Two-stage bilateral laparoscopic adrenalectomy for large pheochromocytomas

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Abstract
A 66-year-old Lithuanian female patient with a history of hypertension was diagnosed with bilateral adrenal tumors during a routine sonoscopy. Scintigraphy with metaiodobenzylguanidine and computed tomography scan revealed right 130/116/93 mm and left 85/61/53 mm pheochromocytomas. The patient suffered from hypertension with blood pressure over 240/100 mm Hg and heartbeat disturbances. Blood adrenaline levels exceeded the norm 10-fold. After possible spread of tumors was rejected, laparoscopic transperitoneal adrenalectomy was planned in 2 stages, starting on the right then followed by the left side. After preoperative treatment with adrenoblockers, 2-stage bilateral laparoscopic adrenalectomy was performed. 13 cm × 12 cm × 9.5 cm right adrenal and, 3 months later, 8.5 cm × 8 cm × 6 cm left adrenal pheochromocytomas were removed. Histologically – radical extirpation, pheochromocytomas with possible malignant potential. Stable remission of hypertension was achieved postoperatively. Laparoscopic transperitoneal adrenalectomy is a safe and feasible method of treatment of large benign and possible malignant, but noninvasive pheochromocytomas.

Key words: pheochromocytoma, large pheochromocytoma, laparoscopic adrenalectomy, bilateral pheochromocytoma, laparoscopic transperitoneal adrenalectomy.

Introduction
Pheochromocytoma is a rare, catecholamine-secreting tumor arising mostly from the chromaffin cells of the adrenal gland, which clinically presents as a potentially curable form of hypertension. The annual incidence of pheochromocytomas is approximately 1–4/10⁶ population or 0.5% of subjects with hypertension [1]. In patients with inherited pheochromocytoma, benign and bilateral tumors are more common [2].

The diagnosis of pheochromocytoma is obtained by biochemical confirmation of catecholamine excess and imaging studies. Imaging studies used for diagnosis are ultrasound and computed tomography (CT) scan subsequently. Moreover, scintigraphy with metaiodobenzylguanidine (MIBG), which is accumulated by adrenergic tissues, helps to determine an active pheochromocytoma diagnosis. Five–six percent of pheochromocytomas can be malignant [3]. The malignancy is confirmed by finding a metastatic spread; otherwise a benign diagnosis is established [4].

The only radical treatment of pheochromocytoma is adrenalectomy. Laparoscopic removal of pheochromocytoma is the preferred surgical technique, but, in case of malignancy, with signs of invasion, a transfemoral approach should be considered [5]. There were concerns about the size of tumor to be treated laparoscopically. However, developing minimally invasive techniques allows this method to be used for larger tumors [3]. The laparoscopic approach can be either transperitoneal or retroperitoneal. Substi-
tutional therapy with corticosteroids postoperatively and long-term clinical follow-up of the patients are essential to obtain a good outcome [4].

We present a case of a patient with large bilateral pheochromocytomas, which were removed in two stages using the laparoscopic transabdominal technique.

Case report

A 66-year-old Lithuanian female patient with a history of high blood pressure was incidentally diagnosed with bilateral adrenal tumors during a routine sonoscopy in December, 2009. Computed tomography (CT) scan showed a right 100 mm × 76 mm adrenal tumor and 87 mm × 58 mm left adrenal tumor with signs of cystic degeneration and no growth into surrounding structures. Blood adrenaline (1100 ng/l) levels exceeded the normal range 10-fold. Body scintigraphy with MIBG revealed right and left active pheochromocytomas (Photo 1). On the other hand, calcitonin levels were normal (5.5 ng/l), which eliminated the multiple endocrine neoplasia type 2A (MEN) syndrome diagnosis together with normal thyroid sonography results.

Operative treatment was suggested to the patient. However, the patient refused operation and further examination. Monitoring was resumed after the CT scan, performed in 2011, showed 10% growth of the right adrenal tumor (110 mm × 87 mm). One year later the patient started to suffer from hypertension episodes with blood pressure over 240/100 mm Hg accompanied by headaches with vertigo and heartbeat disturbances. Computed tomography scan revealed right 130/116/93 mm and left 85/61/53 mm pheochromocytomas with clear boundaries, without invasion or growth into the surrounding structures. The right adrenal tumor was contacting and pressing the right liver lobe, inferior vena cava, right kidney and renal vein. The left adrenal tumor was contacting the pancreatic tail, posterior wall of the stomach, superior pole of the spleen, and also pressing and deforming the superior pole of the left kidney (Photo 1).

By that time the patient agreed to have an operation. Considering the fact that the tumors had not overgrown or invaded the surrounding organs in a 2-year period, the benign type of pheochromocytoma was confirmed, which provided the opportunity to select a laparoscopic approach for the resection of tumors. The laparoscopic adrenalectomy was planned in 2 stages, starting with the right and followed by left laparoscopic adrenalectomy 3 months later. A lateral transperitoneal approach was chosen for both operations. The patient received a 2-month course of the adrenoblockers phenoxybenzamine and metoprolol prior to the operation.

Photo 1. Body scintigraphy with late imaging (MIBG) revealed active pheochromocytomas
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Right laparoscopic adrenalectomy was started by placing the patient in a right lateral position. Four trocars were inserted 2 cm below the right costal arch as follows: midclavicular line (10 mm), anterior axillary line (10 mm), midaxillary line (10 mm), posterior axillary line (5 mm). After the right hepatic lobe was elevated with a retractor, the duodenum was mobilized using Kocher’s approach. The 13 cm × 12 cm × 9.5 cm adrenal gland was visualized between the inferior vena cava and superior renal pole. It was then separated using a harmonic scalpel and electrocoagulation. The 7 mm central adrenal vein was identified, stapled with an endo-GIA stapler and then cut. The separated gland was retrieved in a bag via the midclavicular port expanding the incision to 6 cm (Photos 3 and 4). Blood loss was 130 ml. Operation time was 210 min. No severe blood pressure disturbances were observed during the adrenalectomy. The postoperative period passed without complications. The patient was discharged after 2 days. Histological findings proved the radical extirpation of pheochromocytoma, but with possible malignant potential: invasion to capsule, mitotic activity, focal necrosis.

Left laparoscopic adrenalectomy was performed 3 months later. The same position of trocars on the left side was established. The left colonic flexure and spleen were mobilized using a harmonic scalpel and electrocoagulation. The spleen was pushed with a retractor medially and the pancreatic tail was mobilized. The 8.5 cm × 8 cm × 6 cm left adrenal gland was found under the pancreatic tail medially from the left kidney’s superior pole. The central vein was identified and clipped with a large clip in the site of outflow from the renal vein. The gland was separated and retrieved through the anterior axillary trocar site, expanding the incision to 5 cm length (Photos 3 and 4). Blood loss was 20 ml, operation time 180 min. Histological findings revealed similar radically resected pheochromocytoma with possible malignant potential. The postoperative course passed without complications. The patient was transferred to the endocrinological unit 2 days after surgery for adrenal hormone balance correction and was discharged on 45 mg of hydrocortisone and 100 mg fludrocortisone daily doses afterwards. Three months later chromogranin A level was 121.7 ng/ml (normal < 148.56 ng/ml). Scintigraphy with MIBG did not reveal any active accumulation spots 3 months postoperatively. Stable remission of hypertension and elimination of heartbeat disturbances were achieved.
Discussion

After laparoscopic surgery of the adrenal gland was first described in 1992 by Gagner, who used the transabdominal approach to access the gland in a patient with Cushing syndrome, this method has become a gold standard for managing benign adrenal lesions as well as pheochromocytomas [3, 5].

The initial goal for selection of laparoscopic treatment of pheochromocytoma is to determine whether a tumor is benign or malignant. Despite the concerns that large pheochromocytomas are more likely to be malignant, in our case, imaging studies showed no tumor overgrowth or spread into the surrounding structures and organs, which indicated a benign diagnosis.

A concern considering catecholaminergic discharge caused by the pneumoperitoneum did not seem sufficiently important to discard this technique as a first choice management. However, the procedure should be performed by an experienced surgeon, in order to reduce gland manipulation and shorten the operation time [6]. Early clipping and dividing of the adrenal vein helps to avoid a catecholamine-induced hypertensive crisis [3].

It is very important to control arterial blood pressure, heart rate and arrhythmias and to restore the blood volume to normal prior to surgery [7]. Preparative treatment is usually started with administration of α-adrenoblockers over a period of a fortnight. Additional β-adrenergic blockade can be required to treat any associated tachyarrhythmias. Our patient received an initial therapy with phenoxybenzamine 10 mg 3 times daily and metoprolol 25 mg twice daily prior to the operation.

Which approach to choose? The retroperitoneal approach could be the primary choice in patients with severe cardiopulmonary disease in whom an increase in the CO₂ level may occur, even in the presence of increased pulmonary ventilation [5]. On the other hand, the transperitoneal approach opens more workspace for manipulation. Also it may be beneficial for obese patients [8]. However, there is no significant difference between retroperitoneal and transperitoneal method outcomes, so the operating surgeon can select one of the mentioned approaches depending on his own experience and concerns [5, 8].

The retroperitoneal approach is feasible for tumors up to 6 cm in size. For tumors larger than 6 cm transabdominal laparoscopic technique should be preferred [9, 10]. There is some evidence of successful laparoscopic adrenalectomy for an 18 cm tumor [9]. The main contraindication for laparoscopy is a malignant, 10 cm tumor [9, 11]. Our case report demonstrates that laparoscopy could be used for tumors larger than 10 cm without signs of invasion to adjacent structures and organs.

The main reason for conversion is intraoperative bleeding as the result of splenic injury or tear of the vena cava during dissection of the right adrenal vein [10, 12]. In our case major bleeding was avoided and the blood loss was minimal. Some sources suggest that left-sided adrenalectomy is an independent risk factor for overall and surgical complications after laparoscopic adrenalectomy [12]. It might be explained partly by the lateral transabdominal approach, which requires partial mobilization of the left pancreas and spleen. In our case bleeding during the left adrenalectomy was not significant.

Bilateral tumors may determine attempts to seek a genetic syndrome. Although the MEN syndrome diagnosis was eliminated after normal blood calcitonin and thyroid sonography results were obtained, a precise examination by a geneticist should be performed. Moreover, genetic evaluation can play an important role determining the malignancy and prognosis of pheochromocytoma [13]. Unfortunately, our patient refused genetic evaluation.

Histological findings revealed possible risk of malignancy. Pheochromocytoma of the adrenal gland scaled score (PASS) was 5 points for both sides (invasion to capsule 1 point; focal necrosis 2 points; mitotic activity 2 points). It is held that tumors with PASS below 4 have a benign behavior; on the other hand, those with PASS higher than 4 can act aggressively [14]. This is why a long-term follow-up of an oncologist is essential even if there were no signs of metastatic spread. Alternatively, normal blood chromogranin A level 3 months after the operation indicates a good short-term outcome [15].

Conclusions

Laparoscopic transperitoneal two-stage bilateral adrenalectomy is a safe and feasible method of treatment of large benign or malignant but noninvasive pheochromocytomas.

References

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Received: 31.07.2013, accepted: 5.11.2013.