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HEPATOLOGY - General Hepatology

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A rare concurrence of two immune-mediated pathology; autoimmune hepatitis and lichen planus

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Objectives and Study: Autoimmune hepatitis (AIH) is a progressive inflammatory liver disorder characterized by elevated transaminases and immunoglobulin G levels, as well as positivity of non-specific autoantibodies. Lichen planus (LP) is a disease of unknown origin and considered to have an immune-mediated pathology. Association with diseases of immundysregulation such as alopecia areata, ulcerative colitis, vitiligo, dermatomyositis, morphea, sclerosing cholangitis, thyroiditis, immune thrombocytopenia, celiac disease, and insulin-dependent diabetes previously reported. To the best of our knowledge, association of these two immune-mediated diseases has not been described in childhood before

Results: A previously healthy 16-year-old boy was evaluated for itchy lesions on his knees and lateral parts of lower legs that had been present for 4 months. Dermatological examination showed purple, excorited, lichenified papules and plaques over the knees and lower legs (image 1). White reticular patches are observed on buccal mucosa. A punch biopsy from one of the lesions is performed and histopathological findings were consistent with LP and the lesions were unresponsive to topical treatment. During his follow up, laboratory investigations revealed high transaminase levels; alanine transaminase 203 IU/L and aspartate transaminase135 IU/L, and the patient was referred for further investigation. Infection serology including hepatitis B, hepatitis C, CMV, and EBV were negative. Metabolic tests, including alpha 1 antitrypsin and ceruloplasmin, were normal. The immunoglobulin G level was normal for his age and autoantibodies for AIH were negative, except for ANA (1/320) positivity. A liver biopsy was performed and showed portal and interface hepatitis, with a mixed inflammatory infiltrate composed of lymphocytes, plasma cells, and eosinophils (hematoxylin and eosin), rosette formation, porto-portal bridging and regenerative nodules. Pretreatment autoimmune hepatitis score was 16 (definite diagnosis >15). Oral prednisone 40 mg/day and azathioprine (2 mg/kg/day) were prescribed. Topical clobetasol propionate 0.05% and triamcinolone acetonide 0.1% were prescribed to apply on the skin and oral mucosa, respectively. After two months of follow up, transminase levels returned to normal (ALT 35 IU/L, AST 31 IU/L). Skin lesions also improved markedly with both oral and topical steroid treatment.

Conclusion: Although both diseases have been frequently reported to be associated with other autoimmune diseases, to the best of our knowledge AIH and LP in the same patient has not been reported previously. We are not sure whether this situation is a coincidence or a real concurrence of two diseases, we need to evaluate more patients with OIH or LP during their follow up to reveal the real association between these disorders.