Unusual association of diseases/symptoms

Vanishing adrenal mass

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Adrenal involvement by primary lymphomatous disorders is quite rare. Primary adrenal lymphoma is usually associated with bilateral adrenal involvement and presents as adrenal insufficiency or class B symptoms. The authors report a case who presented with pain in the abdomen and was diagnosed to have unilateral primary adrenal lymphoma. The patient responded to chemotherapy with near-complete resolution of the adrenal mass.

BACKGROUND

Adrenal gland(s) involvement by the systemic lymphomatous disease in non-Hodgkin's lymphoma has been reported in 25–35% of patients.¹ However, primary adrenal lymphoma (PAL) is quite rare and so far only 70 cases and mostly bilateral, have been reported in the English literature.² Class B symptoms and adrenal insufficiency are the main presenting manifestations of PAL. Rarely, it may present with a fever of unknown origin, hypocalcaemia or may be diagnosed incidentally on abdominal imaging.³ With appropriate diagnosis and treatment, the abnormally enlarged gland(s) may eventually regress to normal size with or without alterations in functional status. We report a case of unilateral adrenal mass diagnosed to have PAL and he responded well to chemotherapy.

CASE PRESENTATION

An 80-year-old man, known case of diabetes and hypertension for past 15 years presented with ill-defined left-sided abdominal pain of 3 months duration. The pain was dull aching and had no relation to meals without any alterations in bowel and bladder habits. He had no history of paroxysms.

On examination, his body mass index was 28.2 kg/m² and had BP of 150/90 mm Hg. He had evidence of peripheral neuropathy and bilateral cataract. There were no stigmata of protein catabolism, or any café-au-lait macules. The abdominal examination revealed a bimanually palpable vague non-tender mass in the left lumbar and iliac regions. Other systemic examination was normal.

On investigation, haemoglobin was 10.2 gm/dl; serum potassium was 4.2 meq/l and normal renal and liver function tests. Serum cortisol at 08:00 h and at 16:00 h was 840 nmol/l (N: 400–600) and 600 nmol/l (200–400) respectively; and was non-suppressible after 1 mg dexamethasone (640 nmol/l N:<50 nmol/l). Urinary metanephrines was within normal range. Serum 17 α hydroxyprogesterone (17 α OHP) was 2 ng/ml (N: 0.5–2). Ultrasonography of the abdomen showed a suprarenal mass on the left side and multiple cysts in both kidneys. CT scan showed a lobulated, heterogenous left adrenal mass measuring

12×11×7 cm encasing aorta and left renal vessels (figure 1). The other adrenal gland was normal.

In view of the size of the adrenal mass (>5 cm) with intralesional necrosis and vascular involvement, patient was planned for surgery considering a diagnosis of adrenocortical carcinoma. But he refused to undergo surgery therefore, an ultrasound guided fine needle aspiration cytology (FNAC) from the adrenal mass was performed to establish the cytological diagnosis which showed discrete populations of monomorphic lymphoid cells, round to oval with scanty blue cytoplasm and some of the cells having cytoplasmic vacuolations. The cells were CD 20 positive and CD 3 negative. A diagnosis of high-grade B cell non-Hodgkin's lymphoma was made (figure 2). Bone marrow biopsy from the iliac crest was normal. Serum lactate dehydrogenase (LDH) was 556 U/l (normal: 180–248) and β 2 microglobulin was 7593 μ g/l (670–1310). Cerebrospinal fluid analysis was unremarkable.⁶⁷ Gallium scan did not show uptake anywhere including adrenal mass. After establishing the diagnosis of adrenal lymphoma, patient was treated with cyclophosphamide, adriamycin, vincristine (oncovin) and prednisolone (CHOP) chemotherapy protocol at monthly intervals for eight consecutive cycles with near-complete resolution of the adrenal mass (figure 3). Serum LDH and β 2 microglobulin decreased considerably. The cortisol response to 250 μ g ACTH (Synacthen) a month after the last cycle was 485 nmol/l (N-500 nmol/l) at 30 min, suggestive of decreased adrenal reserve. During follow-up for a period of almost 5 years, there is no recurrence of disease as evidenced by normal level of β 2 microglobulin and no ¹⁸FDG tracer uptake.

DISCUSSION

PAL is a rare disorder.² Men outscore women (2:1–7:1 in different studies) and commonly present during sixth decade and onwards,^{1–4} as was in our case. Class B symptoms and features of adrenal insufficiency are the main presenting manifestations and there is usually bilateral adrenal involvement.^{5–6} However, our patient had neither of them and diagnosis of unilateral PAL was a histological surprise.

Figure 1 Contrast enhanced CT adrenal showing a heterogenous lobulated left adrenal mass encasing aorta. Right adrenal is normal.

Figure 2 Fine needle aspiration cytology from the adrenal mass showing infiltration by non-Hodgkin's lymphomatous cells (MGG×540).

The differential diagnosis for patients with unilateral adrenal incidentaloma include adrenocortical neoplasia both benign and malignant (size >5 cm), pheochromocytoma, granulomatous disorders like tuberculosis and histoplasmosis, adrenal cyst, myelolipoma and rarely oncocytoma. For bilateral adrenal involvement, the usual differential diagnoses are metastatic deposits, bilateral pheochromocytoma, granulomatous disorders and rarely adrenal myelolipoma and adrenocortical carcinoma. The minimum work-up required for adrenal incidentaloma

include overnight dexamethasone suppression test, urinary metanephrines and serum potassium. However, if the patient is hypertensive and/or has hypokalemia, serum aldosterone and plasma rennin activity is to be assessed.

FNAC is usually not indicated for establishing the aetiological diagnosis of adrenal incidentaloma unless there is a possibility of granulomatous disorder or metastatic adrenal involvement. Patients with granulomatous disorders usually present with fever, weight loss and adrenal

Figure 3 Contrast enhanced CT adrenal showing near complete resolution of the adrenal mass after completion of chemotherapy.

insufficiency, while in metastatic adrenal involvement, presence of primary malignancy elsewhere is usually evident. Therefore, the main contender for the differential diagnosis in this case was adrenocortical carcinoma in view of the adrenal mass of >5 cm, intralesional necrosis and vascular encasement. However, typical histology, positive immunocytochemistry with CD20, a B cell marker and raised levels of LDH and β_2 microglobulin substantiated the diagnosis of adrenal lymphoma.

The present case had non-suppressible cortisol level after overnight dexamethasone suppression test, which was consonant with the presence of adrenocortical carcinoma associated with subclinical Cushing syndrome. PAL is usually present with adrenal insufficiency as there is usually bilateral adrenal involvement and serum cortisol at 08:00 h is likely to be suppressed after overnight dexamethasone. Our case had unilateral adrenal involvement and in retrospect, the non-suppressibility of cortisol dynamics in the presence of adrenal lymphoma can be explained by ongoing stress and/or increased glucocorticoid release in response to ongoing infiltrative process.

The diagnosis of PAL was considered in view of absence of superficial or retroperitoneal nodal involvement at presentation, no other organ involvement including bone marrow and lack of uptake of ^{67}Ga tracer anywhere. Lack of ^{67}Ga tracer uptake by adrenal lymphomatous lesion is possibly related to dedifferentiated lymphomatous cells devoid of tracer receptors. The involvement of the other adrenal gland by lymphomatous process is unlikely in view of preserved shape of the other adrenal gland, normal adrenal reserve and probably a somatic mutation, being confined to the involved adrenal gland.

The origin of lymphomatous process in the adrenal gland is akin to adrenal myelolipoma. As primitive totipotent mesenchymal cells in the adrenal gland can undergo differentiation to B cell or T cell under stress,

infection or some other undefined process.³ The common subtype is diffuse large B cell type, as was in our case. However, T cell angiocentric large cell lymphoma type B are exceptional.⁸

The therapeutic modalities include combination chemotherapy, followed by radiotherapy in order to prevent local recurrence. Surgical debulking is not recommended in view of high morbidity and mortality and recurrence of the disease elsewhere later. Our patient received eight cycles of CHOP based chemotherapy at monthly intervals and responded well. Old age, initial presentation with adrenal insufficiency, large tumour burden, elevated serum LDH and β_2 microglobulin and involvement of the other organs during follow-up indicate poor prognosis.^{8,9}

In conclusion, in older patients with unilateral adrenal mass, fine needle cytology may be rewarding.

Learning points

- FNA may be rewarding in cases with unilateral adrenal mass particularly in the older patients.

Competing interests None.

Patient consent Obtained.

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