JEMIS

Journal of Endoscopic, Minimally Invasive Surgery in Newborn, Children and Adolescent - ISSN 2283-7116

Laparoscopic surgery and hemodynamic changes during adrenalectomy for pheochromcytoma in childhood: management of two cases and literature review

published on June 28, 2016 in **Articles**

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DOI: http://dx.medra.org/10.1473/JEMIS20

KEYWORDS:

Pheochromocytoma in children, Laparoscopic adrenalectomy

CONFLICTS OF INTERESTS: none declared.

ABSTRACT

Pheochromocytoma (PCC) is a catecholamine-secreting tumor arising from chromaffin cells of the adrenal medulla and extra-adrenal sites. Approximately 10% of this rare tumor is found in pediatric age group. Perioperative management of pheochromocytoma is a challenge that requires a multidisciplinary approach for optimal care and successful outcome. Surgical resection remains the mainstay treatment of symptomatic and medically refractory PCC. The operative risk comes from hypertensive spikes and arrhythmias due to painful stimulations and mass manipulation. Laparoscopic adrenalectomy (LA) has been accepted as the "gold standard" treatment modality for

small-sized (<6 cm) PCC. Its use for larger PCC (>6 cm) was also reported in the literature. The laparoscopic approach allows minimal handling of the tumor with early control of the adrenal vein, and this results in lesser intraoperative hemodynamic alterations in comparison to open surgery. We report two cases of PCC in children, which we have treated with laparoscopic adrenalectomy with good outcome. The advantages of low morbidity, fewer complications, less intraoperative blood loss, quicker recovery, cure for hypertension and lack of hormonal recurrence, make LA an effective option in PCC.

INTRODUCTION

Pheochromocytoma (PCC) is a catecholamine-secreting tumor arising from chromaffin cells of the adrenal medulla and extra-adrenal sites. About 10% of this rare tumor is found in pediatric age group (mean age between 6-14 years). Most PCCs are sporadic, but can occur as a heritable and family disorder associated with multiple endocrine neoplasia (MEN) 2 or Neuroectodermal syndromes such as von Recklinghausen disease, von Hippel-Lindau disease and Sturge-Weber syndrome.

Perioperative management of pheochromocytoma is a challenging matter and requires a multidisciplinary approach for optimal care and successful outcome [1]. Surgical resection remains the mainstay treatment of symptomatic and medically refractory PCC [2]. The operative risk comes from hypertensive spikes and arrhythmias due to painful stimulations (endotracheal intubation, surgical incision) and mass manipulation. On the other hand, it was reported the risk of hypotension after mass resection (related to lack of catecholamine). Alpha-blocker therapy (doxazosin) almost 10 days earlier the surgery is required [3]. Adequate intravenous volume expansion is necessary to avoid hypotensive events during the anesthesia and after tumor resection. Volume expansion should to be initiated 24 hours prior to the surgical procedure to obtain a hemodynamic stability. In the postoperative period, blood pressure monitoring is important.

Laparoscopic adrenalectomy (LA) has been accepted as the “gold standard” treatment modality for small-sized (<6 cm) PCC and has been used for resection of large-sized (>6 cm) PCC [4]. Out of 173 adrenalectomies that we had done in the last 40 years, just 6 cases were for PCC. We report two of these cases, which we have treated with laparoscopic adrenalectomy.

CASE REPORT

Case 1

A 15-year-old boy suffering from Neurofibromatosis type 1 with

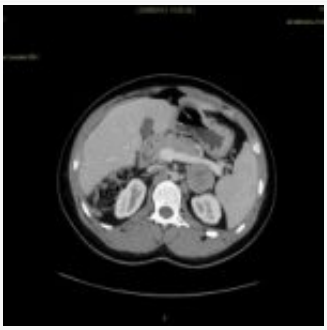

Figure 1 – CT scan shows a left adrenal pheochromocytoma

one-year history of headache, nausea and palpitations. High blood pressure was never documented. A 24-hour urine collection revealed: vanillyl mandelic acid 9.5 mg (normal range 1.8-6.7 mg); norepinephrine 74.4 µg/L (12.1-85.5 µg/L); epinephrine 8.7 µg/L (1.7-22.4 µg/L); plasma norepinephrine 2690.2 ng/L (95.0-446.0 ng/L) and plasma epinephrine 188.6 ng/L (<67 ng/L). Echocardiogram and ECG were unremarkable. Subsequently, abdominal ultrasonography revealed a 48 x 35 mm left adrenal nodular mass. Magnetic resonance imaging (MRI) and CT scan of the abdomen showed a 44 x 42 x 58 mm lesion at the left adrenal region with preserved cleavage planes with splenic vein, pancreatic tail and left kidney (Figure 1). After diagnostic evaluation, he was given doxazosin and bisoprolol for 10 days preoperatively. Intravenous hydration with normal saline was started 24 hours prior to the surgery.

Laparoscopic left adrenalectomy was performed without complications. Induction of general anesthesia was done with propofol and maintained with sevoflurane. A urinary catheter, nasogastric tube, arterial line and central venous catheter for monitoring of central venous pressure were placed. The patient was positioned in supine position slightly turned to his right side. A 30° optic lens was introduced through an umbilical 10 mm port. Other three 5 mm ports were placed: two working ports in both flanks and one insubxiphoid region. The spleen and the pancreatic tail were mobilized and reflected anteromedially, exposing the left adrenal gland. Left adrenal vein was identified and secured with clips. The mass was dissected using 5 mm LigaSure^{TM}, placed into a 10 mm Endobag^{TM} and exteriorized through the umbilical port (Figure 2). The excised mass was 6 x 5 x 4 centimeter. An abdominal drain was placed. Intraoperative blood loss was insignificant. During operation, the patient had only one hypertensive spike (during dissection), treated by sodium nitroprusside. Total operating time was 210 minutes.

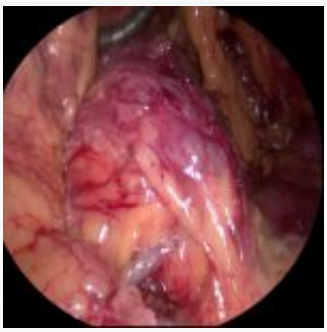

Figure 2 Laparoscopic adrenalectomy of the left adrenal pheochromocytoma

Postoperatively, the patient was admitted to the pediatric intensive care unit and transferred to the floor after 24 hours. Nasogastric tube was removed on postoperative day 2; while urinary catheter and abdominal drain were removed on postoperative day 3. The patient had an initial instability of blood pressure, which necessitated the use of bisoprolol, and resolved before discharge. Psychological support was rendered to the patient and his household by the psychology team. The patient was discharged on the fifth postoperative day.

A 13-year-old girl with a 2-year history of hypertension (140/90 mmHg). A 24-hour holter blood pressure monitoring revealed an average systolic pressure of 170 mmHg and diastolic pressure of 104 mmHg. Abdominal ultrasonography showed a nodular mass measuring 3,7 x 3,1 cm in the right adrenal region. She was started on an ACE-inhibitor

but with no improvement, therefore shifted into amlodipine. MRI scans showed a 4 x 3.8 cm right adrenal mass (Figure 3). PCC was confirmed by MIBG scintigraphy and by the highest values of 24-hour urinary catecholamines. The patient was given doxazosin and bisoprolol for 9 days, and intravenous hydration with normal saline was initiated 24 hours before surgery.

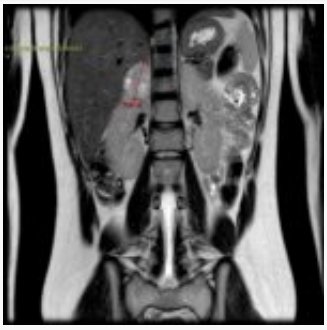

Figure 3 a) MRI scan shows a right adrenal pheochromocytoma. b-d) Laparoscopic adrenalectomy of the right adrenal pheochromocytoma.

Laparoscopic right adrenalectomy was performed without complications. Anesthetic technique and agents, patient position, and trocars placement were all the same as in the first case. Arterial and central venous catheters were also placed. Insufflation was performed through the umbilical port with a pressure of 8-10 mmHg. It was not necessary to mobilize the hepatic flexure. The mass was isolated from the peritoneal adherences and the surrounding fat using 5 mm LigaSure^{TM}, placed into a 10 mm Endobag^{TM} and exteriorized through the umbilical port (Figure 3). A drain was placed in the right adrenal region. Intraoperative blood loss was insignificant. During the procedure, the patient had only one hypertensive spike associated with tachycardia (at the beginning of dissection), treated by a beta-blocker. Total operating time was 100 minutes.

Postoperatively, the patient was admitted to the pediatric intensive care unit and transferred to the floor after 24 hours. The postoperative period was uneventful. Nasogastric tube was removed on postoperative day 1; urinary catheter and abdominal drain were removed on postoperative day 2 and day 5, respectively. Blood pressure was normal during the hospital stay and she did not require any anti-hypertensive therapy. Psychological support was given to the patient and her family by the psychology team. The patient was discharged on the fifth postoperative day.

DISCUSSION

Laparoscopic adrenalectomy has wide acceptance as the “gold standard” treatment modality for small-sized (< 6 cm) PCC [4]. Reports of surgical treatment of PCC that are greater than or equal to 6 cm have demonstrated significant associations with longer operative times, greater intraoperative blood loss, higher conversion rates and more hypertensive crises than the surgeries to treat PCC less than 6 cm [5]. However, some studies have found no significant correlation between the size of PCC and the severity of an intraoperative hemodynamic event [6].

In our experience with adrenalectomies for diverse adrenal pathologies, we had performed 173 adrenalectomies in the last 40 years, of which, we had operated 161 in open technique and 12 by laparoscopic technique. Out of all these cases, only 6 were PCCs, where 4 were treated by

Table 1 Demographic data, surgical approach, intraoperative monitoring parameters and operative time of our adrenalectomies for PCC over the last 40 years

open adrenalectomy (OA) and 2 by LA (Table 1). LA for PCC is a technically challenging procedure and puts patients at high risk of LA-related complications and death; however, the pathology of PCC alone does not increase LA-associated morbidity [4]. When LA is applied to larger sized adrenal tumors, there is a high risk of conversion to OA and of surgery-related morbidities. Conversion to OA is the major intraoperative complication of LA. Compared with other adrenal gland tumors, PCC has been associated with a lower conversion rate (by approximately 10%). Tumor size is a primary, independent risk factor leading to conversion [7]. Perry et al. reported a higher likelihood (37.6%) of conversion to OA in patients with greater than or equal to 6 cm PCC compared with those with less than 6 cm PCC (9.1%) [6]. A previous study showed that larger tumor size alone was not necessarily associated with a higher risk of conversion [8]. The most frequent incidental cause of conversion has been reported as iatrogenic vascular injury, while a larger sized PCC is more likely to involve major vessels, such as the renal and splenic veins. Another common cause of conversion is intraoperative hemodynamic instability [5]. In our cases, either the OA or the LA groups, tumor size was <6 cm and none of our two LA cases required conversion or had any intra- or postoperative complication.

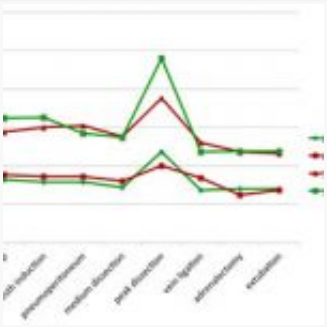

Figure 4 Comparison of average systolic and diastolic blood pressures during VLS versus Open approach.

Carbon dioxide pneumoperitoneum can result in respiratory acidosis, hypercapnia and cardiovascular instability [9]. Iatrogenic acidosis stimulate the release of catecholamines into the circulation and causes acute bouts of hypertension [10]. The largest increase in circulating catecholamines occurs during tumor manipulation regardless of the use of LA or OA [11]. Some studies have reported that the episodes of hemodynamic instability with LA in PCCs are equal to or lesser than during open surgery [12].This could be because there is minimal handling of the tumor with early control of the adrenal vein in LA in comparison to OA. Clinically, significant hemodynamic alterations during manipulation were more commonly encountered during conventional OA than during LA [13]. It has been suggested that a low intra-abdominal pressure of 8-10 mmHg causes less catecholamine release and fewer hemodynamic fluctuations [14]. In our cases, it was obvious that number of hypertensive spikes, heart rate and average systolic and diastolic blood pressures were a bit more with the LA group as demonstrated in (Table 1) and (Figure 4). However, this finding could not be statistically significant due to the low number of cases, and we should bear in mind our early experience in LA. In the contrary, LA offers some advantages over OA, such as minimal invasiveness, less intraoperative blood loss, and faster postoperative recovery. The most important point is to do minimally-invasive adrenalectomy for well-encapsulated masses without image-defined risk factors [15]. LA is associated with a smaller incision, shorter operative time, less intraoperative bleeding, a lower incidence of postoperative intestinal

obstruction, faster postoperative recovery regardless of PCC size [4, 16].

LA for tumors larger than 6 cm is controversial because of the perceived increased risk of malignancy. This controversy continues despite the fact that no data support the assumption that, when performed correctly, laparoscopic complete adrenalectomy is inferior oncologically to open adrenalectomy for malignant PCC [17].Differentiation between benign and malignant PCC is not possible preoperatively, even by histopathology, without evidence of local invasion, vascular invasion, or metastases [17]. A laparoscopic exploration can be a useful tool to determine the extent of the disease and estimate the resectability. Kravarusic et al. recommend at least diagnostic laparoscopy for cases of intra-abdominal, extra-adrenal pheochromocytoma/paraganglioma for identifying the exact tumor origin, relation to surrounding structures and vascular elements with appropriate estimation of laparoscopic resectability [18]. Currently, the choice of surgical approach for PCCs with the apparent local disease is dictated by tumor size, absence of signs of invasion or metastases, and surgeon experience [19].

CONCLUSION

Laparoscopic adrenalectomy is an effective and safe treatment for small-sized pheochromocytoma (< 6 cm) in children. Under adequate anesthetic control, PCC could be safely managed through a minimally invasive approach. The laparoscopic approach allows a minimal handling of the tumor with early control of the adrenal vein in comparison to open surgery. In literature, clinically significant hemodynamic alterations were more common during OA than during LA. In our cases, hemodynamic alterations were a bit more in the LA cases which could not be of statistical significance due to the low number of cases, considering also our early experience in LA.However, LA was associated with less intraoperative bleeding and shorter length of hospital stay, in addition to its advantages of low morbidity, fewer complications, quick recovery, cure of hypertension and lack of hormonal recurrence, which make LA an effective option in PCC.

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