Laparoscope resection of ectopic corticosteroid-secreting adrenal adenoma

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Abstract

Tumors originating from ectopic adrenal tissue are relatively rare. In this article, we describe a case with Cushing's syndrome caused by an ectopic adrenal adenoma. A 38 year-old male patient presenting with cushingoid appearance for 2 years was diagnosed to have ACTH-independent Cushing's syndrome based on endocrinological evaluation. Mutiple radiological examinations detected bilateral adrenal atrophy. When the images were investigated in a more expanded scope, a 3.0×3.5×5.3 cm mass was detected in the anterior of left renal hilum and left renal vein. The mass was successfully resected with intraoperative endoscopy and pathological evaluation revealed an ectopic adrenal tumor. It is suggested that when the endocrinlogically confirmed adrenal neoplasm could not be well and definitely localized, the possibility of ectopic adrenal should be presumed and further radiography examinations should extend to the field where ectopic adrenal usually presents.

INTRODUCTION

Ectopic adrenal tissues are usually present close to the adrenal glands, along the path of descent or in association with the gonads due to the close spatial relationship between the adrenal and urogenital primordial (Ayala *et al.* 2000). Tumors originating from ectopic adrenal tissue are relatively rare. In this article, we describe such a case with Cushing's syndrome caused by an ectopic adrenal adenoma.

CASE REPORT

A 38 year-old male patient presenting with cushingoid appearance was referred to Endocrinology Department. Since September 2009, he had gradually developed moon facies, hirsutism, bruise easily and weakness. His blood pressure elevated to 140–150/100–110 mmHg, with bad response to anti-hypertensive drugs. Later, he suffered from polydipsia, polyuria, with confirmed persistent hyperglycemia. The blood glucose could not got be well controlled with insulin Aspart 30 (54 IU per day). In March 2011, compression fractures in lumbar 2,3 occurred when he raised something only slightly heavy.

On arrival, his blood pressure was 144/114 mmHg. He presented with typical Cushing's syndrome, including moon facies, a moderate cervicodorsal fat pad, bilateral supraclavicular fullness, and truncal obesity. Multiple purple striae and ecchymoses were present over the lower abdomen and thighs.

Biochemical examination showed persistent hypokalemia and metabolic alkalosis. Even repeated administration of chloratum Kalium

CASE REPORT

for 4.5 g/d, the serum kalium level was still lower than normal range (2.70–3.31 mmol/l). HbA1c level was much higher (9.1%).

Endocrinological evaluation showed much higher levels of urinary free cortisol (5319.3 nmol/24h, reference range (RR) 98–500.1 nmol/24h) and serum free cortisone (731.52 mmol/L, RR 198.7–797.5 mmol/L at 8:00 am). Dynamic endocrine tests revealed altered circadian rhythm of cortisol with complete suppression of plasma ACTH (<1.1 pmol/L at 8:00 am and 16:00 pm, RR 2.2–10.12 pmol/L). High dose dexamethasone suppression test showed serum cortisol was not suppressed (878.5 mmol/L) at all.

Collectively, these clinical features were consistent with the diagnosis of ACTH-independent Cushing's syndrome. Subsequently, ultrasononic examination, as well as computed tomography (CT) scan and magnetic resonance imaging (MRI) scan were performed. However, all these 3 assays only detected bilateral adrenal atrophy, without tumors or nodules in adrenals (Figure 1). When the images were investigated in a more expanded scope, a 3.0×3.5×5.3 cm mass displayed in the anterior of left renal hilum and left renal vein, with abundant fatty component and plentiful vascular supply (Figure 2).

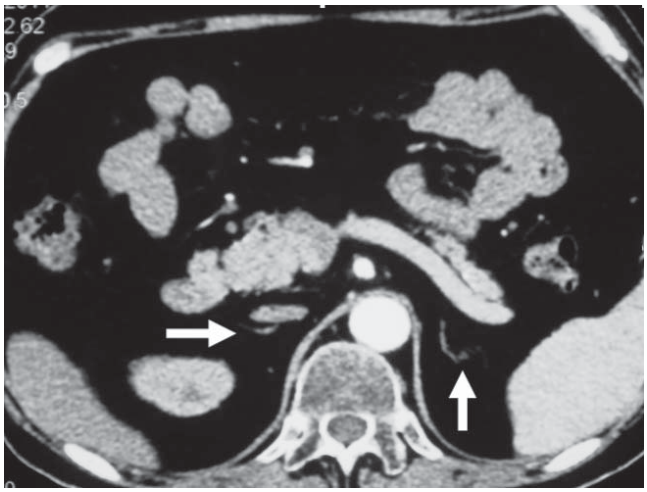

Fig. 1. Abdomen CT scan detecting bilateral adrenal atrophy, without tumors or nodules in adrenals.

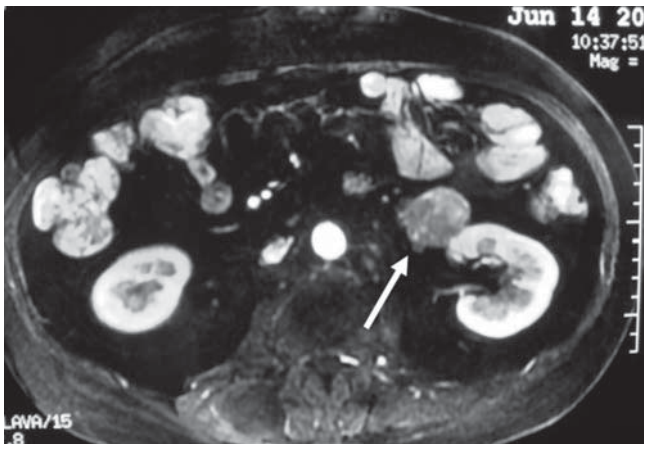

Fig. 2. Abdomen MRI scan detecting a 3.0×3.5×5.3cm tumor in the anterior of left renal hilum and left renal vein, with abundant fatty component and plentiful vascular supply.

Taken together, the endocrinological and radiological data were highly suggestive of ACTH-independent Cushing's syndrome due to an ectopic adrenal adenoma.

Later, this patient underwent laparoscopic endoscopic approach. The laparoscopic wedge resection of the tumor was successfully performed using the Endo-GIA roticulator. The resected tumor measured 4.0×3.5×3.0 cm, with yellow brown cut surface. Microscopical examination revealed the tumor cells arranged in nests and trabeculae.

Serum cortisone level decreased to 474.5 mmol/L abruptly in the second hour after operation. He was then administrated with hydrocortisone intravenous for 5 days to prevent adrenal crisis, and later transferred into prednisone 5 mg at 8:00 am and 2.5 mg at 16:00 pm for supplement. The postoperative course was uneventful. Drug-resistant hypertension and hypokalemia were resolved and hyperglycemia was well controlled with the same dose of insulin as before. In the follow-up, the cushingoid manifestation disappeared gradually.

DISSCUSION

The adrenal gland arises from primordial mesenchyme in the wall of the dorsal coelom adjacent to the dorsal mesentery and urogenital structures (Ayala *et al.* 2000; Makino *et al.* 2010). The unusual location of ectopic tissue may be related to misplaced mesothelial cells or autonomous differentiation of mesodermal elements. As a result, most ectopic adrenocortical tissue is found along the path of embryonic migration within the urogenital tract, including celiac axis (32%), broad ligament (23%), adnexa of testis (7.5%), and spermatic cord (3–8%) (Ayala *et al.* 2000; Iyengar & Pittman 2007; Yoon *et al.* 2010).

Ectopic adrenal tissue is estimated to occur in about 1% of the adult population and up to 50% of neonates. With advancing age, this accessory tissue atrophies because it is physiologically unnecessary in the presence of normal adrenal glands (Ketata *et al.* 2008; Louiset *et al.* 2010). Ectopic adrenal tissue may contain both cortex and medulla, if the breaking event occurs after migration of neural (Ye *et al.* 2009; Rodriguez *et al.* 2009).

Adrenocortical tumors, both benign and malignant, can also arise at ectopic sites. The vast majority of the cases reported are non-secreting ectopic adrenal tumors. Ectopic adrenal tissue becomes symptomatic if it over-secrets hormones such as aldosterone, cortisol, or androgens, which can lead to the presentation of Cushing's syndrome, aldosteronoma, Conn's syndrome or extreme virilization (Louiset *et al.* 2010; Mavroudis *et al.* 2007). Primary cortisol-secreting tumors of the

adrenal glands account for the final 15% of cases with Cushing's syndrome. The case with Cushing's syndrome caused by ectopic adrenal adenoma is rarely reported.

As for this case, initially presented with Cushing's syndrome, endocrine evaluation indicated the diagnosis of ACTH-independent Cushing's syndrome caused by adrenal tumor or nodular hyperplasia. Unexpectedly, only adrenal atrophy but no adrenal tumor or nodular hyperplasia was detected. When the detected scope was expanded, the presumed offending tumor was localized in the anterior of left renal hilum and left renal vein. The ectopic tumor was removed and serum free cortisone reduced significantly after operation. The pathology and immunohistological examination confirmed our initial judgment. To our best knowledge, this is the first report on successful laparoscope resection of ectopic adrenal adenoma.

In fact, the ectopic endocrine glands such as thyroid, parathyroid, and pheocromacytoma may be encountered sporadically. When an active endocrine gland tumor or hyperplasia was confirmed according to biochemical and endocrinological examination, both usual location and unusual ectopic location of endocrine gland should be detected thoroughly with radiological assays. It may be helpful for reaching the correct preoperative diagnosis of adenoma originating from an ectopic endocrine gland, thereby possibly averting aggressive surgery.

CONCLUSION

In summary, we described a rare case with Cushing's syndrome caused by ectopic adrenal cortisol-secreting adrenal adenoma, who underwent successful laparoscope operation. We would like to emphasize that the possibility of ectopic adrenal adenoma should be presumed when endocrinologically confirmed adrenal adenoma could not be detected in the usual adrenal location. Also, the radiography examinations should extend to the field of ectopic adrenal. Laparoscope resection of ectopic adrenal tumor seems to be a feasible and effective approach (Lehrfeld *et al.* 2010).

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