G36 Undetected Polyglandular Autoimmune Syndrome Type II (Schmidt Syndrome) as a Cause of Sudden Death

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The goals of this presentation are to discuss the clinical features and disease association characteristics of the Polyglandular Autoimmune Syndrome Type II, and to increase the recognition of the morbidity and mortality arising from polyglandular autoimmune disorders.

This presentation will impact the forensic community and/or humanity by demonstrating how polyglandular autoimmune (PGA) syndromes are associated with a diversity of related diseases. Diabetes mellitus Type I is a common manifestation of PGA Types II and III. Less frequently observed is Addisonian crisis arising from co-occurring thyroiditis and adrenalitis in the absence of pancreatic involvement and resulting in sudden, unexpected death. The premorbid clinical and laboratory manifestations of this disorder can be subtle and non-specific, yet are critical benchmarks to be recognized in order to avoid a potentially lethal outcome.

Immunologic syndromes impacting multiple endocrine organs and giving rise to other nonendocrine immune disorders are a rare cause of sudden, unexpected death. Endocrine deficiency due to Polyglandular Autoimmune Syndrome can be brought on by infection, infarction, or tumor that results in the destruction of all or a large part of an endocrine gland. In most cases, however, the activity of an endocrine gland is depressed as a result of an autoimmune reaction that produces inflammation, lymphocytic infiltration, and partial or complete destruction of the gland. There are three patterns of Polyglandular Autoimmune (PGA) Syndrome, referred to as types I, II, and III. PGA Type I usually occurs in childhood and is characterized by hyperparathyroidism (79% of cases), followed by adrenal cortical failure (72%). Diabetes mellitus Type I seldom occurs. PGA Type II generally occurs in adults and always involves the adrenal cortex and frequently the thyroid gland (Schmidt syndrome) and the pancreatic islets. Type II is the most common of the syndromes and is characterized by the occurrence of autoimmune Addison's disease in combination with thyroid autoimmune diseases and/or diabetes mellitus Type I. The most frequent clinical association is between Addison's disease and Hashimoto's thyroiditis, while the least frequent clinical combination is Addison's disease, Graves' disease, and diabetes mellitus Type I. PGA Type III occurs in adults and does not involve the adrenal cortex, but includes at least two of the following: thyroid deficiency, diabetes mellitus, pernicious anemia, vitiligo, and alopecia.

Following the sudden death of a 38-year-old Caucasian female an autopsy revealed findings consistent with the diagnosis of Schmidt syndrome, or Polyglandular Autoimmune Syndrome Type II. While diabetes mellitus Type I was not diagnosed, as is the case in 50% of PGA Type II presentations, her past medical history included an ill-defined thyroid disorder and recent premorbid history of listlessness to extreme fatigue and non-specific somatic complaints. Her healthcare professional failed to make the correct premortem diagnosis. The differential diagnosis of this disorder is addressed, as is a brief discussion of the polyglandular autoimmune syndromes, with particular emphasis on the etiology, epidemiology, morbidity, and mortality associated with Polyglandular Autoimmune Syndrome Type II.

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