

·Case Report·

Complete recovery after the removal of an ectopic testicle in a case of primary reninism and retroperitoneal hemangioma

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Abstract

A 32-year-old man recovered completely from hypokalemic hypertension that had been caused by primary reninism after the ablation of an ectopic left testis, epididymis and ductus deferens. For several years, severe hypertension has been resistant to treatment, even the concurrent administration of up to seven antihypertensive agents. In this case, cryptorchidism was associated with an indirect inguinal hernia and an open peritoneo-vaginal process on both sides, aplasia of the posterior wall of the inguinal canal on the right side, an umbilical hernia, and a retroperitoneal tendrilar hemangioma. (*Asian J Androl* 2006 Mar; 8: 247-250)

Keywords: primary reninism; cryptorchidism; peritoneo-vaginal process; Sertoli-cell-only syndrome; hypertension; secondary aldosteronism; hemangioma

1 Case report

A 32-year-old man who was dyspneic and in a state of hyponatremic hyperhydration was admitted to the hospital in November 2001 for an adrenalectomy due to hypokalemic arterial hypertension. For several years, hypertension had been refractory to combinations of up to seven concurrently administered antihypertensive agents. Spironolactone in a dosage of 400 mg/d, administered because of a massive potassium loss, had caused the onset of pulmonary edema. The aim of the adrenalectomy was to reduce the aldosterone antagonist to a

non-sodium sensitive dosage so as to reduce the risk of pulmonary edema.

An anamnesis revealed cryptorchidism associated with an indirect inguinal hernia and a bilateral open peritoneo-vaginal process, aplasia of the posterior wall of the right inguinal canal, status post GnRH therapy resulting in descending of the right testis, post implantation of a lattice covering a defect of the posterior wall of the inguinal canal and orchidopexy on the right side, post umbilical herniotomy, and post left inguinal herniotomy.

Previously, at the age of 21, the patient had suffered from a sudden onset of arterial hypertension. His blood pressure reached levels as high as 240/130 mmHg. Laboratory analyses revealed a moderate hypokalemia and cortisolism, as well as more pronounced aldosteronism and reninism prior to the administration of medication. Before the diagnosis was reached on the basis of this constellation, alpha and beta blockers and an Angiotensin-Converting-Enzyme (ACE) inhibitor had been adminis-

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tered in an attempt to control the excessive blood pressure peaks. There were subsequent unsuccessful attempts between 1991 and 2001 to control hypertension using various quadruple, quintuple and sextuple combinations of antihypertensive agents. Twice, therapy had been escalated to a septuple combination consisting inter

alia of spironolactone or trilostane, an 11 beta-hydroxylase blocker, clonidine, ACE inhibitors, angiotensin receptor (AT) blockers, calcium channel blockers, dihydralazine, furosemide, hydrochlorothiazide and alpha and beta blockers (Figure 1). A conspicuous feature of the graphs in Figure 1 is the excessive and increasing

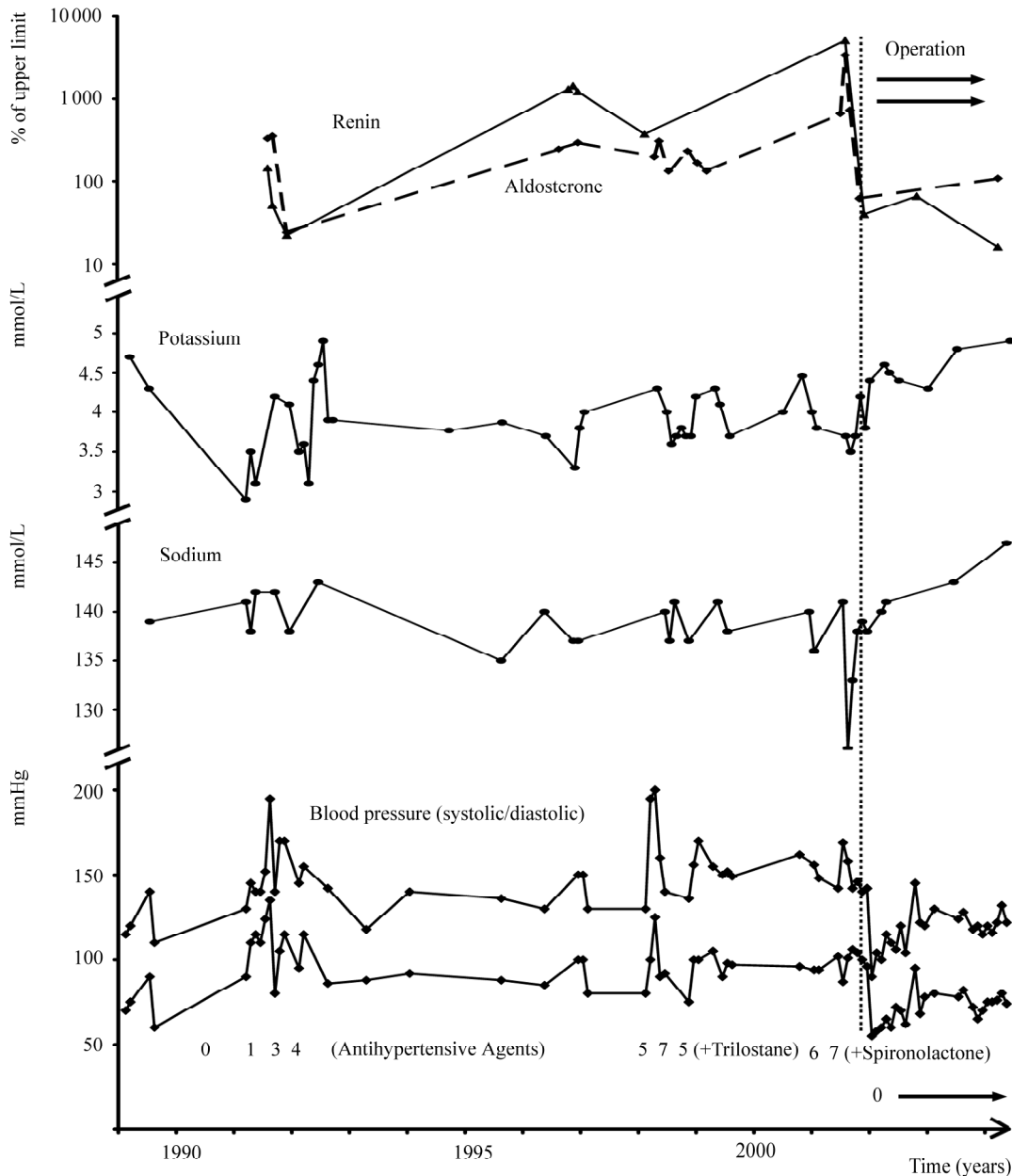


Figure 1. On the ordinate, serum renin and aldosterone in the percentage of the upper limit of the normal range (logarithmic), the serum potassium and sodium in mmol/L, the systolic and diastolic blood pressure in mmHg, the number of antihypertensive agents taken concurrently, and the date of the operation are plotted against the time (in years) on the abscissa. Blood pressure is expressed as the weekly mean of up to 34 single measurements where available retrospectively.

aldosteronism and reninism as well as the necessity for massive potassium and magnesium substitution during the course of the disease. Pheochromocytoma, aortic and renal artery stenosis, and renal diseases had been repeatedly ruled out. The adrenal glands were morphologically inconspicuous. No computed tomography (CT) scan of the lower abdomen had been carried out at that time.

A CT scan revealed a left iliacal mass on the measuring $2.5 \times 1.5 \times 1.5$ cm (Figure 2A), and retroperitoneal lymph nodes (Figure 2B) enlarged and also increased in number, especially the interaortocaval and in the area of the renal hili.

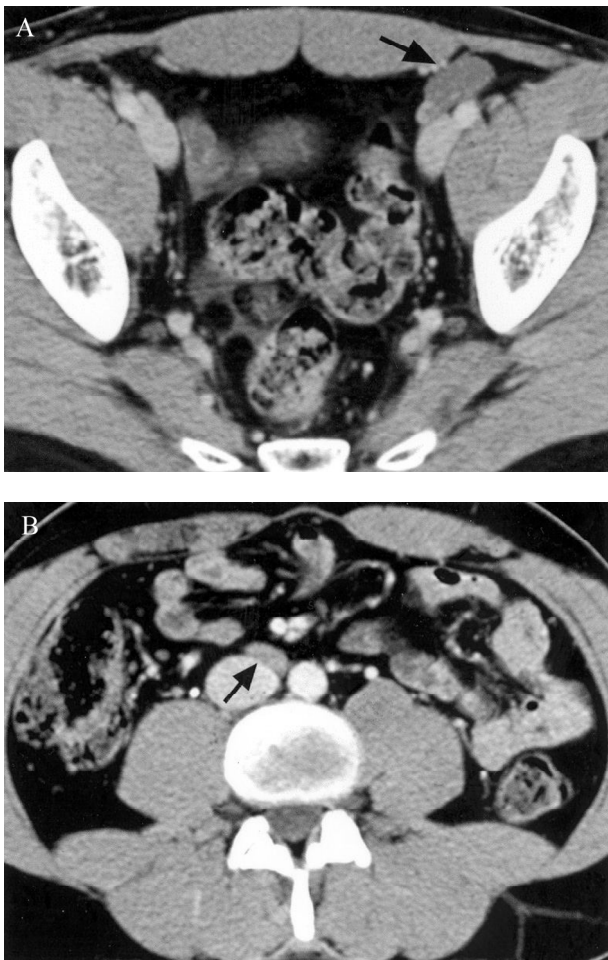


Figure 2. (A): Abdominal computed tomography scan. Left iliacal mass measuring $2.5 \times 1.5 \times 1.5$ cm. (B): Abdominal computed tomography scan. Retroperitoneal lymph node.

The iliacal mass was removed. The histological examination revealed an undescended testis with a Sertoli-cell-only-syndrome-like pattern. Immature Sertoli cells and persistent immature tubules without germinal cells were found (Figure 3). There were no signs of malignancy. Retroperitoneoscopic interaortocaval lymph node dissection revealed the presence of a retroperitoneal tetrillar hemangioma in which the lymph nodes were located. The histological examination revealed a sinus histiocytosis. The hemangioma was not removed.

After removal of the testis, we were astonished by the immediate cessation of arterial hypertension. Renin levels were normalized. It became possible to discontinue both antihypertensive medication and potassium substitution. Serum potassium increased until it was within the normal range (Figure 1). The patient recovered completely. He gained 16 kg in the following year, and completed a marathon in October 2004 with a time of 4:04:34.

2 Discussion

Reninomas is a rare cause of arterial hypertension. In the majority of the approximately 50 cases reported in the published literature, autonomous renin production was caused by intrarenal juxtaglomerular tumors, of which only six were malignant [1]. The extrarenal entities, some of which were described several times, included a liver cell carcinoma [2], a hepatoblastoma [3], a pelvic ter-

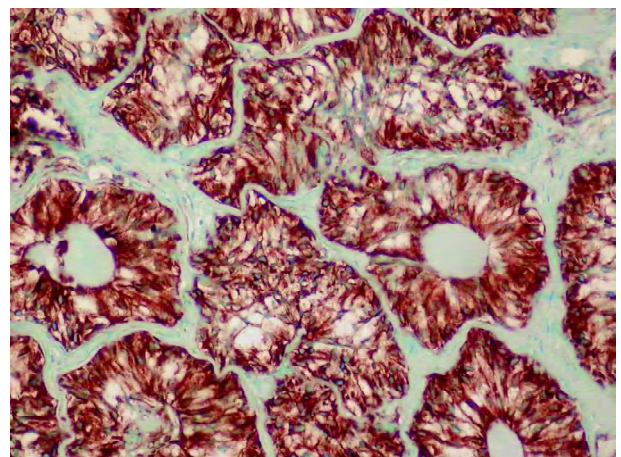


Figure 3. Anti-vimentin immunoperoxidase stained slide of the resected iliacal mass showing an undescended testis with a Sertoli-cell-only syndrome-like pattern. Immature Sertoli cells and persistent immature tubules are without germinal cells ($\times 400$).

atoma [4], a serous cystadenocarcinoma of the ovary [5], a leiomyosarcoma of the uterus [6], a leiomyosarcoma of the lung [7], a pancreas carcinoma [8], an adrenocortical carcinoma [9], a malignant paraganglionoma [10], a pheochromocytoma [11], a tumor of the fallopian tube [12] and a nephroblastoma [13].

Corresponding to our case, there is a striking evidence that Sertoli cells can produce renin, as shown by the case of a renin-producing ovarian Sertoli cell tumor [14]. Interestingly, in that case the arterial hypertension was, as in our case, refractory to medication.

Regarding the renin-angiotensin system during morphogenesis, renin-containing cells have been observed on the walls of the renal, the mesonephric, the adrenal and abdominal arteries, the adrenal glands, and the testes in male mouse embryos on the 13th day of gestation [15]. The testicular isoform of the angiotensin-converting enzyme (tACE) is expressed exclusively during spermatogenesis and fetal germ cell development [16]. Human chorionic cells secrete significant amounts of prorenin [17], and the ovaries are the source of both elevated plasma prorenin and renin in pregnant women [18]. Little is known about the late involution of the widespread embryonic renin system, especially in genitourinary tissues, or its later extrarenal occurrence and function. For the first time, the case presented here provided evidence for the persistence of the mesonephric and metanephric renin-producing potential not only of female, but also of male juvenile gonadal tissue. The renin production of the Sertoli-cell-only syndrome testis raises questions regarding the extent of renin expression in gonadal tissue in general, and its regular function in human gonadal tissue in particular. The relevancy during development as well as differentiation of gonadal tissue in arterial hypertension remains to be determined.

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