中文題目：IgG4 相關性硬化膽道炎案例報告

英文題目：IgG4-related sclerosing cholangitis-case report

作　　者：凌旺業[1] 陳奕霖[1] 張學豪[1]徐永勳[2] 張廷安[3] 黃博琪[4]林志陵[5]
陳冠仰[5] 廖麗瑛[5] 王鐘貴[5]

服務單位：台北市立聯合醫院仁愛院區 一般內科[1]腎臟內科[2]病理科[3]消化內科[5]
台北市立聯合醫院和平婦幼院區 病理科[4]

Background:

IgG4-related sclerosing cholangitis is the most frequent extrapancreatic manifestation of type 1 autoimmune pancreatitis (IgG4-related). It rarely occurs in the absence of pancreatitis. Distinctions between primary sclerosing cholangitis and IgG4-related sclerosing cholangitis are crucial because of the drastically different prognoses. Currently, the differentiation is based upon tissue biopsy, increased IgG4 serum levels, and characteristic responsiveness to glucocorticoids. Here we will present a case of a man with poor appetite for 2 months and mild jaundice. A series of image studies was arranged and revealed dilation of intrahepatic bile ducts and common bile duct (CBD) lower end narrowing. With elevated serum IgG-4 level and proof of liver biopsy, IgG4-related sclerosing cholangitis is impressed. We recommend that IgG4-related disease should be included in the differential diagnosis of unexplained jaundice.

Case:

This 68 year-old man with past medical history of diabetes mellitus and chronic kidney disease came to our outpatient clinic due to generalized weakness and body weight loss with poor appetite for 2 months. Jaundice was then found. Blood tests showed abnormal liver function panel and elevated level of tumor marker of CA-199. Image studies of abdominal sonography and Magnetic resonance cholangiopancreatography (MRCP) revealed dilation of intrahepatic bile ducts (IHDs). Under the suspicion of hepatic hilar neoplasm or cholangiocarcinoma, endoscopic retrograde cholangiopancreatography (ERCP) was performed and showed saccular dilation of IHDs near the hilar region. Biliary stricture caused by sclerosing cholangitis was then suspected. The result of liver biopsy also compatible with sclerosing cholangitis. Serum IgG-4 level was then checked and showed highly elevated. IgG4-related disease was impressed. Further clinic follow up showed worsening renal function and renal biopsy was done as concerning new involvement to kidney. Result of renal biopsy revealed diabetic glomerulosclerosis. IgG4-related sclerosing cholangitis was finally diagnosed.