Primary adrenal lymphoma with adrenal insufficiency: report of three cases and review of literature

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Introduction

Primary adrenal lymphoma (PAL) is an extremely rare disease with a high incidence of bilateral adrenal involvement whose prognosis is usually poor even with aggressive chemotherapy such as (R)-CHOP. Here we report three patients with PAL. All of them initially presented with adrenal insufficiency, which later turned out to be due to destruction of both adrenal cortices by diffuse large B-cell lymphoma (DLBCL).

Case 1

A 69-year-old man was referred to our hospital by an internist for further examination of lack of appetite, a 4.4-kg weight loss in one month, slight low back pain and bilateral 50-mm adrenal lesions. Before admission, dexamethasone 6.8 mg/day had been administered intravenously by the internist. Physical examination was normal, but laboratory evaluation showed hyponatremia and a low cortisol level of 0.5 µg/dl. Adrenal insufficiency was confirmed by a cosyntropin stimulation test. Hydrocortisone 15 mg/day was started orally. A CT-scan revealed 43-mm right adrenal mass and a 35-mm left adrenal mass. The patient underwent laparoscopic left adrenalectomy and was diagnosed to have DLBCL by microscopic and immunohistological examination although the right adrenal lesion responded to R-CHOP and radiation therapy, para-aortic lymph node swelling indicative of lymphoma relapse was newly found. Despite salvage chemotherapies, his general condition deteriorated and he died.

Case 2

A 68-year-old man presented with facial pigmentation, anorexia, a 4.5-kg weight loss in 2 months and low back pain lasting for one month. Laboratory analysis showed hypoglycemia and a high plasma ACTH level. Adrenal insufficiency was diagnosed by a cosyntropin stimulation test. Hydrocortisone 15 mg/day was started orally. A CT-scan revealed 43-mm right adrenal mass and a 35-mm left adrenal mass. The patient underwent laparoscopic left adrenalectomy and was diagnosed to have DLBCL by microscopic and immunohistological examination although the right adrenal lesion responded to R-CHOP and radiation therapy, para-aortic lymph node swelling indicative of lymphoma relapse was newly found. Despite salvage chemotherapies, his general condition deteriorated and he died.

Case 3

A 63-year-old man was referred and admitted to our hospital because of progressive anorexia, 4.8-kg weight loss in 4 months, and high fever and nocturnal sweating lasting for 3 weeks. Laboratory findings included a high ACTH level of 86.5 pg/ml, and serum cortisol levels responded inadequately to cosyntropin-ACTH administration, confirming the biochemical diagnosis of primary adrenal insufficiency. Hydrocortisone 15 mg/day was started orally and his symptoms were improved. An abdominal CT scan revealed bilateral adrenal masses. The right adrenal mass was 83 mm in diameter and left masses were 86 mm and 78 mm large, respectively. A whole body PET/CT showed FDG uptake in both adrenal masses, stomach wall, cardiac lymph nodes, and lymph nodes along the major and minor omentum. A CT-guided percutaneous needle biopsy of the right adrenal mass led to the diagnosis of DLBCL. After six cycles of R-CHOP therapy, the adrenal masses have shrunk remarkably (Partial Response).

Discussion

Epidemiology

- Rare disease <200 cases worldwide
- North America 20%
- The mean age = 52 years old
- Male: Female = 1.8:1

Chemotherapy Regimens

- CRP: 129 /mL
- C reactive protein: 129 mg/dl
- 2-5 year DFS rate: 51%
- 5-year EFS rate: 68%
- 5-year OS rate: 65%
- Complete remission: 55%
- Partial remission: 30%