PRIMARY BILATERAL ADRENAL LYMPHOMA: A CASE REPORT

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ABSTRACT

Primary lymphoma of both adrenal glands in a 59-year-old man is reported. The patient presented with fever and weight loss for 5 months. Computed Tomography (CT) scan of the abdomen revealed bilateral adrenal masses. Opened biopsy at right adrenal gland was done and immunostaining histologic report revealed large cell type of malignant lymphoma, whereas the bone marrow aspiration was negative. After initiation of chemotherapy, the patient developed febrile neutropenia, which was complicated by pneumonia and expired shortly thereafter. Literature about imagings of primary adrenal lymphoma was reviewed.

INTRODUCTION

Adrenal masses rarely result from involvement by lymphoma. Diffuse non-Hodgkin's disease is the most common type.[1-4] This may be found at presentation or at follow-up. In CT series, about 1-4% of patients being followed for lymphoma developed adrenal involvement.[2-4] Adrenal involvement is most commonly seen in conjunction with an extra-adrenal disease site particularly retroperitoneal adenopathy.[1, 5] Involvement of the adrenal glands were demonstrated up to 25% of patient with lymphoma in an autopsy series.[6] Primary adrenal lymphoma is rare and is believed to arise from hematopoietic cells in the adrenal glands.[7] The true incidence of these neoplasms is not known.[8] To our knowledge, this is the first reported case of primary adrenal lymphoma in Thailand.

CASE REPORT

A 59-year-old man, who had had underlying NIDDM type II for 10 years, presented with prolonged fever and weight loss for 11 kilograms in 5 months. The patient developed freguency of fever with chill, sweating, and pain at periumbilical area in the last 2 months. Neither splenomegaly, hepatomegaly nor lymphadenopathy was present. The computed tomography of the abdomen demonstrated bilateral adrenal masses (Fig 1,2). The right and left adrenal masses were 7x4.5 cm and 5x3 cm in size respectively, measured about 25 to 34 HU on non-contrast enhanced images. On enhanced images, these masses were inhomogeneously enhanced to about 73 to 79 HU.

The symptoms were not improved, so the patient was referred to King Chulalongkorn Memorial Hospital. At presentation, the patient looked chronically ill without cushingnoid appearance. The body temperature was about 39°c, whereas the pulse rate, respiratory rate and blood pressure were normal. No lymphadenopathy, hepato-splenomegaly or abdominal mass was detected. Routine laboratory tests were within normal limits.

NIDDM = Non-insulin dependant Diabetes Mellitus.

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Bone marrow aspiration was performed on the 3rd day of admission and the result was negative for hypercellular bone marrow. Unsatisfied specimens were obtained from 2 times of fine needle aspiration of the adrenal glands so the opened biopsy at the right adrenal gland was performed on the 20th day of admission and the histological examination revealed large cell type of malignant lymphoma by immunostaining for LCA (Leukocyte Common Antigens and Vimentin).

After initiation of chemotherapy, the patient developed febrile neutropenia and sepsis and was complicated by pneumonia on the 35th day of hospital course. The patient expired 3 days later.

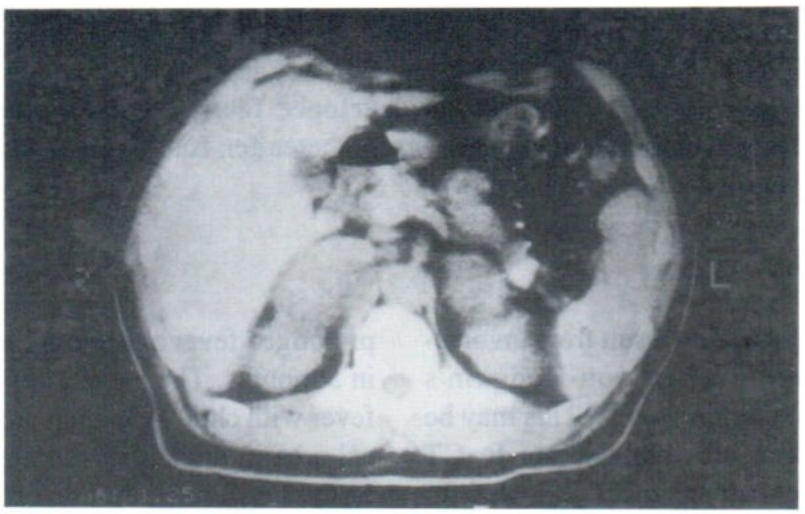

Fig. 1. Unenhanced CT scan shows bilateral adrenal masses. The masses are slightly hypodense than renal parenchyma.

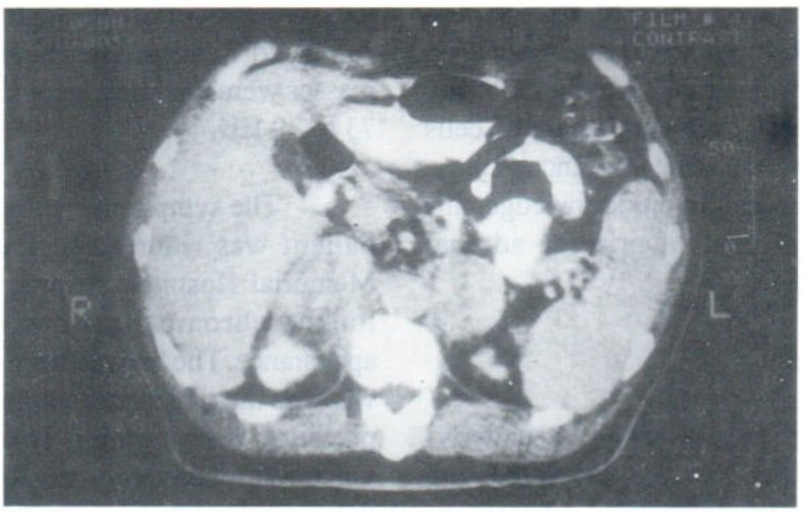

Fig. 2. Constrast-enhanced CT scan shows inhomogeneous enhancement of bilateral adrenal masses.

DISCUSSION

Bilateral adrenal enlargement can be associated with hyperfunction or normal to diminished function of the gland. Hyperplasia and pheochromocytoma are causes of enlargement with hyperfunction. If the function of the gland is normal or diminished, common causes are granulomatous infection (tuberculosis and histoplasmosis), metastasis and hemorrhage.[9] Metastasis to the adrenal glands are most common from lung and breast cancer.[1]

Primary lymphoma of the adrenal gland is rare. Since lymphoma arises from hematopoietic tissue which may be present in the adrenal glands, primary lymphoma arising in the adrenal gland can be expected.[7] The presenting symptoms are non-specific which may include fever, anorexia, malaise and a palpable abdominal mass. Over 70 cases were reported in the literature over the past 40 years and bilateral primary adrenal lymphoma appears to predominate over unilateral.[8,10-12] The definite diagnosis of adrenal lymphoma is by electron microscopic examination with immu-nostaining techniques. Review of the literature indicates diffuse histiocytic lymphoma to be the most common histology of primary adrenal lymphoma.[1-4, 7]

There is no pathognomonic appearance on the imaging findings. Neither ultrasonography, CT scan nor Magnetic Resonance Imaging (MRI) can indicate lymphomatous involvement of the adrenal gland. The sonographic appearance of primary adrenal lymphoma may vary from anechoic or hypoechoic to mixed hypoechoic and hyperechoic lesions. The hypoechoic variety is the most common pattern. Variable sonographic appearances is probably due to the presence of hemorrhage or necrosis.[7]

Using CT, adrenal lymphomas usually are seen as large soft tissue masses (46 to 60 HU) replacing the adrenal glands. They usually alter the shape of the adrenal glands, but the glands may markedly expand while retaining a somewhat adrenaliform shape.[1] The growth pattern sometimes can suggest lymphoma, as it is more likely to infiltrate or insinuate around the upper pole of the kidney rather than displace it, as would be typical for carcinoma.[1] Mild to moderate enhancement is seen after intravenous administration of iodinated contrast. The primary adrenal lymphoma tends to have necrosis and hemorrhage which may cause a cystic appearance on computed tomography.[7] However, CT appearances of adrenal enlargement due to lymphomas are non-specific and may mimicked by metastases.[13] In general, calcification in untreated lymphoma is uncommon, but a case of primary adrenal lymphoma with CT appearace of a cystic mass with multiple punctate calcifications has been reported.

MR imaging has considerable promise in the evaluation of adrenal glands.[14] In some cases, such as pheochromocytoma and adrenal adenoma, MRI helps to increase imaging accuracy by using the signal characteristics of the masses.[1,15] MRI is better than CT to demonstrate IVC patency and periadrenal blood vessels can be easily distinguished from adrenal nodules.[14,15] However, MR appearance of adrenal lymphomas are in distinguishable from other malignancies.[1] They are usually heterogeneous, with low signal on T1-weighted images (less intense than normal liver, but more intense than muscle) and more intense than fat on T2-weighted images. Kato et al. described a case of primary non-Hodgkin's B-cell lymphoma of diffuse large cell type at both adrenal glands, which MRI revealed bilateral adrenal masses with some enhancing septa.[16]

In conclusion, primary adrenal lymphoma is a rare disease, which has no specific appearance on sonography, CT nor MRI. However, in a

patient with unilateral or bilateral adrenal enlargement even in the absence of lymphadenopathy, primary adrenal lymphoma should be included in the differential diagnosis.

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