Case Report

# Symptomatic cycling Cushing disease managed by simultaneous bilateral laparoscopic adrenalectomy in a 11-year-old boy

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ABSTRACT

We report symptomatic cycling Cushing disease in a 11-year-old boy that was managed with simultaneous bilateral laparoscopic adrenalectomy. Positioning and the surgical technique have been fully described. Excellent results were achieved. Recent application of laparoscopic adrenalectomy for various adrenal pathology is highly effective and offers better results than open surgery. Post-operative recovery after laparoscopic technique is significantly shorter than the open technique.

**Key words:** Cycling Cushing disease, laparoscopic adrenalectomy, laparoscopic adrenalectomy in children, simultaneous bilateral adrenalectomy

## INTRODUCTION

Advanced laparoscopic surgery has played a significant role in urological practice in the 21st century.

Laparoscopic adrenalectomy has become the standard technique for the surgical removal of the adrenal gland at many centers worldwide.[1] Open surgical approach to the adrenal gland requires a large flank incision which carries higher morbidity and mortality particularly in patients with morbid obesity.

Laparoscopic surgery offers a shorter length of hospital stay, a decrease in post-operative pain, faster return to pre-operative activity level, improved cosmetic appearance and reduced overall morbidity.[2,3]

## CASE REPORT

The present case is about a 11-year-old Saudi boy who was referred to our service for bilateral adrenalectomy. The patient had been diagnosed by the pediatric endocrinologist with cycling Cushing disease. He presented with a history of rapid weight gain, decreased school performance and emotional disturbances in the previous 18 months. These complaints came in cycles, each lasting 2-4 months before the patient returned to normal. Adrenal ultrasound and computed tomography scan showed normal adrenal glands, magnetic resonance imaging of the brain also showed normal pituitary and hypothalamic area. During one of these cycles, his weight increased by 4 kg, blood pressure was 140/100 and he exhibited the typical signs of Cushing's disease. Blood work confirmed Cushing's disease. His serum cortisol at 1 PM was elevated at 16.6 mcg/dL. Electrolytes showed sodium 143 mEq/L, potassium 4 mEq/L, chloride 103 mEq/L, total $CO_2$ 27, blood urea nitrogen 9 mg/dL and creatinine 0.3 mg/dL. 24 h urine for free cortisol was high at 542 mcg/24 h (normal 5-55 mcg/24 h). At 2 months later, the patient presented with a third episode and was admitted for a full work-up. Urine free cortisol was elevated at 350 mcg/24 h, at midnight serum cortisol was 19.9 mcg/dL and serum adrenocorticotropic hormone (ACTH) was <4 pg/ml. Am cortisol was 15.7 mcg/dL and ACTH was 5.4 pg/ml. The patient was subjected to small and high doses of dexamethasone suppression tests (1 mg and 5 mg respectively) but there was no change in cortisol or ACTH serum levels. These findings confirmed a diagnosis of Cushing's disease

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of adrenal origin and its cyclic nature presented Carney's complex syndrome as a possibility. Further investigations to exclude Endocrine and cardiac tumors as part of Carney's complex included ultrasound of the thyroid and echocardiography which were normal. Ultrasound of the testes showed bilateral diffuse macro-calcification as well as 0.8 × 0.8 cm hypo-echoic mass within the left testicle, which strengthened the diagnosis of Carney's complex. The patient had no abnormal skin rash and the family history was negative for any endocrine problems. Deoxyribonucleic acid analysis on our patient did not show the known mutation in Carney's complex (PRKAR1A) which is found in only 50% of those patients.

Bilateral adrenalectomy was the treatment of choice in this patient. The plan was to start with right laparoscopic adrenalectomy and proceed to left adrenalectomy with the possibility of converting to open. The patient underwent a successful simultaneous bilateral adrenalectomy; the operative time was 3 h and 30 min. The estimated blood loss was 50 ml. The patient resumed his diet and ambulated on the 1st post-operative day and was discharged from hospital on the 2nd post-operative day. The post-operative course was smooth and uneventful and he was started on replacement therapy post-operatively. At 3 months later, he was doing well with a remarkable improvement in his behavior and school performance. Histopathology report of his adrenal gland was consistent with micronodular adrenocortical hyperplasia.

## SURGICAL TECHNIQUE

Clear fluid and mechanical bowel prep were started 24 h prior to surgery. Prophylactic intravenous (IV) antibiotic was given 1 h pre-operatively. Peri-operative steroids coverage planned by the pediatric Endocrinologist included hydrocortisone stress dose IV (100 mg/m$^2$) with induction of anesthesia then 100 mg/m$^2$/day IV every 6 h; the doses were switched to oral once he was well enough to take oral medications.

After the induction of the general anesthesia, a nasogastric tube and Foley catheter were inserted. The patient was placed in a supine position and the port sites marked. The plan was to place a total of five ports for both right and left adrenalectomy [Figure 1]. The preferred approach to the adrenal gland was the lateral transperitoneal. The patient was then placed on his left flank after the abdomen was prepped [Figure 2]. Three ports placed in the upper mid-line and the 4th at the left mid-clavicle line at the level of the umbilicus [Figure 3]. One of the ports was used to retract the liver, the other two ports used for the instruments. The camera was placed at the umbilical port and 5 mm laparoscopic clips used to secure the right adrenal glands. For the left laparoscopic adrenalectomy, the patient was repositioned to the right flank position. We did not add any port as planned and we managed to perform the laparoscopic left adrenalectomy through the three ports at the upper mid-line [Figure 4].

The adrenal glands were completely excised, placed in a plastic bag and removed through the supra-umbilical trocar site.

## DISCUSSION

A total of 34 laparoscopic adrenalectomy have been performed in our urology unit. More experience is being acquired in laparoscopic surgery in children including laparoscopic nephrectomy and excision of urachal cyst.

The role of minimally invasive surgery (MIS) in children with solid neoplasms is slowly evolving. MIS appears to be an ideal way to obtain diagnostic information (i.e. tissue

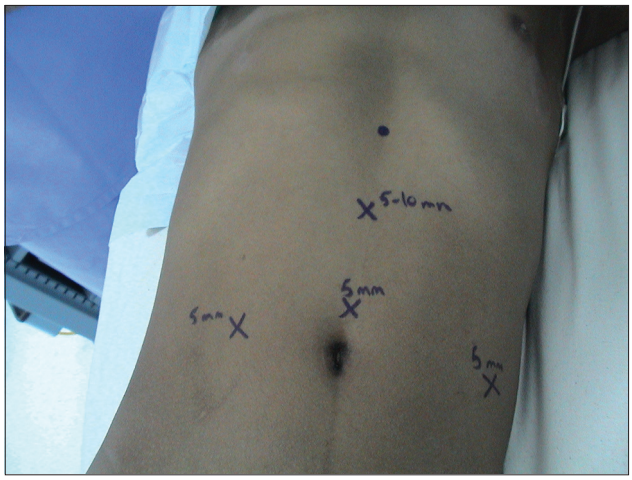


**Figure 1:** Planned port sites for bilateral laparoscopic adrenalectomy

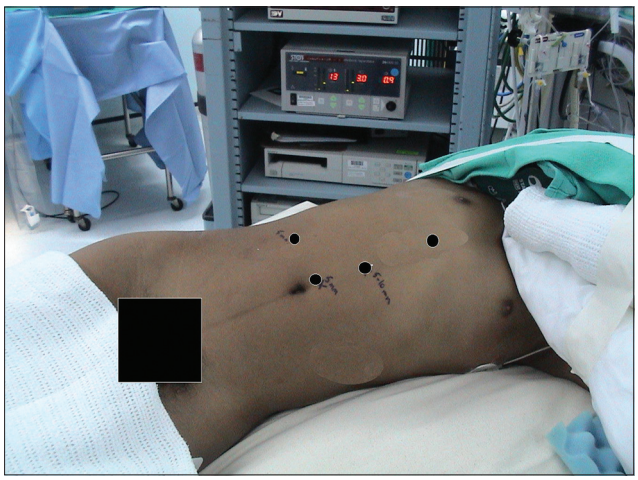

**Figure 2:** Position for right laparoscopic adrenalectomy

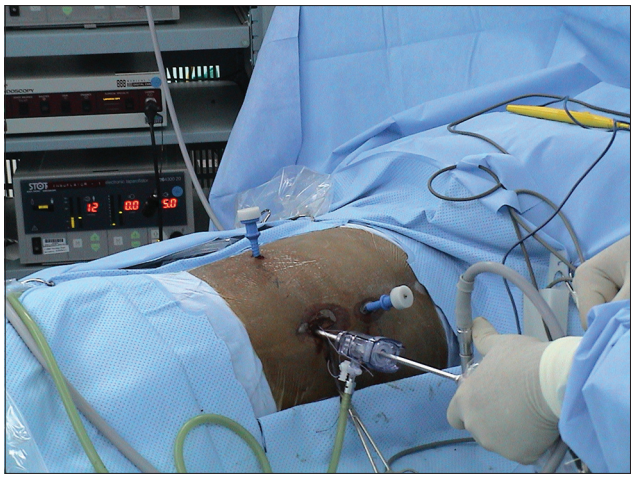

**Figure 3:** Trocar sites for right laparoscopic adrenalectomy

**Figure 4:** Position and port sites for left laparoscopic adrenalectomy

biopsy) in children with solid neoplasms, but its role as an ablative/curative technique is controversial.[4]

Simultaneous bilateral laparoscopic adrenalectomy is performed to minimize the number of trocar sites and operation time. Success was attributed to the careful selection of trocar sites to permit safe dissection.[5]

The role of laparoscopy in children with Neuroblastoma has not been fully defined. The laparoscopic approach to the adrenal gland is already largely used in adults and a few cases have been reported in children.[6]

Laparoscopic procedures in pediatric urology are gaining popularity, with an increasing number of centers performing advanced surgery. Indications have expanded from diagnostic to ablative surgery and more recently to reconstructive procedures.[7]

Conventional adrenalectomy for tumor excision is one of the challenging tasks in adults as well as children with mounting post-operative complications. Advancing laparoscopic techniques with the new era of diagnostic image developments aids this kind of procedure performed mainly in adults with only a few reported cases in children.[8]

In our patient, a simultaneous bilateral laparoscopic adrenalectomy was performed successfully without fragmentation, with uneventful post-operative course. The laparoscopic technique assures less morbidity, faster recovery and appears to be equally effective in eradicating functioning and non-functioning adrenal masses.[9] Although only a few reports indicate the feasibility of laparoscopic adrenalectomy,[10] the results indicate that this procedure is comparable with open adrenalectomy in controlling symptoms of Cushing's disease.[11] This procedure is safe and effective with minimal blood loss and patients are discharged post-operatively in a relatively short time with few complications.[12-16]

## CONCLUSION

We believe that laparoscopic adrenalectomy, if done by skilled surgeons, is now the first choice for treating most adrenal tumors, Although commonly performed in adults, laparoscopic adrenalectomy in children including simultaneous bilateral adrenalectomy is performed only in centers with advanced laparoscopic expertise.[17] This is comparable to open adrenalectomy to control symptoms of Cushing's disease.

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