Primary adrenalin sufficiency due to bilateral adrenal lymphoma

Insuficiencia suprarrenal primaria por linfoma adrenal bilateral

To the Editor:

A patient with adrenal insufficiency secondary to a bilateral primary adrenal lymphoma (PAL) is reported. A brief review of the literature on the most relevant clinical and pathological characteristics of this condition is also provided.

An 80-year-old female patient with a history of high blood pressure complained of constitutional symptoms, nausea, and vomiting during the previous four months. Supplementary tests performed revealed persistently elevated serum LDH levels (2,000 IU/L) and normal electrolyte levels. A CT scan of chest and abdomen (Fig. 1) showed masses in both adrenal glands. A study of the adrenal axis showed ACTH levels ranging from between 300 and 500 pg/mL (normal range, 10-50 pg/mL) and basal cortisol levels ranging from between 10 and 14 µg/dL (normal range, 3-18 µg/dL). Following glucocorticoid replacement therapy, laparotomy was performed, at which a biopsy could only be performed because both lesions were surgically non-resectable. A pathological study reported a large diffuse B-cell non-Hodgkin lymphoma, CD20 positive and Ki-67 positive in approximately 90% and P53 positive in approximately 80%. No bone marrow infiltration was found. Chemotherapy was started and well tolerated, and, at the time of writing, response to it is pending evaluation.

Primary lymphoma of the adrenal gland accounts for approximately 1% of extranodal lymphomas, and there are less than 100 tumors of this type reported in the literature. It is more common in males than in females (2/1), and mean age at tumor occurrence is approximately 68 years. Bilateral adrenal involvement is found in 65% of cases. Ninety percent of PALs are large B-cell non-Hodgkin lymphomas. The most common initial symptoms include abdominal pain, lumbar pain, fever, and weight loss. In patients with bilateral involvement, some degree of adrenal insufficiency may be found in up to 60% of cases, but insufficiency is usually subclinical in most of them.

Despite its low incidence, PAL should be included in differential diagnosis of an adrenal gland together with other malignant tumors such as carcinoma of the breast and lung, gastrointestinal tract tumors, and malignant melanoma, in which the incidence of adrenal metastases, often bilateral, is high. Differential diagnosis should also include other con-
ditions such as primary adrenal carcinoma, pheochromocytoma, and some infectious conditions. PAL is the disease with the poorest prognosis among extranodal B-cell lymphomas. Ninety percent of reported patients die within one year of diagnosis despite treatment. The chemotherapy schemes routinely used with curative intent for the treatment of high-grade B lymphomas provide very poor results in PAL. Radiotherapy may improve local control of the disease, but its impact on the survival of these patients is unknown. Finally, glucocorticoid replacement therapy should be part of the treatment when some degree of adrenal insufficiency exists.

References