Large adrenal leiomyoma presented as adrenal incidentaloma in an AIDS patient: A rare entity

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SUMMARY

The literature on adrenal gland tumour in HIV-infected patients is scarce. We report a 46-year-old Malay man with HIV and Hepatitis C infection presenting with a large non-functioning adrenal tumour. Computed tomography showed a large right adrenal tumour with heterogeneous enhancement and central necrosis. A high index of suspicion of a malignant tumour or pheochromocytoma led us to surgical removal of the adrenal gland. In this case report, we highlight important features to look for during pre-op evaluation of a large adrenal mass. Appropriate action should be taken when there is a suspicion of a pheochromocytoma or malignancy.

KEY WORDS:
Adrenal incidentaloma, adrenal leiomyoma, HIV infected patient

INTRODUCTION

Due to the introduction of highly active combination antiretroviral therapy (HAART), some cancers which were previously not associated with HIV appears to be increasing in incidence due to the longer survival of these patients. These are collectively termed as non–AIDS-defining cancers (NADCs). It is rare to see a case of adrenal tumour in HIV-infected patient, though the incidence was reported to be 3% in a series of autopsy.[1]

CASE REPORT

We present a 46-year-old Malay man, an intravenous drug user, diagnosed in the year 2010 with HIV and hepatitis C infection when he first presented with weight loss and lethargy. Subsequently, he had multiple opportunistic infections which include syphilis, pneumocystis carini, oral candidiasis and varicella zoster. He was started on second line HAART regimen and then on a salvage regimen with Tenvir (Emtricitabine and Tenofovir) and Kaletra (Lopinavir and Ritonavir). In May 2013, he had an incidental adrenal mass detected by a computed tomography, when imaging was planned for hepatoma screening due to his hepatitis status.

Physical examination revealed a thin built man with normal blood pressure. There were no episodic headache, sweating, palpitation, cushingoid features or diabetes mellitus. Abdominal examination revealed a ballotable mass over the right upper quadrant of the abdomen. Laboratory investigation revealed immune deficient state with low CD4 count and no hypokalemia or evidence of excessive adrenal hormone production (Table I). Renin and aldosterone level was not taken by the internist as the patient was normotensive with normal potassium levels. The level of morning serum cortisol was very low which can be due to the replacement of normal adrenal tissue with the tumour (Table I).

Computed tomography revealed a well-defined mass at right suprarenal region measuring 70 mm x 79 mm x 88 mm (AP x W x CC) with a clear plane to the surrounding structures (Figure 1). The mass showed heterogeneous enhancement on portovenous phase with areas of necrosis. The periphery of the lesion measured 45 HUs pre-contrast. The centre of the mass was approximately 5 HUs. These features suggested the possibility of an infectious aetiology, malignancy or pheochromocytoma. He was scheduled for surgery to remove the tumour via right subcostal incision. The adrenal tumour was handled with great caution as there was a suspicion of pheochromocytoma. An arterial line was inserted at induction by the anaesthetist for intraoperative arterial blood pressure monitoring, and labetalol infusion was kept on standby. Adrenal veins were cauterised with ligasure and clipped with liga clips during surgery. The right adrenal gland was mobilised and the adrenal tumour was removed. Intraoperative blood loss was 300 mL. No surge of blood pressure noted intraoperatively. Post-operative recovery was uneventful and he was discharged on post-op day three. Subsequent follow up until one year, he was well with a healed scar over the right upper abdomen.

Pathology

Grossly, the right adrenal gland consists of a well circumscribed, lobulated mass measuring 95x60x45 mm. The external surface was smooth and cut section showed cream coloured mass with an area of mucoid change. Histological examination revealed a fairly circumscribed, encapsulated lobulated lesion consisting of highly cellular lobules alternating with hypocellular myxoid and hyalinized areas. The lobules showed spindle cells proliferation arranged in haphazard fascicles with foci of concentrically around blood vessels (Figure 2). The spindle cells have ovoid, plump or irregularly shaped vesicular, moderately pleomorphic nuclei with occasional small nucleoli. Mitotic activities was rarely seen (1/10 HPF, Olympus BX43). These spindle cells are diffusely positive for Desmin and SMA but negative for other immunostains panCK, S-100, Myogenin, CD117, ALK-1 and CD34.

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Table I: Serial anterior pituitary hormonal assays of the patient

	Value	Normal Range
White Cell Count	5.0 x10^3/UL	(4.0 -12) x10^3/UL
Hemoglobin Level	14.4 g/dL	(13.0 -17.0) g/dL
Platelet Count	191 x10^3/UL	(150 - 400) x10^3/UL
Serum Urea	4.3 mmol/L	(2.5 -7.1) mmol/L
Creatinine	87 µmol/L	(80 -115) µmol/L
Sodium	137 mmol/L	(135 -145) mmol/L
Potassium	4.1 mmol/L	(3.5 -5.0) mmol/L
Corrected Calcium	2.20 mmol/L	(2.15 -2.55) mmol/L
Total Bilirubin	10µmol/L	(3 -25) µmol/L
Albumin	36g/L	(35 -50) g/L
Alkaline Phosphatase	79 u/L	(44 -100) u/L
Alanine Transaminase	34 u/L	(10 -40) u/L
24hr Urinary Catecholamine[a]		
Norepinephrine	62.1µg/day	(12.1 -85.5) µg/day
Epinephrine	2 µg/day	(1.7 -22.4) µg/day
Dopamine	422 µg/day	(<498) µg/day
AM Serum Cortisol[a]	0.412 µg/dl	(7-25) µg/dl
CD4 Count[b]	88 cells /mm^3	(500-1500)cells /mm^3
Viral Load	12894 c/ml	

[a] Biochemical test done for adrenal function reflects a non- functioning gland.
[b] Low CD4 count suggestive of an immunodeficient state

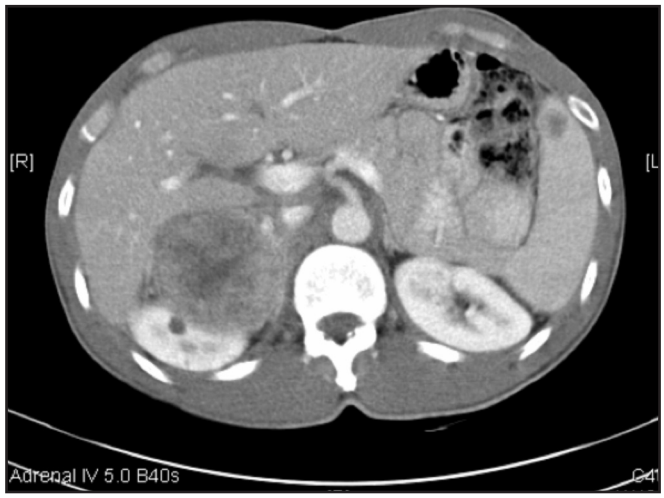

Fig. 1: Computed tomographic scan of the abdomen, showing right adrenal mass compressed the right kidney.

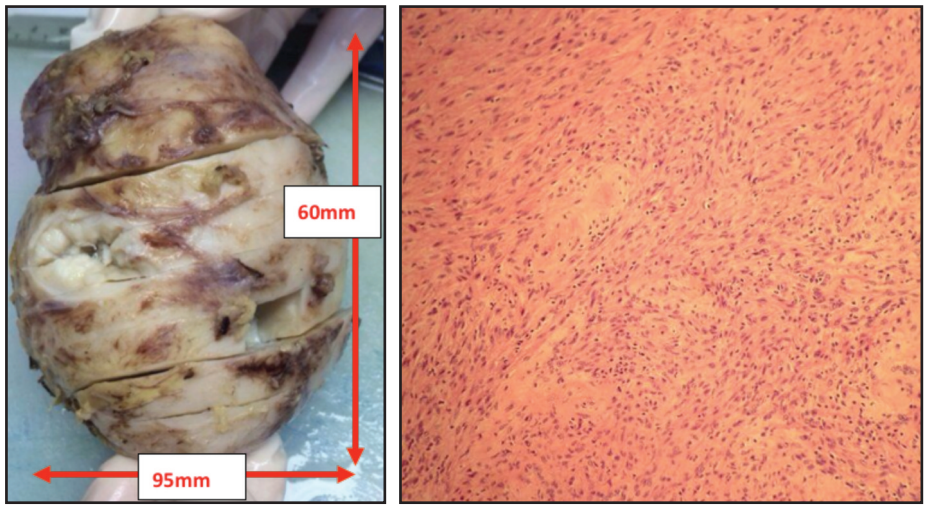

Fig. 2: Gross and microscopic findings of the removed specimen
(Left) - Right adrenal tumour removed (After soaked in formalin and sliced)
(Right) - Spindles cells arranged in haphazard fascicles. (Hematoxylin-eosin stain; x10 magnification)

DISCUSSION

Adrenal leiomyoma is a rare solid tumour of unknown aetiology. Reported cases in the literature were less than 20.2 The age of presentation varied from 2 to 72 years, and the young patients tend to develop bilateral adrenal gland tumour.[2,3] About two third of the cases occur in female and largest diameter reported ranged between 3 to 11 cm.[2] In this case, the patient had HIV infection which is also seen in eight out of 18 cases in another review.[2]

Adrenal mass in HIV-infected patients can have an infectious aetiology such as Cytomegalovirus necrotising adrenalitis, Mycobacterium or Cryptococcus.[3] But the likelihood of infective aetiology in the current case was excluded by infectious disease physician evidenced by the patients' good clinical condition and normal cell counts.

Computed tomography features to diagnose pheochromocytoma or adrenal cancer include size (>3 cm), attenuation of >10 HU on unenhanced CT, heterogeneous texture and increased vascularity with decreased contrast washout at 10 to 15 minutes.[4] In our case, these features were present which led to the surgical removal of the adrenal tumour.

In the presence of a large adrenal tumour, a normal level of urinary catecholamines is insufficient to rule out a pheochromocytoma as most of it is metabolised within the tumour to metanephrines. A collection of urinary or plasma free metanephrine would have been best, but the availability of these tests is limited at our institution. As such, we proceeded with extra caution during surgery.

CONCLUSION

Open surgery with a right subcostal incision to remove a large adrenal leiomyoma is safe even in an immune-deficient HIV-infected patient. To our knowledge, this is the first case being reported in Malaysia.

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