Unsuspected adrenal masses, or incidentalomas, are increasingly found with the widespread use of thoracic and abdominal imaging; most cases do not pose a significant risk to a patient’s health. We describe a case of a patient with enlarged adrenals who was initially diagnosed with nonclassic congenital adrenal hyperplasia (NCAH), but 1 year later died from primary adrenal lymphoma dissemination.

In January 2013, an 83-year-old woman visited a general practitioner due to nonspecific chronic abdominal pain. Abdominal ultrasound suggested an adrenal mass and computed tomography (CT) scanning revealed bilateral adrenal enlargement. The glands measured up to 69 mm and had a baseline density of 38 HU and contrast washout rate below 60% (FIGURE 1A). No other abdominal lesions were detected.

In April 2013, the patient was admitted to our hospital for diagnostic workup. Physical examination was unremarkable except for mild abdominal tenderness. Routine laboratory test results were normal. The circadian cortisol profile, urinary cortisol excretion, low-dose dexamethasone suppression test results, as well as serum testosterone and androstenedione levels were normal. The baseline and stimulated 17-hydroxyprogesterone levels were increased to 6.94 ng/ml and 20 ng/ml, respectively.

The patient was diagnosed with NCAH and received no treatment. Because of the indeterminate CT features, adrenal biopsy was considered but the patient refused.

In September 2013, she presented with classic symptoms of adrenal crisis, and steroid replacement was initiated. Adrenal CT was ordered; however, due to the patient’s personal reasons, the scan was performed as late as in April 2014. It revealed enormous adrenals measuring up to 156 mm (FIGURE 1B) with periadrenal fat tissue infiltration.
Laboratory tests revealed elevated lactate dehydrogenase and β2-microglobulin in serum (1076 U/l and 8.6 mg/l, respectively). A core needle biopsy of the adrenal gland was performed, and a histopathological examination confirmed diffuse large B-cell lymphoma (DLBCL, FIGURE 1C–1E). The patient died before receiving the first dose of chemotherapy due to severe infectious complications. On autopsy, bilateral adrenal infiltration as well as involvement of the kidney, liver, stomach, peritoneum, and multiple periaortic lymph nodes were found.

In most cases, bilateral adrenal masses represent a benign adrenocortical disease: bilateral adenomas, macronodular hyperplasia, or distinct bilateral nodules with normal or atrophic cortex intervening. A differential diagnosis includes metastases, adrenal hemorrhage, some infections (eg, tuberculosis), and lymphoma. Current guidelines recommend a determination of whether an adrenal mass can be classified as a lipid-rich adenoma based on CT or magnetic resonance imaging. Other types of tumors increase the likelihood of malignancy and require in-depth diagnostic workup, careful follow-up, or surgery.

The initial CT findings in our patient were not consistent with adrenal adenoma. However, given the patient’s asymptomatic presentation and the hormonal test results, NCAH was diagnosed. The diagnosis was reconsidered after the patient developed adrenal crisis, and because of her poor compliance, the control CT and the resulting biopsy were delayed. Rapid tumor growth could have been probably detected by any suitable imaging modality, including ultrasound. The point-of-care ultrasound examination performed at the time of adrenal crisis could lead to a more timely diagnosis of adrenal DLBCL.

Although most patients with adrenal tumors have good prognosis, a thorough initial evaluation and periodic follow-up are necessary to identify those at risk for poor outcome. The presented case underscores the importance of the recently published guidelines concerning adrenal tumors.