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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.

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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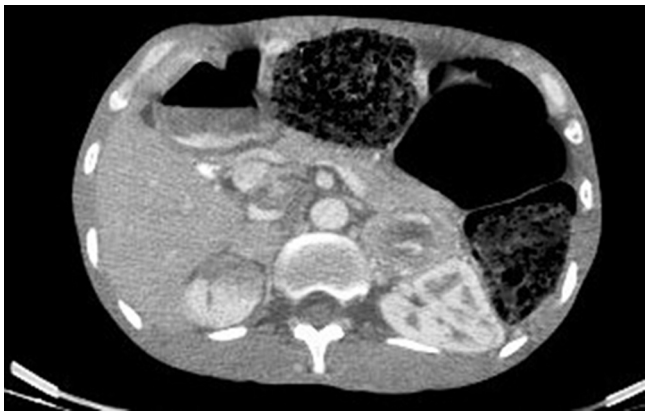

Fig. 1. CECT scan: B/L adrenal pheochromocytoma.

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

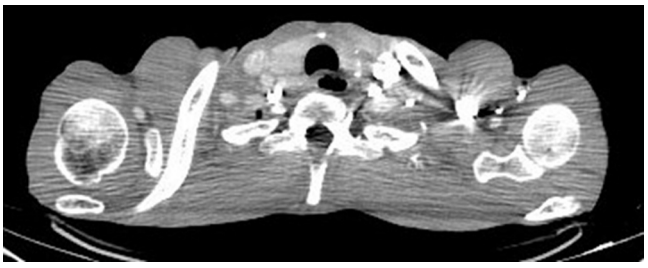

Fig. 3. CECT scan: thyroid nodule.

cut. The gland was then dissected and specimen delivered through endobag from lower port. Operating time was 5 h, blood loss was 300 cm^3. 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

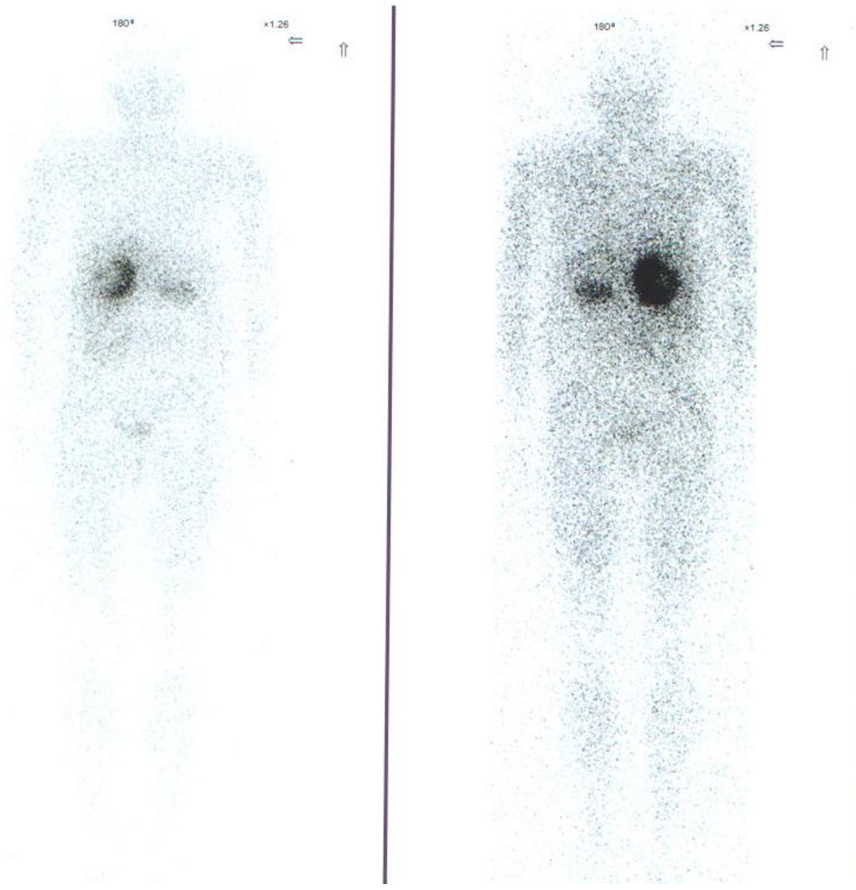

Fig. 2. MIBG scan: uptake in B/L adrenal pheochromocytoma.