveled global hepatomegaly with alteration of parenchymal intensity and stenosis of the intrahepatic bile duct. Liver biopsy showed architectural distortion, portal fibrosis, portal spaces with inflammatory infiltrate composed by histocytes, lymphocites, plasma cells, foamy histiocytes and abundant eosinophils, granulomas and bile duct damage with aberrant expression of cytokeratin 7 at periportal level and ductular reaction with acute cholangiolitis. Hepatocanalicular cholestasis, xanthomatous change with periportal copper deposits. CD1a and s100 revealed positivity in the portal histiocytes (Figure 1). The microscope used was an Olympus BX51 series 9C13849 with an Olympus DP72 camera and the Quick Photo Camera 3.2\RN.exe program to take the microscopic images. The patient was subjected to chemotherapy and with morphologic remission pending for transplantation.

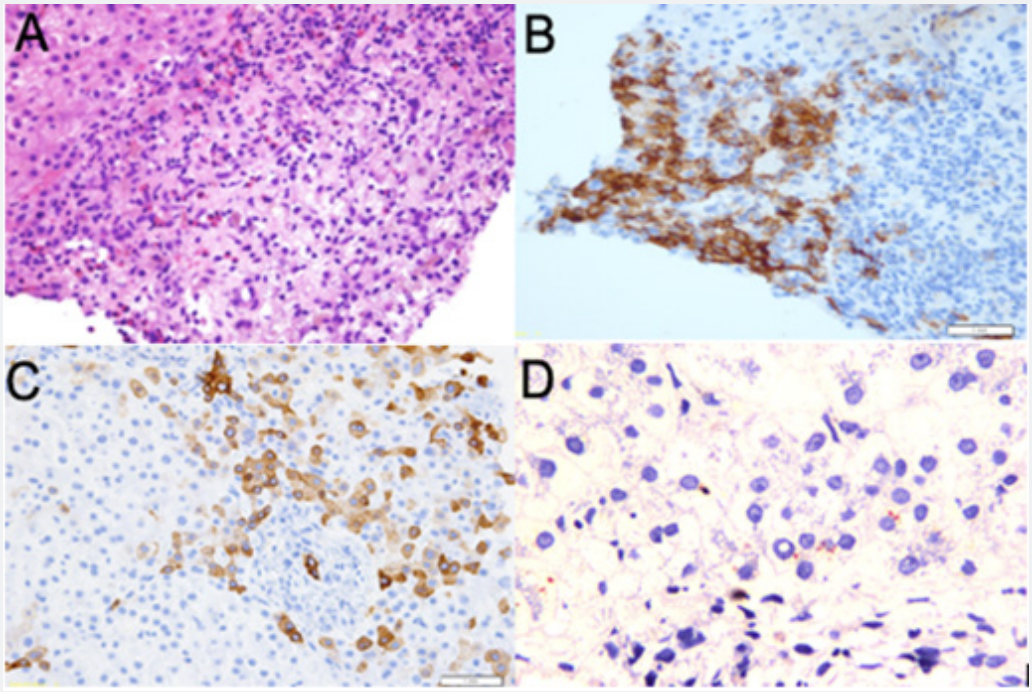

Figure 1: Liver Biopsy: (A). HE 10X, portal fibrosis and granulomas with histiocytes and eosinophils, (B). CD1a 10X, presenting positivity in the portal histiocytes, (C). Cytokeratin 7 10X, with damage at periportal level and ductular reaction with acute cholangiolitis, (D). Copper 40X, Periportal copper deposits.

Discussion

LCH is a rare disorder with an estimated incidence of 3 - 5 per million children and 1 - 2 per million adults annually in the United States [4]. We present a unique patient with a 47-year-old with liver LCH, diagnosis in adults is extremely uncommon and even more rare in adult Hispanic males [5]. Clinical Presentation as in our case may manifest as unifocal or multisystem disease, including large variety of symptoms (dermatologic manifestations, endocrinopathies, neurological and even oral symptoms) [6] . The pathogenesis of LCH is not well understood and is probably related with clonal proliferation of Langerhans cells, some authors refers LCH as an atypical immunoreaction [7] and immortalization of Langerhans cells rather than uncontrolled cell proliferation [1]. Histopathologically, these immortalized cells infiltrate not looking as benign histocytes counterpart resembling tissue macrophages [8]. LCH has highly variability on its presentation as a consequence it should always be considered as a differential diagnosis, our reported case highlights this variability, a 47-year-old Hispanic male who initially presented diabetes insipidus and multiple episodes of acute cholangitis. The liver has a resident population of histiocytes that are related in local and systemic processes leading to increase in their numbers and activity [9] and LCH has particular tropism for the bile ducts leading to a sclerosing cholangiopathy and biliary cirrhosis [10]. Although adult presentations of LCH is described in the literature this patient's age and both biliary duct damage are unusual. As a morphological finding, we shown that the presence of granulomas with histiocytes and eosinophils in biopsies should be considered as a suspicious finding for LCH; Hodgkin lymphoma and Mastocytosis should however, be consider in the differential diagnosis.

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