Case Report

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Addisonian crisis in a patient with schmidt's syndrome

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Introduction

Schmidt's syndrome, also known as polyglandular syndrome type II, is a rare disease with potential life threatening consequences. It is characterized by multiple immune endocrinopathies, including Addison's disease with thyroid disease and/or type I diabetes.[1–3] We describe a case of Addisonian crisis in a patient with known Addison's disease. Interestingly, she also has type 1 diabetes mellitus and Graves' disease but was never previously diagnosed with Schmidt's syndrome.

Case

EPI: Exocrine Pancreatic Insufficiency; CCK: Cholecystokinin; HIV: Human Immunodeficiency Virus; FE-1: Fecal Elastase-1; PERT: Pancreatic Enzyme Replacement Therapy; PPI: Protompump Inhibitors; CF: Cystic Fibrosis; CFTR: Cystic Fibrosis Transmembrane Conductance Regulator; IBD: Inflammatory Bowel Disease; CD: Chron's Disease; UC; Ulcerative Colitis; AIDS: Acquired Immunodeficiency Syndrome; CT: Computed Tomography; MRCP: Magnetic Resonance Cholangiopancreatography; S-MRCP: Secretin-Enhanced Magnetic Resonance Cholangiopancreatography; EUS: Endoscopic Ultrasound; FNA: Fine Needle Aspiration; ERCP: Endoscopic Retrograde Cholangiopancreatography; Fcht: Fecal Chymotrypsin; PLT: Pancreolauryl Test.

Discussion

Schmidt's syndrome is a compendium of polyendocrine autoimmune diseases (Addison's disease, autoimmune thyroiditis/ Graves' disease, type I diabetes) and can be associated with other non-endocrine autoimmune disorders, such as myasthenia gravis, Sjogren's syndrome, and rheumatoid arthritis.[3] It is autosomal dominant with variable penetrance and usually affects middle-aged females (female-to-male ratio 3:1).[1] The diagnosis of this condition can be challenging due to its rarity (1.4 - 4.5 cases/100,000 population), atypical presentation, and variance in clinical symptoms depending on the type and severity of gland involved. Thus, a high index of suspicion is warranted for early diagnosis and appropriate hormonal therapy.[1]

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Conflicts of interset

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