Abstract

Primary adrenal lymphoma is extremely rare. We report a young patient who presented with non-specific symptoms of fever and abdominal pain. Conventional imaging modalities demonstrated bilateral bulky adrenal masses, and whole-body fluorine-18-fluorodesoxyglucose (18F-FDG) positron emission tomography/computed tomography showed intense 18F-FDG-avid bilateral adrenal masses with no evidence of extra-adrenal spread. A pathological diagnosis of non-Hodgkin lymphoma of peripheral T-cell type was made. The present case indicates that primary adrenal lymphoma should be included in the differential diagnosis of bilateral adrenal masses.

Introduction

Adrenal lymphoma is often bilateral and in most of the cases B-cell type. Adrenal lymphoma of T-cell type is exceptionally rare [1]. The prognosis is always bad and the patient can die early because of acute adrenal insufficiency. We report here a rare case of primary bilateral adrenal T-cell lymphoma in which whole body fluorine-18-fluorodesoxyglucose positron emission tomography/computed tomography (18F-FDG-PET/CT) played an important role in the diagnosis.

Case report

A 33 years old male presented with history of abdominal pain, fever and loss of weight, around 15kg in one year. Ultrasound of the abdomen revealed enlarged bilateral suprarrenal masses. However, the patient did not have any signs or symptoms of adrenal insufficiency. Mantoux test was negative, excluding the possibility of tuberculosis. Whole body 18F-FDG PET/CT (Fig. 1) showed intense 18F-FDG uptake in the heterogeneously enhancing bilateral adrenal masses with right adrenal gland measuring 8.3x9.0x6.0cm with SUVmax 13.3 and the left adrenal gland measuring 5.4x7.2x4.0cm with SUVmax 14.1. There was no other abnormal focus of 18F-FDG uptake and a provisional diagnosis of primary adrenal lymphoma was made based upon the previously reported cases of bilateral PAL which presented as enlarged bilateral homogeneous adrenal masses showing increased FDG uptake [1].

Figure 1. MIP (A), coronal CT (B) and coronal fused PET/CT (C) images respectively showing increased 18F-FDG uptake in the enlarged bilateral adrenal masses (arrows), with no other areas of abnormal 18F-FDG uptake. The right adrenal mass is larger than the left. Photopenic area (arrow head) is also seen in the right adrenal mass suggestive of necrotic changes, which was confirmed by histopathological findings.
Subsequent laparoscopic biopsy of the right adrenal showed infiltration by atypical large lymphoid cells (Fig. 2A) which had irregular vesicular nuclei and occasional conspicuous nucleoli. No granulomas or any fungal profiles including histoplasma were seen. Acid fast bacteria stain (AFB) and Periodic acid-Schiff (PAS) stain did not reveal any acid fast bacilli or any fungal profiles or histoplasma. The atypical lymphoid cells were positive for CD3 on immunohistochemistry (Fig. 2B); indicating a T-cell lymphoma. A few reactive CD20+ B-cells were also seen among the lymphoid infiltrate. The patient has been treated with 3 cycles of cyclophosphamide, hydroxy urea, Oncovin and prednisolone chemotherapy and become symptom-free. Now, he is under follow-up with ongoing chemotherapy and has also gained weight around 5kg.

**Discussion**

Disseminated nodal non-Hodgkin’s lymphoma can involve adrenals in 25% of the cases [1]. However, a primary adrenal lymphoma is a rare entity. A review of literature has documented 70 reported cases of primary adrenal lymphoma out of which 56 had bilateral adrenal involvement, with only 5 cases being T-cell type [1]. A diffuse large B-cell lymphoma is the commonest primary adrenal lymphoma. 18F-FDG-PET/CT is a valuable technique for the diagnosis, staging, and evaluation of the response to treatment of adrenal lymphoma [1]. It has shown encouraging results in differentiating benign from malignant adrenal masses [2]. Primary adrenal T-cell lymphoma has also been demonstrated by 18F-FDG-PET [3]. Primary adrenal non-Hodgkin’s lymphoma commonly occurs in older age group, usually above 70 years and most of these cases present with features of adrenal insufficiency. In a review of six patients with primary adrenal lymphoma of T-cell type, only one who had no clinical hypoadrenalism had survived beyond 8 years [4]. He was treated with right adrenalectomy and radiotherapy. The rest of the patients in that review had expired within 4 months of diagnosis [4, 5].

In conclusion, unlike previous cases reported in the literature, our patient with bilateral adrenal involvement by T-cell lymphoma presented with non-specific symptoms at an early age of 33 years. 18F-FDG-PET/CT had also been reported to demonstrate metabolic response to chemotherapy in such patients [1].

**Bibliography**