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Simultaneous bilateral laparoscopic adrenalectomy for pheochromocytoma in multiple endocrine neoplasia (MEN) syndrome: Case report with review literature



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ABSTRACT

INTRODUCTION: Laparoscopic adrenalectomy has gained favour as a preferred surgical approach in the multiple endocrine neoplasia (MEN) type 2 patients. Currently, there is limited literature on bilateral simultaneous laparoscopic adrenalectomy in MEN 2 syndrome. We reported two cases of bilateral pheochromocytoma associated with MEN 2 syndrome cured by simultaneous bilateral laparoscopic adrenalectomy.

PRESENTATION OF CASE: First patient presented with big lips since childhood and episodic abdominal pain. On investigations, he was diagnosed with features of MEN 2B syndrome. Second patient was hypertensive and presented with abdominal pain. On evaluation she had features of MEN 2A syndrome.

DISCUSSION: Minimally invasive approach was preferred in both cases. Bilateral simultaneous adrenalectomies were uneventfully done with acceptable operative time and blood loss with rapid perioperative recovery. These cases highlighted the feasibility of laparoscopic simultaneous bilateral adrenalectomy for pheochromocytomas in MEN 2 syndrome.

CONCLUSION: Laparoscopic simultaneous bilateral adrenalectomy is a safe feasible and preferable technique for pheochromocytomas associated with MEN 2 syndrome.

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1. Introduction

Pheochromocytomas are neoplasms arising from chromaffin cells in the adrenal medulla and produce catecholamines.¹ In patients of MEN 2 syndromes approximately 40–50% develop pheochromocytomas, at the age range of 30–40 years and rarely at 5–10 years.² In MEN 2 syndrome pheochromocytomas are usually benign. They have multifocal medullary lesions and are bilateral in more than 50% of cases.³ Considering these features, minimal invasive surgical approach should be applied here.

Since the first description of pheochromocytoma by Felix Frankel and its first successful surgical management by César Roux and Charles Mayo, the management of pheochromocytoma has evolved greatly.⁴ Today, laparoscopic adrenalectomy is preferred surgical approach for MEN 2 patients. The conversion rate to an open procedure is less than 10%.⁵ Although the benefits of unilateral laparoscopic adrenalectomy have been well documented, less

experience has been reported in literature with simultaneous bilateral laparoscopic adrenalectomy. We describe two cases of bilateral pheochromocytoma in patients with MEN 2 syndrome treated with simultaneous laparoscopic bilateral adrenalectomy with a review of literature.

2. Presentation of case

2.1. Case 1

45 years, diabetic male with big lips as his identification mark since childhood, presented with episodic upper abdomen pain for 1 year. On workup, plasma free metanephrines and normetanephrines levels were 2905 pg/ml and 2240 pg/ml, respectively. Serum calcitonin was 6243 pg/ml. Contrast enhanced CT imaging showed bilateral enlarged adrenal, thyroid nodule and rest normal (Figs. 1–3). Thyroid nodule was diagnosed as medullary carcinoma thyroid on FNAC. His ophthalmologic evaluation detected eyelid neuroma and thickened corneal nerves. His anal manometric evaluation patient was suggestive of Hirschsprung's disease.

He was diagnosed to have MEN 2B syndrome. Patient was optimized with alpha blockers and underwent surgery two weeks later. The patient was placed in supine position with a 30° tilt on the right

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Fig. 1. CECT scan: B/L adrenal pheochromocytoma.

side and pneumoperitoneum was created. Four ports were used, one for camera and one for retracting liver. The peritoneum over the vena cava and in the Morrison space was incised by a hook. The adrenal gland was dissected from inferior vena cava with the help of harmonic ultrasound scalpel and appropriate use of clips. Climbing on this plane, adrenal vein was dissected, hemlock clips applied and cut. Right adrenal was then dissected off the superior pole of kidney and an endobag was used to remove the specimen through lower port. A drain was placed in the Morrison space and the right sided ports were closed. The patient was then tilted 30° on the left side. Three ports were used. After pneumoperitoneum was established again, the splenic flexure and colon was reflected to approach left adrenal. Left adrenal vein was identified, secured and

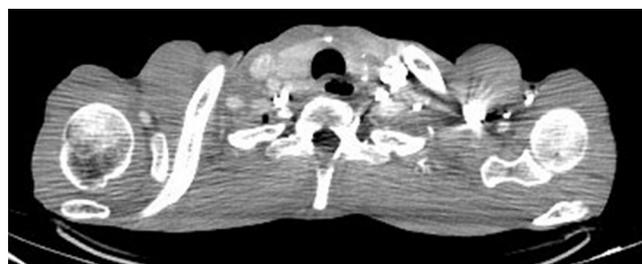


Fig. 3. CECT scan: thyroid nodule.

cut. The gland was then dissected and specimen delivered through endobag from lower port. Operating time was 5 h, blood loss was 300 cm³. Postoperative period was uneventful. Patient was started on steroid replacement. The blood sugar also normalized. Patient was started on diet on postoperative day 1. Drains were removed on postoperative day 3 and patient discharged. Total thyroidectomy with bilateral neck dissection was done for medullary carcinoma thyroid after one month. The patient is doing well on follow-up.

2.2. Case 2

43 years, hypertensive female was having paroxysmal episodes of abdominal pain, headache, giddiness and palpitation since 15 years. Her imaging detected bilateral adrenal enlargement. Her family history revealed thyroid disorder in two of four siblings and early death of elder brother due to hypertension. Hypertension was recorded in both supine and standing position. She was having an interscapular 8 cm × 7 cm lichen amyloidosis which was later

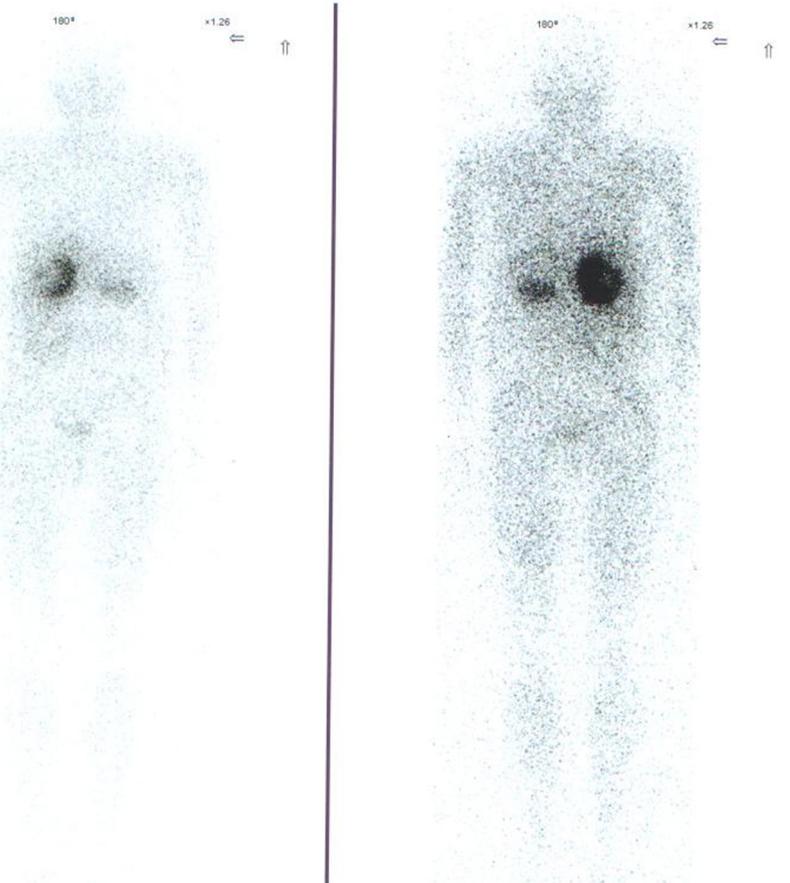


Fig. 2. MIBG scan: uptake in B/L adrenal pheochromocytoma.

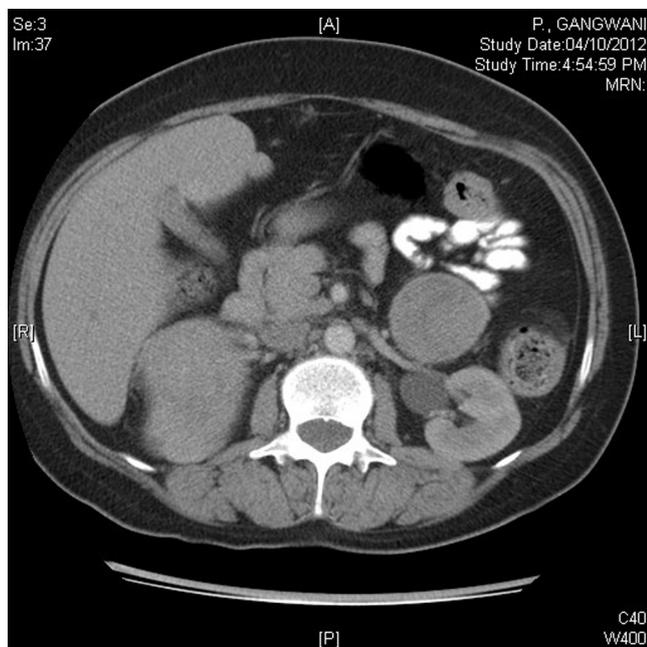


Fig. 4. PET-CECT: B/L pheochromocytoma.

confirmed by skin biopsy. Her plasma free metanephrines, plasma free normetanephrines, serum calcitonin and serum parathyroid hormone were 923 pg/ml, 4289 pg/ml, 597 pg/ml and 346.7 pg/ml, respectively, all raised. PET-CECT showed bilaterally enlarged adrenals and thyroid nodule (Figs. 4 and 5). MIBG scan showed increased tracer uptake in bilateral adrenal masses and a left para aortic mass (Fig. 6).

Clinical evaluation lead to the diagnosis of MEN 2A. Patient was optimized with alpha blockers. Laparoscopic bilateral adrenalectomy was done along with excision of left para aortic paraganglioma. Operative time was 4 h and blood loss was 300 cm³. Histopathologic examination confirmed bilateral pheochromocytoma with left paraganglioma. Total thyroidectomy with bilateral neck dissection and excision of enlarged right superior, right inferior and left inferior parathyroid glands was done after two months. Histopathology confirmed medullary carcinoma thyroid and parathyroid hyperplasia. Patient is doing well on followup.

3. Discussion

The reported frequency of bilateral pheochromocytoma is variable in published reports, ranging from 35% to 80%.^{6,7} It is also known that the mutation in the codons 634 and 618 is related to bilaterality.⁷ Patients suspected to have MEN 2 syndromes should be screened for pheochromocytoma. North American Neuroendocrine Tumour Society (NANETS) recommends that initial



Fig. 5. PET-CECT: thyroid nodule.



Fig. 6. MIBG scan: B/L adrenal uptake.

testing must include measurements of fractionated metanephrines in plasma, urine, or both, as available.⁸ Anatomical imaging is most widely used in the initial evaluation of patients as it offers the advantage of low cost, universal availability and easy interpretation. CT scan can be used to localize adrenal tumours > 1 cm and extra adrenal tumours > 2 cm.⁹ Adrenal tumours should be imaged with either 123 I-MIBG scintigraphy, 18 F-DA PET or 18 F-DOPA PET to detect/exclude multifocal or metastatic disease. Patients are also screened for all associated MEN 2 abnormalities. Usually pheochromocytomas are dealt first. Thyroid medullary carcinoma and other conditions can be addressed later after excision of pheochromocytomas.

Although laparoscopic adrenalectomy is preferred therapy for pheochromocytoma, experience with simultaneous laparoscopic bilateral adrenalectomy is limited. Also open bilateral adrenalectomy is associated with a high rate of surgical complications including intraoperative and postoperative bleeding (22%), pancreatic fistula, incisional hernia (10%), and wound healing impairment (13.5%) as well as general complications. Bilateral laparoscopic adrenalectomy is feasible and seems to be associated with a lower operative risk.^{5,10} However the reported rate of complications is higher than for unilateral laparoscopic adrenalectomy.¹¹ In laparoscopic approach as compared with the open one, there is less manipulation of the adrenal tumour.

Case series has been reported for unilateral laparoscopic adrenalectomy in pheochromocytoma associated with MEN syndrome; but bilateral simultaneous laparoscopic adrenalectomy sparsely reported in literature. Absence of intraoperative and postoperative complications in our cases contributes to lower rate of complications for this surgical procedure. Surgical approaches described for laparoscopic synchronous bilateral adrenalectomy include; “anterior” transperitoneal approach, lateral transperitoneal or retroperitoneal approach and the “posterior” retroperitoneal approach.^{12,13} No randomized trial has been published yet to confirm superiority of one approach over another. We used transperitoneal laparoscopic approach as it allows proper identification of anatomic landmarks, especially early ligation of the suprarenal vein and thus avoid vena caval bleeding that constitutes the most dangerous complication of the entire procedure.¹⁴ This approach also saves time as it does not require a changing in the patient position but only a 30° lateral tilt achievable by operating table. Laparoscopic lateral approach has been frequently described

for this kind of operation. It makes mobilization of abdominal organs less difficult due to effect of gravity upon organs and when performed with retroperitoneal approach provides direct access to the adrenal gland. Disadvantages of the lateral approach include the time spent in changing patient's position and longer learning curve. Even the posterior approach has been described for bilateral laparoscopic adrenalectomy. It does not require the time for repositioning of the patient as it allows a direct access to the adrenal glands vessels, and it is useful in case of patient with multiple intra-abdominal adhesions. Disadvantages of this approach are the very narrow operation field, which allows operating only small tumour, a difficult vascular control and inability to explore intraperitoneal structures. Our operating time, similarly to those reported literature was fairly long. This time duration was probably due to the fact that our experience, like most of the cases reported in literature, was the initial experience dealing with such cases. Postoperatively our patients were given steroid replacement. These two cases highlight the safety, feasibility, early patient recovery, patient comfort and good outcome in doing simultaneous bilateral adrenalectomy. In future minimal invasive simultaneous bilateral adrenalectomy should be surgery of choice in MEN 2 patients.

Conflict of interest

None.

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None.

Consent

Due consent was taken for video and photo recording and case publication for academic purpose. Consent can be produced at any time required.

Author contributions

DMG urooncology staff contributed for data collection and analysis.

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