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CASE REPORT

Probable autoimmune hepatitis and systemic sclerosis complicated by reversible posterior leukoencephalopathy syndrome: An unusual association

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Summary Systemic sclerosis is a multisystem autoimmune rheumatic disorder characterized by fibrosis in the skin and internal organs but rarely with hepatic or brain involvement. Reversible posterior leukoencephalopathy syndrome is characterized by a unique pattern of brain vasogenic edema in the setting of neurotoxicity predominantly in the parietal and occipital regions. We report a case of systemic sclerosis associated with probable autoimmune hepatitis that progressed rapidly to reversible posterior leukoencephalopathy syndrome with loss of vision, seizures, and coagulopathy. Brain computed tomography showed faint low density in the bilateral occipital lobes and posterior parietal lobes with edematous change. Fortunately, the patient's clinical condition considerably improved 2 days following the initiation of 100 mg intravenous hydrocortisone infusion.

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Introduction

Systemic sclerosis (SS) is a systemic disorder characterized by inflammation, fibrosis and degeneration of the skin, blood vessels, and internal organs [1]. Nearly all patients with SS have peripheral vascular involvement with Raynaud syndrome, and >80% experience gastrointestinal function change such as dysphagia. SS rarely has hepatic involvement especially autoimmune hepatitis. Rodrigues et al [2] reported a patient with diffuse SS who was diagnosed with autoimmune hepatitis and who had an excellent response to immunosuppressive drugs.

Reversible posterior leukoencephalopathy syndrome (RPLS), or posterior reversible encephalopathy syndrome, is characterized by a unique pattern of brain vasogenic edema in the setting of neurotoxicity predominantly in the parietal and occipital regions [3]. It may develop in patients with immunosuppressed conditions and various rheumatologic diseases. Fugate et al [4] noticed that a substantial proportion of patients with RPLS had underlying autoimmune disorders. They described focal regions of symmetric hemispheric edema on computed tomography (CT) or magnetic resonance images [5]. However, the etiology of presentation was unknown; underlying vasculitis, cerebral autoregulation disorder, and hypertensive encephalopathy were considered [6]. RPLS is reversible and is characterized by headaches, decreased alertness, altered mental status, seizures, and visual loss.

We present a patient with underlying SS who developed acute liver injury, seizure, and visual loss, and whose condition considerably improved following hydrocortisone intravenous infusion.

Case report

A 71-year-old woman experienced nausea, vomiting, and intermittent epigastric pain for 10 days accompanied by

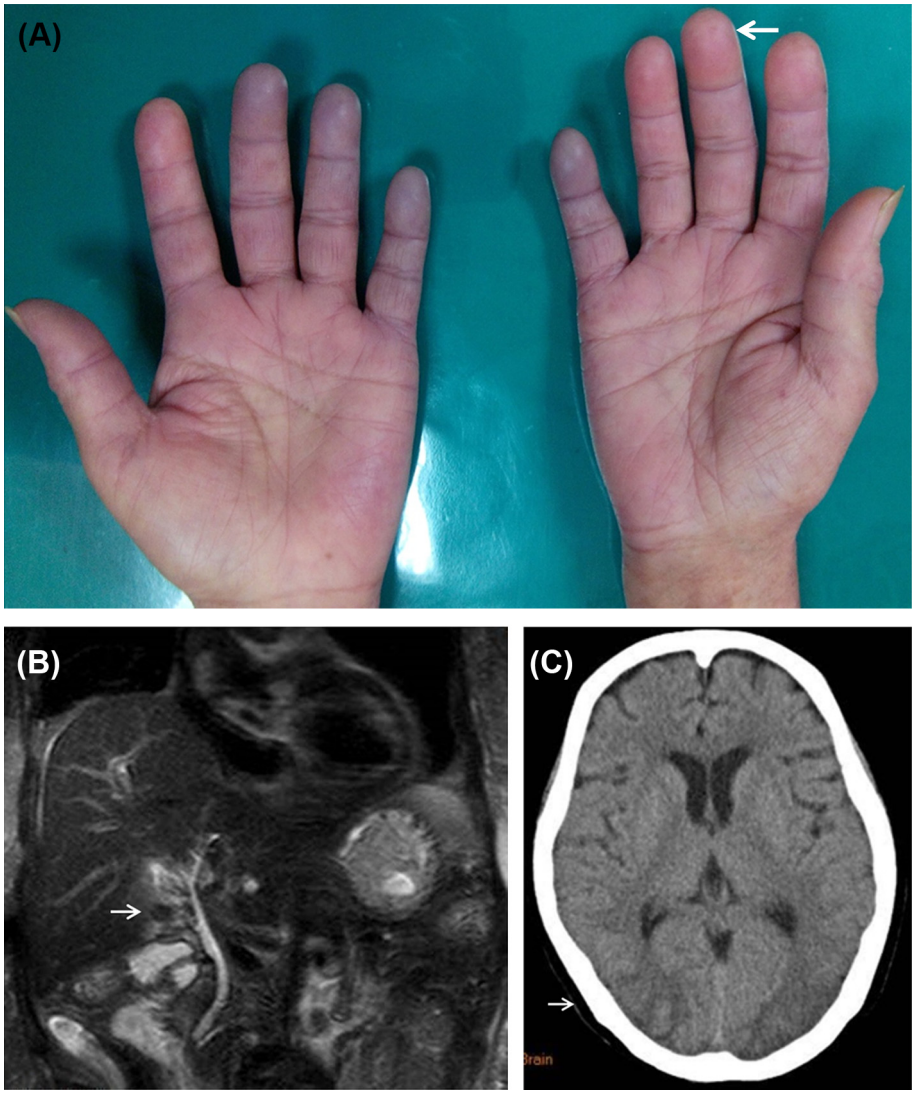

Figure 1 (A) The patient had Raynaud's phenomenon and digital pitting scar (white arrow). (B) The magnetic resonance cholangiopancreatography image shows gallstones (white arrow) with intact biliary duct. (C) The brain computed tomography images show a hypodense area in the occipital lobe, which is a characteristic of posterior reversible encephalopathy syndrome (white arrow).