Primary sclerosing cholangitis in a child

朱娟秀

Lin WT;Lin SJ;Ni YH;Chen HL;Wang HP;Chu JS;Chang MH

摘要

Abstract

Primary sclerosing cholangitis (PSC) is a rare disease in Taiwan and has not been described in Taiwanese children previously. We report a 4-year- old girl who presented with prolonged fever, eosinophilia (11%), hepatomegaly, and markedly elevated serum levels of alkaline phosphatase (3, 318 IU/L) and γ-glutamyl transpeptidase (475 IU/L). Subsequent investigations including endoscopic retrograde cholangiopancreatography and liver histology confirmed the diagnosis of PSG. Treatment with a low dose of prednisolone for 2 months and ursodeoxycholic acid during 32 months of follow-up resulted in clinical remission and halted disease progression. A high index of suspicion is necessary for physicians to diagnose this disorder I children with chronic liver disease. Our experience in this case indicates that therapy with prednisolone and ursodeoxycholic acid may be helpful for the treatment of PSC in children, and suggests the need for more trials of combined therapy..