Primary Hyperaldosteronism Induced by Aldosterone-Producing Adenoma Co-Existing with A Left Suprarenal Accessory Spleen: Two Case Reports and A Review of The Literature

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We encountered 2 patients (a 33-year-old woman and a 66-year-old man) with an aldosterone-producing adenoma (APA) and a left accessory spleen. The patients' primary symptoms were hypertension and hypokalemia, and both had elevated serum aldosterone levels. Preoperative computed tomography a left suprarenal retroperitoneal mass and laparoscopic left adrenalectomy was performed in both cases. The postoperative microscopic examination revealed splenic tissue. Both patients experienced relief of their hypertension and hypokalemia, with an uneventful recovery.

Keywords: adrenal adenoma; hyperaldosteronism; accessory spleen

INTRODUCTION

Ectopic splenic tissue can exist in two forms: an accessory spleen or splenosis. An accessory spleen is normal splenic tissue that has evolved from embryological remnants of splenic tissue[(1)], and is typically asymptomatic, although it may grow to the size of a normal spleen after splenectomy is performed[(2)]. The co-existence of an aldosterone-producing adenoma (APA) with a left accessory spleen would be difficult to diagnose, and we are not aware of any reported cases of primary hyperaldosteronism (PHA) that were induced by APA and a left accessory spleen. Thus, we report our experience with two cases of APA and a left accessory spleen that was identified after laparoscopic adrenalectomy.

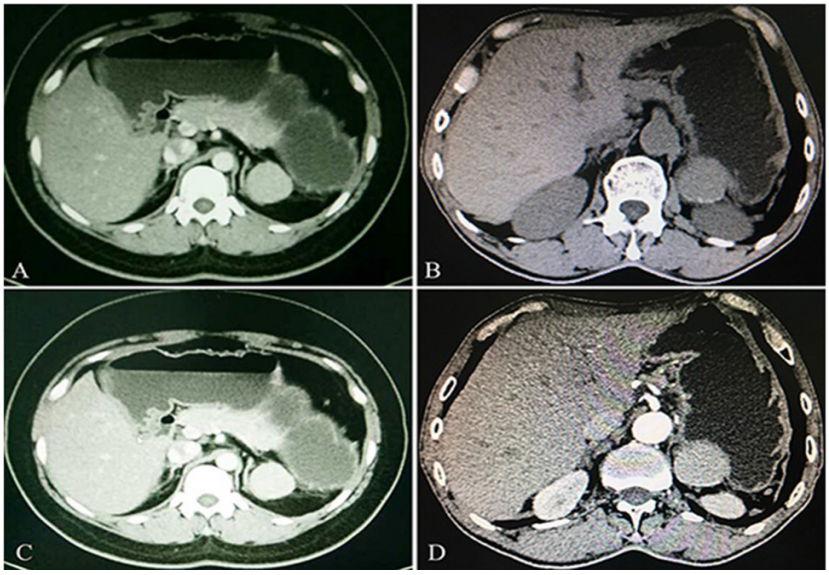

Figure 1. Preoperative abdominal computed tomography of case 1 (A, C) and case 2(B, D).

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Table 1. Basic information of the 2 patients.

	Case1	Case2	Normal Value
Gender	Female	Male	-
Age	33	66	-
Splenectomy	Yes	Yes	-
Blood pressure(mmHg)	170/110	181/119	140-90/90-60
Serum potassium(mmol/L)	3.16	2.98	3.5-5.5
Mass location	Left suprarenal	Left suprarenal	-
Mass size(cm*cm)	4*4	4*3	-
Serum aldosterone(pg/ml)	158.65	202.96	45-175
Serum renin(pg/ml/h)	0.19	2.65	0.55 ± 0.09
Serum angiotensin(pg/ml)	87.49	90.36	26.0 ± 1.9
Serum cortisol(ng/ml)	166.23	175.83	66-286
Serum ACTH(pg/ml)	25.9	175.83	5-50
Urine aldosterone(ug/24h)	3.5	27.8	2.0-13.3
Urine cortisol(ug/24h)	69.65	60.53	36-455

CASE REPORT

Case 1

A 33-year-old woman (**Table 1**) had previously undergone splenectomy and was subsequently referred to our hospital because of hypertension (170/110 mmHg) and hypokalemia (3.16 mmol/L). Abdominal computed tomography (CT) revealed a left retroperitoneal suprarenal mass with approximate dimensions of 4 × 3.5 cm (Figure 1). Laboratory testing revealed a serum aldosterone level of 158.65 pg/mL, a renin level of 0.19 ng/mL/h, an angiotensin level of 1,187.49 pg/mL, a cortisol level of 166.23 ng/mL, an adrenocorticotrophic hormone level of 25.90 pg/mL, a urinary aldosterone level of 3.5 μg/24 h, and a urinary cortisol level of 69.65 μg/24 h. The patient had a pale complexion and finger beds, and received a 1-week treatment using α-receptor blockers (phenoxybenzene) and spironolactone, which controlled her blood pressure, serum potassium level, and heart rate (< 80 beats/min). During this period, the patient did not exhibit paroxysmal hypertension, but developed warm fingers and toes, pink nail beds, and nasal obstruction. Left laparoscopic adrenalectomy was performed under general anesthesia, and revealed that the suprarenal mass had a distinct capsule (**Figures 2,3**). No uncontrollable hypotension was encountered during or after the surgery (blood pressure: 153/102 mmHg), and the patient did not report experiencing postoperative discomfort.

A pathological evaluation revealed that the mass was non-malignant ecchymotic splenic tissue (**Figure 4**). The patient recovered uneventfully and was discharged on postoperative day 5. At the 6-month follow, the patient had normal values for blood pressure, urinary aldosterone, and renin, as well as normal findings from CT and routine blood testing of liver and kidney functions.

Case 2

A 66-year-old man (**Table 1**) presented to our hospital with hypertension (181/119 mmHg) and hypokalemia (2.98 mmol/L). The patient had undergone total splenectomy at the age of 25 years because of a traffic accident. Preoperative CT revealed a 4 × 3 cm mass in the left retroperitoneal suprarenal region (**Figure 1**). Laboratory testing revealed a renin level of 2.65 ng/mL/h, an angiotensin level of 1,190.36 pg/mL, an aldosterone level of 202.96 pg/mL, a cortisol level of 175.83 ng/mL, an adrenocorticotrophic hormone level of 30.32 pg/mL, a urinary aldosterone level of 27.8 μg/24 h, and a urinary cortisol level of 60.53 μg/24 h. The patient had a pale complexion and nail beds, and received a 1-week treatment using phenoxybenzene and spironolactone, which controlled his blood pressure, serum potassium level, and heart rate (< 80 beats/min). The patient also developed warm fingers and toes, pink nail beds, and nasal obstruction. Laparoscopic left adrenalectomy was performed under general anesthesia (**Figures 2,3**), and his blood pressure remained stable during and after the surgery (145/98 mmHg). The postoperative pathological results suggested that the removed mass was accessory spleen tissue (**Figure 4**). The patient recovered well and was subsequently discharged from the hospital. At the 6-month follow-up, the patient had normal values for blood pressure, urinary aldosterone level, and renin level, as well as normal findings from routine blood testing of liver and kidney functions.

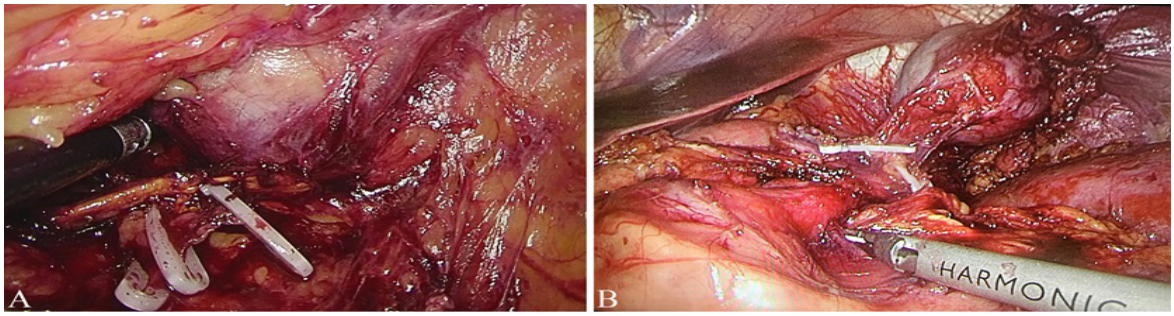

Figure 2. The left adrenal area mass was discovered during laparoscopic surgery (Case 1: A, Case2: B).

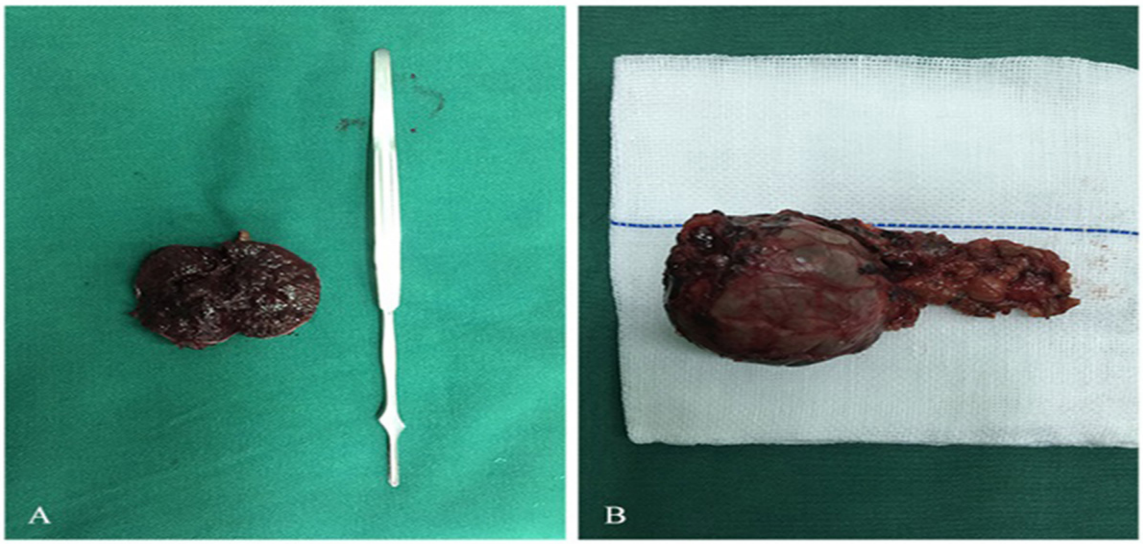

Figure 3. The removed mass of case 1 (A) and case 2(B).

Surgical technique

The retroperitoneal approach was used in Case 1 and the intraperitoneal approach was used in Case 2. In Case 1, a roughly 4 × 3.5 cm round mass was discovered at the left adrenal area. The mass had an intact capsule and was closely attached to the adrenal gland. In Case 2, a mobile round mass was discovered near the left adrenal gland and behind the pancreas. That mass also had an intact capsule but was farther away from the left adrenal, compared to the first mass (**Figure 2**). In both cases, the masses were completely removed and sent for pathological examination. The sectioned surfaces exhibited a homogenous solid structure with a "fish meat" appearance, and no cavity or hematoma was observed within the structure. Thus, the post-operative pathological diagnoses were accessory spleens with chronic congestion.

DISCUSSION

Twenty percent of the population has an accessory spleen, which is supplied by vessels from the splenic hilum and is usually discovered adjacent to the spleen in the peritoneum[3]. Between 1 and 4 wandering accessory spleens can be detected in a patient, and they usually have a diameter of 1–3 cm[4]. The manifestations of these accessory spleens can mimic an adrenal, pancreatic, gastrointestinal, or even testicular tumor. Surgical intervention is not necessary in asymptomatic cases, although laparoscopic resection is recommended if the patient experiences anemia, pain, rupture, or infarction[5].

We are only aware of three reported cases of an accessory spleen in the suprarenal region. Rosenblatt et al.[6] performed laparoscopic adrenalectomy for a 72-year-old woman with a suspected adrenal tumor, although postoperative microscopic examination revealed splenic tissue that included lymphoid follicles. Tsuchiya et al.[7] reported the case of a 66-year-old woman who underwent laparoscopic adrenalectomy because of a suspected non-functional adrenal tumor, although the surgical specimen was not a tumorous lesion. Chen et al.[8] performed laparoscopic resection for a patient with a suspected adrenal tumor, although the pathological report confirmed the diagnosis of an accessory spleen. However, we are not aware of any reports regarding PHA induced by APA and an accessory spleen.

During plain or contrast-enhanced CT, the accessory spleen has the same density as normal splenic tissue[7], and an adrenal mass with a high-intensity T2-weighted signal is generally a malignant tumor. Magnetic resonance angiography could provide more detailed anatomical information to facilitate the diagnosis, and damaged red blood cells during scintigraphy can be used to identify a small asymptomatic accessory spleen.

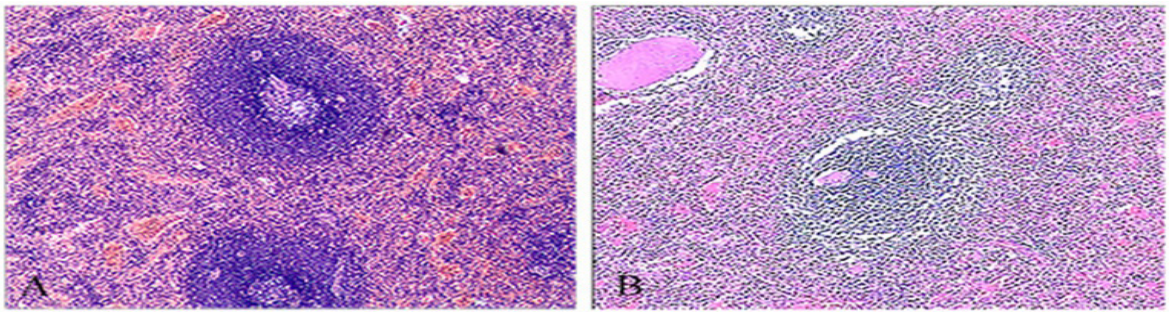

Figure 4. The post-operative pathological diagnoses were accessory spleens with chronic congestion. (Case 1: A, Case2: B)

Table 2.Reports of accessory spleen in the adrenal region.

No.	Reporter	Year	Age	Gender	Location	Size(cm)	Splenectomy History	Chief Complaint	Preoperative Diagnosis	Treatment	Pathology
1	Rosenblatt et al	2010	72	female	left adrenal	5*5	yes	abdominal pain	adrenal tumor	Laparoscopic left adrenalectomy	splenic tissue
2	Tsuchiya et al	2000	66	female	left adrenal	2*2	no	asymptomatic	adrenal tumor	Laparoscopic left adrenalectomy	splenic tissue
3	Chen et al	2005	41	male	left adrenal	7*6	no	asymptomatic	adrenal gland cancer	Laparoscopic left adrenalectomy	splenic tissue

Furthermore, in patients who have undergone splenectomy, the absence of Howell-Jolly bodies and surface indentations on erythrocytes can reveal the immunological and physiological effects of the accessory spleen[9]. Scintigraphy using 99mTc-nanocolloid provides high specificity for confirming the presence of spleen tissue in patients who had experienced splenic trauma.

In the present cases, we performed laparoscopic surgery based on the assumption that the PHA had been induced by APA, but did not consider the possibility of an accessory spleen. This is because the patients' symptoms and serum aldosterone levels fulfilled the PHA diagnostic criteria, which led to our misinterpreting the significance of the masses during the preoperative imaging. Furthermore, the patients' histories of splenectomy made it impossible to compare the accessory spleen to their spleen during the CT. Therefore, clinicians should be aware of the possibility of an accessory spleen when an adrenal mass is detected using conventional imaging techniques. Intraoperative findings of a dark-red mass with a smooth surface may also indicate the presence of an accessory spleen.

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