CASE REPORT

The Use of Minimally-invasive Cortical Sparing Adrenalectomy as an Approach to Bilateral Adrenal Masses in a Patient with von Hippel Lindau Syndrome: Learnings from a Lower Middle-income Country Setting

Cesar K. Jacinto III, MD, FPUA[1]; Joel Patrick A. Aldana, MD, FPUA[1]; Julia P. Young, MD[2] and Elizabeth T. Paz-Pacheco, MD, FPCP, FPSEDM[2]

[1]Division of Urology, Philippine General Hospital, University of the Philippines Manila
[2]Division of Endocrinology, Philippine General Hospital, University of the Philippines Manila

von Hippel Lindau syndrome is a rare genetic disease which may present with bilateral adrenal masses requiring surgical intervention. Previous practice at UP-PGH was to perform outright total adrenalectomy on pathologic adrenal glands and rely on lifelong steroid replacement for patients who had both adrenals removed. Presented here is a case of a patient diagnosed with von Hippel Lindau syndrome with bilateral adrenal masses, surgically managed initially with open adrenalectomy on the right side, followed by the first ever performed minimally invasive cortical sparing adrenalectomy at UP-PGH on the left side.

Key words: Adrenalectomy, von-Hippel Lindau syndrome

Background

von Hippel Lindau (vHL) syndrome is a rare inherited disorder involving a genetic mutation in the vHL gene. It is characterized by tumor formation in different parts of the body such as the eyes, brain, spinal cord, kidneys, pancreas, and adrenal glands. Adrenal tumors of patients with vHL are usually of a pheochromocytoma etiology.[1] Surgical excision of the pheochromocytoma remains to be the primary treatment for these sets of patients. Previous practice involved total adrenalectomy on pathologic adrenal glands. Difficulties arise in patients who require bilateral adrenalectomy as these patients would require a lifetime of steroid replacement. Steroid replacement requires close follow-up to monitor and adjust doses of exogenous steroids. Failure to do so may result in complications such as weight gain, loss of libido, overall depreciation of quality of life and the most dreaded and potentially fatal adrenal crisis.[2]

Cortical sparing adrenalectomy involves excision of the tumor and leaving normal-functioning adrenal tissue behind. Most important benefit of this procedure is that if enough adrenal tissue is left behind, patients may be spared from lifelong steroid therapy and avoid its possible dreaded complications.[3]

The Case

A 19-year-old male presented with hypertensive spikes and headaches 3 years prior to consult. He was initially managed with 3 anti-hypertensive drugs but was advised to seek consult at a specialty institution. Further workup included abdominal imaging which revealed bilateral adrenal masses, elevated 24-

hour urine metanephrine (3.782 mg/24 hours) and genetic testing which yielded a positive result for a pathologic gene, confirming vHL syndrome. Considering the elevated 24-hour urine metanephrine result and a confirmed diagnosis of vHL syndrome, our primary impression for the adrenal masses was pheochromocytoma.

Representative cuts of the abdominal CT scan are shown below (Figures 1 & 2). The right adrenal gland contained 2 tumors: a 4.3cm x 4.2cm mass on the lateral limb (plain HU: 25.2, absolute washout 53%, relative washout: 44%), and a 2.1cm x 2.1cm mass in the medial limb (plain HU: 27.1, absolute washout 70%, relative washout: 59%). On the contralateral side, the left adrenal gland contained a solitary tumor measuring 2.2cm x 2.0cm in its body (plain HU: 20.5, absolute washout 57%, relative washout: 57%)

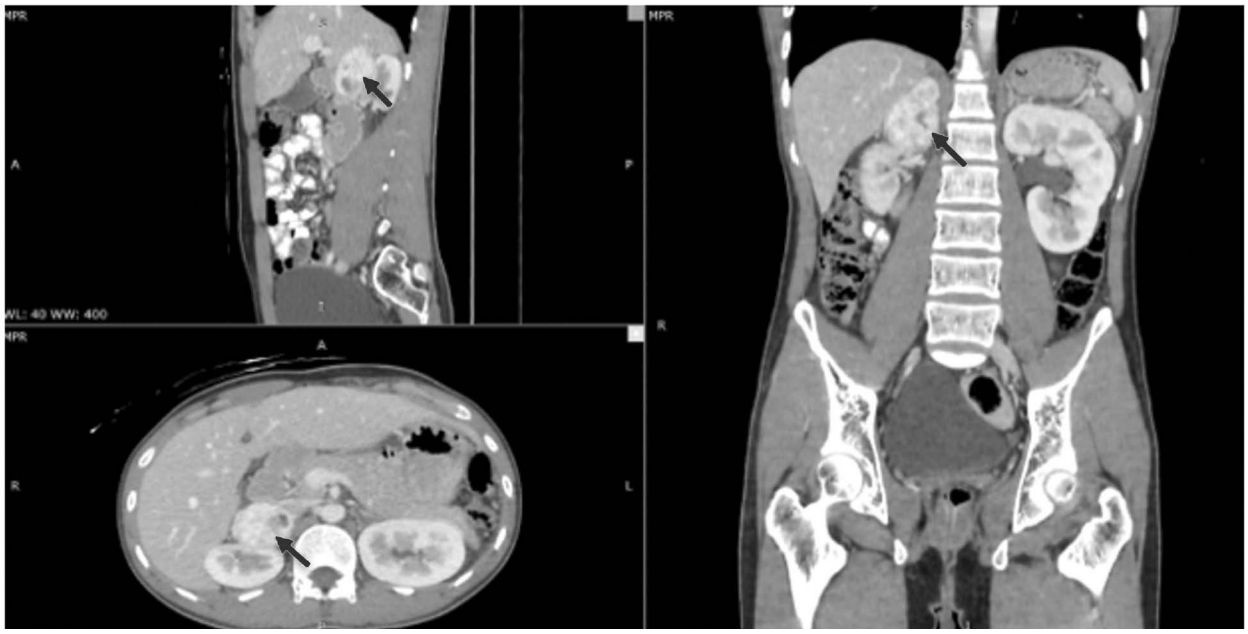

Figure 1. Pathologic right adrenal gland shown on different cuts from abdominal CT scan (arrows).

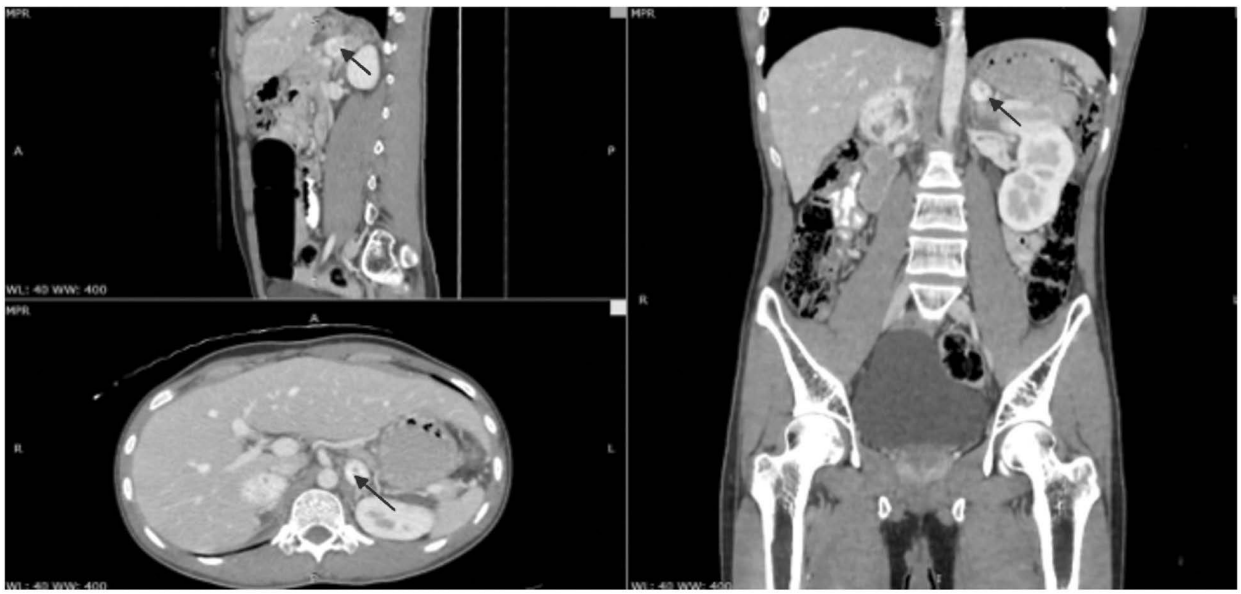

Figure 2. Left adrenal gland solitary tumor shown on different cuts from abdominal CT scan (arrows).

Treatment

After extensive discussions in multiple multidisciplinary team (MDT) meetings involving the Urology and Endocrine services, the patient underwent a total (open) adrenalectomy for the right adrenal gland, and an interval laparoscopic cortical sparing adrenalectomy for the left adrenal gland tumor after 4 weeks. A joint decision to perform an open adrenalectomy for the right side was established due to the size of the adrenal mass wherein the authors could not totally rule out a malignant process.

Laparoscopic cortical sparing adrenalectomy was performed transperitoneally and proceeded as standard practice similar to a laparoscopic total adrenalectomy as described by Stechman, 2022.[3] However, mobilization was minimized to just around the tumor, preservation of the adrenal vein was done, and only the tumor was excised, leaving normal adrenal tissue behind. The patient was placed in a left lateral decubitus position. Trocar placement was done as shown in Figure 3. The authors proceeded first with releasing the descending colon up to just distal to the left crus of the diaphragm. They ensured that the bowels and the tail of the pancreas would be deflected medially to avoid injury to surrounding structures. The Gerota's fascia was incised, and the main adrenal vein was identified. Dissection was continued superiorly to expose the superior aspect of the left adrenal along with the tumor (Figure 4). Once adequate exposure was achieved, the tumor was dissected off the normal adrenal gland with the use of ultrasonic shears, ensuring that a 5mm margin of normal tissue was included. Approximately 70% of normal adrenal tissue was preserved (Figure 5). After adequate hemostasis, the tumor was removed with the use of a sterile retrieval bag. Trocars were then removed and insertion sites were sutured. The patient had an unremarkable surgery. His vital signs were normal perioperatively, with no intraoperative adverse events encountered. In addition to this, no other pathologic lesions were seen in nearby organs and no blood transfusions were required. He was discharged 3 days after his surgery.

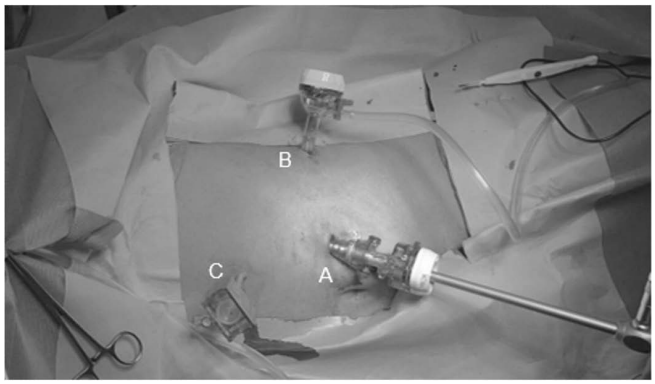

Figure 3. Patient positioning (left lateral decubitus) and trocar placement. (A) Visual port (12mm) was placed in the left periumbilical region, (B) 11mm trocar in the left subcostal region, and a (C) 5mm trocar in the subxiphoid region.

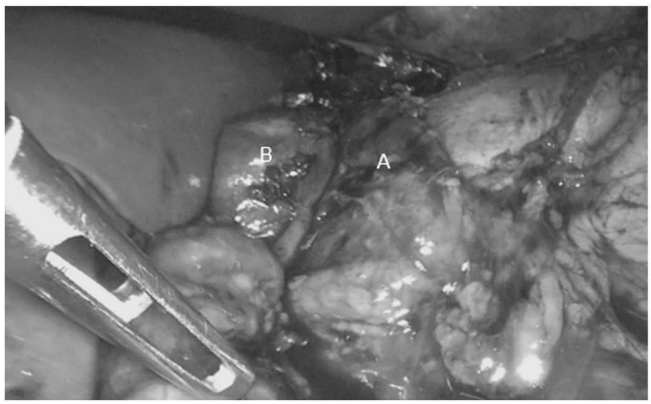

Figure 4. Exposure of the (A) left adrenal and the (B) tumor.

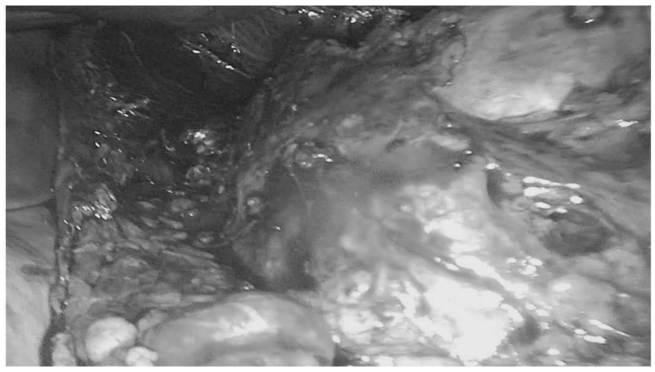

Figure 5. Left adrenal gland post-resection showing ~70% of residual normal adrenal tissue.

The pathology report for the right adrenal gland revealed pheochromocytoma, 5.0 centimeters in greatest dimension, with no lymphovascular invasion and negative tumor margins (Figure 6). The pathology report for the left adrenal gland revealed pheochromocytoma, 2.8 centimeters in greatest dimension, with no lympovascular invasion and negative tumor margins (Figure 7).

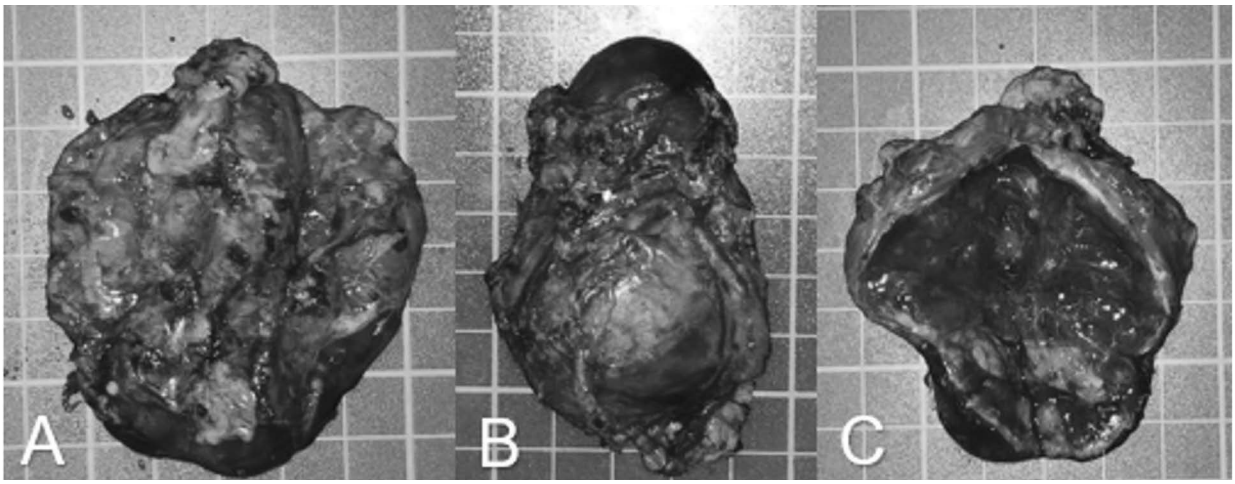

Figure 6. Right adrenal gland almost totally converted into a tumor (A) Anterior, (B) posterior, and (C) cut section views of the specimen.

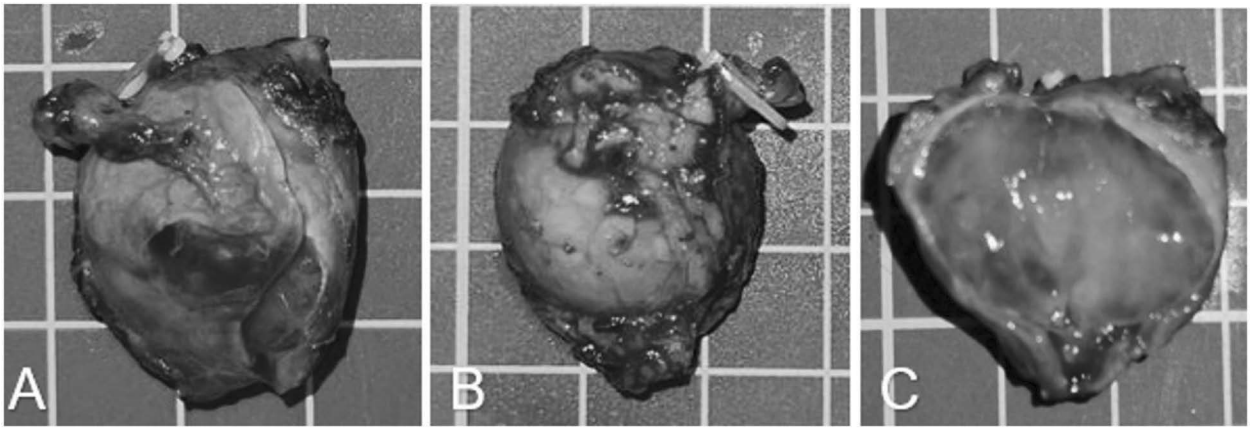

Figure 7. Left adrenal mass. (A) Anterior, (B) posterior, and (C) cut section views of the specimen.

He has been on constant follow-up with both the Urology and Endocrinology services and advised extensively regarding follow-up consults. He has had good blood pressure control since the surgery despite being off his anti-hypertensive medications and has had no symptoms of adrenal insufficiency despite not receiving any steroid replacement therapy.

Discussion

This report presents a rare case of vHL syndrome with bilateral adrenal tumors, managed with bilateral adrenalectomy in two separate surgeries wherein the laparoscopic cortical sparing adrenalectomy for the left adrenal tumor was the first ever performed at UP-PGH.

Cortical sparing adrenalectomy, which is also referred to as partial adrenalectomy or subtotal adrenalectomy, involves the resection of the pathologic lesion while leaving a significant amount of adrenal tissue. Two essential steps in performing a cortical sparing adrenalectomy are first, good exposure of as much of the adrenal gland without full mobilization, and second, preservation of its vascular structures.[4] A good margin of healthy adrenal tissue approximately 3 to 5 millimeters should be resected with the pathologic lesion. With regard to the amount of residual tissue that must be left behind after cortical sparing adrenalectomy, previous reports mentioned in a study by Perysinakis, et al. in 2020 recommend a volume of 15-30% in order to still be steroid-independent. Also mentioned was that if

a minimum of 15% residual adrenal tissue cannot be achieved, it is best to just perform an outright total adrenalectomy to decrease the chances of recurrence.[5] Cortical sparing adrenalectomy can be performed via open technique or via a minimally invasive approach. However, in recent times, the minimally invasive approach is preferred when feasible as it is proven to improve postoperative pain, shorten hospital admissions and hasten the recovery period.[4]

Cortical sparing adrenalectomy is recommended especially for patients with bilateral adrenal pathologies requiring surgical excision, to avoid the consequences of life-long steroid replacement which can lead to multiple complications if not monitored closely. During the early period of its conceptualization, surgeons initially would have to weigh the risks and benefits of performing a seemingly incomplete resection but with a patient safe from life-threatening Addisonian crisis. Some experts would also recommend cortical sparing adrenalectomy for unilateral tumors, attributing this to the fact that approximately 30% of patients are expected to develop contralateral adrenal disease over time hence the need to spare as much normal adrenal tissue as possible.[5] A review in 2010 by Kaye, et al. showed that cortical sparing adrenalectomy has a very low morbidity rate, very low recurrence rate (3%), and has approximately a 95% chance of granting patients freedom from steroid dependence.[6] Gumbs, et al. in 2006 specifically reviewed laparoscopic cortical sparing adrenalectomies mentioned in literature and found that mean operating times between total versus cortical sparing techniques did not differ significantly.[7]

In the setting of a lower middle-income country (LMIC) and that patient in the present study is from a rural area with fair access to healthcare, freedom from lifelong steroid replacement would greatly benefit him and his family as their funds can be reallocated to more basic needs. A retrospective study by Gunnarsson, et al. in 2017 explored the healthcare burden of patients with adrenal insufficiency from different etiologies. Aside from the fact that these patients must spend on the drugs, Gunnarsson, et al. found that patients suffering from adrenal insufficiency on steroid replacement had more frequent hospitalizations than patients in the control group. They concluded that irrespective of the cause of adrenal insufficiency, these patients had a significantly more substantial healthcare burden compared to their matched controls.[8]

This case of the first successfully performed minimally invasive cortical sparing adrenalectomy at UP-PGH, will pave the way for more cases to be performed using this technique. By doing so, surgeons can provide their patients the best quality of their care during their admission for their surgeries and at the same time, provide them with better quality of life, independent of exogenous steroids and the complications that come with it.

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