Objective: We report one case of autoimmune adrenalitis and one case of X-linked adrenoleukodystrophy (X-ALD) to highlight different presentations, possible etiologic and diagnostic challenges and the management of primary adrenal insufficiency.

Case presentation: Case 1, a 33 years old man was admitted to the Cardiac Intensive Care Unit with hypovolemic shock unresponsive to vasopressor agents and severe hyponatremia (117 mEq/L), after three days of pleuritic chest pain, myalgias and fever. He had physical signs of undiagnosed long-standing adrenal insufficiency and so hydrocortisone was administered with remarkable clinical and laboratorial improvement. Basal ACTH was 515 pg/mL, cortisol was undetectable, and aldosterone was 1,24 ng/dL. Fludrocortisone was also administered. Viral pericarditis was the presumed trigger of adrenal crisis, relapsing two more times during follow-up. Abdominal computed tomography revealed adrenal glands thinner than normal, adrenal autoantibodies couldn’t be measured, there were no findings suggestive of tuberculosis and the measurement of very long chain fatty acids (VLCFA) was normal. At the present time, he is asymptomatic without evidence of other glands involvement.

Case 2, a 28 years old man was referred to the Outpatient Endocrine Clinic with the diagnosis of primary adrenal insufficiency. Dexametasone at bedtime added to his previous hydrocortisone and fludrocortisone daily treatment significantly improve his well-being during early morning. He had a past history of seizures in childhood attributed to “meningitis”. Neurological examination and magnetic resonance were normal. VLCFA values were diagnostic for X-ALD. The patient has a female child.

Discussion: Autoimmune adrenalitis is responsible for 70 to 90% of primary adrenal insufficiency cases, half of these having one or more other autoimmune endocrine disorders. It is the initial manifestation in about 50% of patients with poliglandular autoimmune syndrome type II, with most cases occurring between age 20 and 40 years. The viral pericarditis presented could in fact be an autoimmune serositis, and the periodic follow-up should include an echocardiography besides the assessment of other endocrine glands. X-ALD, affecting the nervous system white matter, is an unusual cause of adrenal insufficiency. ‘Addison disease only’ is one of the three main phenotypes and most males only develop Adrenomyeloneuropathy by middle age. Phenotype variability often coexists in the same family. At-risk males should be informed of their risk of X-ALD. Approximately 20% of female carriers develop mild to moderate neurological symptoms.

Conclusion: Primary adrenal insufficiency is a rare and potentially life threatening disorder, but easily treatable. Correct etiologic diagnosis is important for adequate follow-up and possible genetic counselling.