Laparoscopic Resection of an Interaortocaval Paraganglioma: Diagnosis Following a Needle Biopsy

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ABSTRACT

We present the case of a 45-year-old male with an interaortocaval paraganglioma. The tumor was incidentally diagnosed during magnetic resonance imaging (MRI) of the lumbosacral spine to investigate back pain. The MRI was followed by fine needle aspiration cytology that confirmed the diagnosis. The patient, however, did report a history of intermittent episodes of headaches, increased sweating, palpitations, and elevations in blood pressure. The tumor was excised laparoscopically without complications. Needle biopsy of a retroperitoneal mass that could be a paraganglioma must be avoided.

Key Words: Paraganglioma, Extra-adrenal, Retroperitoneal neoplasm, Laparoscopy.

INTRODUCTION

Paragangliomas are often referred to as extraadrenal pheochromocytomas. They present with many of the same features as adrenal pheochromocytomas including paroxysms of headache, sweating, and palpitations. Blood pressure is often elevated, and hypertensive crises can occur. These symptoms are due to the secretion of catecholamines by the tumor. In the past, these tumors have been resected by open surgery. Because of the advances in localization of these tumors by computed tomographic (CT), magnetic resonance imaging (MRI), and 123I-metiodobenzylguanidine (MIBG) scans, laparoscopy is increasingly being used.

CASE REPORT

A 45-year-old Caucasian male was investigated with a lumbosacral MRI for chronic backache. MRI revealed an incidental mass in the aortocaval region. During fine needle aspiration cytology of this mass, the patient developed severe hypertension. Pathology confirmed paraganglioma. The patient also reported intermittent symptoms of severe headaches occurring approximately once per week, excessive sweating, palpitations, and elevations in blood pressure. His blood pressure was 140/90. The diagnosis was confirmed by a plasma normetanephrine level of 4.26 nmol/L (upper limit of normal level is 0.66 nmol/L) and by (MIBG) scan that showed an increased uptake in the mid abdomen (Figure 1). CT revealed a 2.4x2.7x2.2-cm interaortocaval soft-tissue density mass at approximately the L2-L3 level without invasion of the aorta or inferior vena cava (IVC) (Figure 2). The patient’s past medical history is significant for lumbar disk disease, melanoma, and seizures. Laparoscopic excision of the neoplasm was planned.

The patient was prepared for surgery with alpha-adrenergic blockade with doxazosin. At surgery, pneumoperitoneum was established with a Veress needle. A 10-mm trocar was inserted just below the level of the umbilicus in the midclavicular line. Another 12-mm trocar was inserted at the umbilicus. Three 5-mm trocars were also inserted: midline in the epigastrium to retract the liver, at the lateral border of the rectus abdominis mid way between the
umbilicus and the costal margin, and at the anterior axillary line in the subcostal region.

The patient was in a left lateral position. The right colon and the duodenum were mobilized to expose the inferior vena cava (IVC). The right and left renal veins were dissected. The IVC was dissected from the left renal vein caudally. The mass was then exposed between the IVC and aorta with care taken not to manipulate the tumor initially, to minimize intraoperative hypertension. The IVC and the tumor were mobilized and, 10-mm and 5-mm Hem-o-lok (Weck-Teleflex, Medical Research Triangle Park, NC) clips placed on the venous tributaries from the tumor. Dissection was then carried out between the aorta and the tumor.

Figure 1. $^{123}$I-metaiodobenzylguanidine (MIBG) scan showing increased uptake in the mid abdomen at the L2-L3 level.

Figure 2. Axial (A) and coronal (B) views from a contrast enhanced computed tomographic (CT) scan revealing the position of the tumor between the inferior vena cava and aorta without evidence of invasion.
and the IVC, which mobilized the tumor from the interaortocaval region. Finally, the tumor was mobilized from the aorta. Hem-o-lok clips were again used to control the multiple arteries feeding the tumor. The mass was placed in a specimen retrieval bag and removed through the umbilical incision. During mobilization of the tumor, there was significant elevation of the patient’s blood pressure with a maximum BP of 220/90. The blood pressure was controlled by the anesthesiologist with a total dose of 50 mg of Labetolol and 1.5 mg of Nicardipine hydrochloride. A few hours following surgery, the patient produced some coffee-ground emesis. He was not allowed anything by mouth, and on postoperative day 1 (POD 1) an esophagogastroduodenoscopy was performed that revealed erosions in the stomach with no signs of recent bleeding. He was started on a proton pump and inhibitor. On POD 2, the patient was advanced to a regular diet and discharged home without further complications.

One month after surgery, the headaches had not recurred. The patient’s plasma metanephrine and normetanephrine levels were 0.15 and 0.72, respectively. These levels are within normal limits.

**DISCUSSION**

Paragangliomas are catecholamine-secreting tumors that are also known as extraadrenal pheochromocytomas. They develop from chromaffin cells of sympathetic paraganglia, which are derivatives of the neural crest.1,2 The most common sites for these tumors include the carotid body, jugular foramen, mediastinum, organ of Zuckerkandl, and the periaortic region but can occur anywhere paraganglia are present.3 They occur more often in men and usually in the fourth or fifth decade of life but can occur at any age.4,5 They can be either functional or nonfunctional in nature, and are functional in the majority of cases of retroperitoneal paragangliomas.5 When functional, they produce symptoms identical to adrenal pheochromocytomas including paroxysms of hypertension, diaphoresis, headaches, tachycardia, and palpitations secondary to increased circulating catecholamines.1,6 Most paragangliomas are sporadic in nature, but approximately 10% of these tumors may be associated with genetic disorders, such as familial paraganglioma, neurofibromatosis Type 1, von Hippel-Lindau, Carney triad, and multiple endocrine neoplasia (MEN) Type 2.1,5 When compared with adrenal pheochromocytomas, paragangliomas are more likely to be malignant, multicentric, and metastatic, with 29% to 40% being malignant at the time of diagnosis.7 There is no definitive test to differentiate benign from malignant tumors. Often, malignancy can only be confirmed by evidence of metastatic disease.2

The diagnosis of a catecholamine-secreting tumor can be made by a select number of tests. Fractionated plasma catecholamines and metanephrines, along with 24-hour urine catecholamines, will be elevated in the vast majority of cases.6 Localization of the tumor is accomplished by imaging studies. CT and MRI both have high sensitivity, but are not specific to paraganglioma. MIBG scintigraphy is less sensitive than CT or MRI, but is highly specific for paragangliomas and adrenal pheochromocytomas.7 MIBG can also differentiate functional from nonfunctional tumors and identify metastases.3 When all 3 modalities of imaging are used in concert, the accuracy of localization has been reported to be 97%.8

The diagnosis of our patient was made by a needle biopsy. This is unusual and in retrospect unnecessary and potentially dangerous because of the severe hypertensive episode that followed. This report underscores the importance of a thorough history and biochemical evaluation to rule out a paraganglioma, rather than proceeding with a biopsy.

Definitive treatment of paragangliomas and pheochromocytomas is by surgical resection. Classically, paragangliomas have been removed by a transabdominal approach. Posterior and flank approaches and transabdominal laparotomy have been utilized for open surgery.8 Laparoscopic surgery via the transperitoneal or retroperitoneal approaches has been used for retroperitoneal paragangliomas.9 Some of the reported benefits of laparoscopic resection over open surgery include shortened hospital stay, less postoperative pain, decreased adhesion formation, better cosmesis, and lower complication rates.10,11 Another benefit of laparoscopy is that the magnified view provided by the endoscope can theoretically improve operative precision.9 However, preoperative imaging of the retroperitoneal vessels is important, and conversion to open surgery may be occasionally required. Prior to any surgical manipulation of the tumor, preoperative α-, sometimes followed by β-, adrenergic blockades should be performed to aid in preventing intraoperative hypertensive crises.1

**CONCLUSION**

Paragangliomas are rare tumors that can occur in multiple locations and can have severe consequences if not removed in a timely manner. Needle biopsy of a retroperitoneal mass that could be a paraganglioma must be
avoided. They have classically been treated by open surgical resection. With advances in localization, laparoscopy has become a viable solution to the treatment of these tumors with fewer adverse side effects compared with that of open surgical resection.

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