Complete laparoscopic excision of a giant retroperitoneal paraganglioma

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ABSTRACT

Paragangliomas (or 'extra-adrenal pheochromocytomas') are rare tumours arising from neural crest cells. They occur most commonly in the abdomen along the paraspinal sympathetic ganglion. The malignancy rate can be up to 35% and surgical resection is the recommended treatment. While laparoscopic excision of pheochromocytomas is now well established, the overall number of cases of paragangliomas reported is much smaller owing to their rarity (even more so for giant paragangliomas of ≥ 8 cm) and controversy remains over the completeness of excision. Furthermore, the risk of malignancy and recurrence rate are higher in paragangliomas, with incomplete excision giving rise to recurrent endocrinopathy and carcinomatosis. We report the case of a 26-year-old woman who underwent laparoscopic excision of an 8.2cm functioning paraganglioma. Complete resection of the tumour was achieved and the patient's symptoms resolved. Twelve months later, she remains well with no evidence of recurrence.

KEYWORDS

Paraganglioma – Laparoscopic excision – Extra-adrenal pheochromocytoma – Retroperitoneal

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Case history

A 26-year-old woman attended the emergency department complaining of acute left-sided chest pain, palpitations and dizziness. She had experienced similar episodes previously but these were self-limiting. At presentation, her blood pressure was 187/128mmHg and her heart rate was 118bpm. Troponin was elevated at 539ng/L, and electrocardiography demonstrated sinus tachycardia and T wave inversion. She was admitted with a diagnosis of myocarditis. On further questioning, it was discovered that she had presented to her general practitioner several times over a six-month period complaining of unintentional weight loss, general lethargy, headaches and hot flushes, and was found to be hypertensive. During her admission, she underwent further cardiac investigations including 24-hour electrocardiography, echocardiography and cardiac magnetic resonance imaging but these were all normal.

Computed tomography of the chest, abdomen and pelvis demonstrated a round, well defined, heterogeneously enhancing lesion within the porta hepatis and adjacent to the head of the pancreas (Fig 1). Both the gallbladder and pancreas were displaced by the mass. The differential diagnoses of soft tissue sarcoma, neuroendocrine tumour and Hodgkin's lymphoma were considered. Subsequently, the patient underwent ultrasonography guided biopsy and endoscopic ultrasonography to clarify the location of the tumour relative to the duodenum and pancreas. Biopsy confirmed a

paraganglioma (PG). The patient's 24-hour urinary catecholamine level was also found to be elevated. After discussion at our multidisciplinary team meeting, a plan was made for laparoscopic excision of the retroperitoneal tumour by the senior laparoscopic surgeon in our department (LRJ).

Preoperative blood pressure was controlled with alpha and beta-blockade (phenoxybenzamine and propranolol). The patient was initially positioned supine and the first 12mm trocar was inserted below the umbilicus using the open Hasson technique. A 30° laparoscope was inserted and carbon dioxide insufflated to achieve an intra-abdominal pressure of 12mmHg, and a further port was inserted in the epigastrium. Three additional 11mm ports were inserted, all on the right side (the side of the tumour). One was inserted just below the costal margin lateral to the rectus abdominis muscle, one in the right flank and one in the pelvis, again lateral to the rectus abdominis muscle.

Dissection was performed using monopolar hook diathermy and the LigaSure™ ligation system (Medtronic, Minneapolis, MN, US) (Fig 2). The overlying peritoneum was initially incised and the lesion was freed from the retroperitoneal fat. Circumferentially, the surrounding feeding vessels were dissected, and individually ligated and divided between Hem-o-lok® clips (Teleflex, Morrisville, NC, US) or using the LigaSure™ device directly. During the dissection, the patient's position was changed intermittently to allow the effects of gravity to aid in the procedure. The specimen was removed in one piece using an Endo Catch™ pouch

(Medtronic) and delivered through a Pfannenstiel incision, and a subhepatic drain was placed. Blood loss was 40ml and the operating time was 120 minutes.

The patient's postoperative course was uneventful and she was discharged on day 5. Histology confirmed a retroperitoneal PG measuring 8.2cm × 7.1cm × 5.1cm, completely excised with clear margins. At 12 months following the procedure, she remains well with no evidence of recurrence.

Discussion

PGs are rare tumours arising from extra-adrenal paraganglionic cells of the sympathetic nervous system. They are classified as functioning or non-functioning tumours, defined by the production of catecholamine, hence the alternative name of 'extra-adrenal pheochromocytoma'. Patients with functional PGs may present with symptoms of headache, palpitations and diaphoresis. They make up 15% of all phaeochromocytomas (PCCs) in adults.¹ While they may arise at any site in the paraganglion system, they occur most commonly below the diaphragm, in the para-aortic region above the lower renal poles. Complete surgical excision is the treatment of choice for primary tumours as well as recurrence or malignancy.

Initial concerns surrounding the use of laparoscopic surgery for PCCs were based on increased risk of cardiovascular complications, due to intraoperative hypertensive crises brought on by insufflation of carbon dioxide leading to an increase in intra-abdominal pressure and hormone release. However, this has been evaluated in several studies and laparoscopic excision of PCCs is now well established. Despite

GV = gonadal vessels; PG = paraganglioma

Figure 2 Laparoscopic approach for resection of a paraganglioma: exposure of the liver hilum and adjacent paraganglioma (A); dissection of overlying peritoneum using hook diathermy (B); gonadal vessels adjacent to paraganglioma (C); identification of pedicle of paraganglioma (D); division of pedicle (E); and retrieval of paraganglioma and placement in an Endo Catch™ pouch (F)

tumour size being a factor influencing the failure of the laparoscopic approach and a higher recurrence rate after laparoscopic excision, it has been advocated even for large PCCs in the hands of an experienced laparoscopic surgeon.

In comparison, with an incidence of PGs in the general population of between 0.005% and 0.1%,² the amount of data on the outcomes of laparoscopic resection of PGs is limited. PGs have a higher risk of recurrence than PCCs³ and the rate of malignancy is also higher (35% vs 10%)⁴ so long-term follow-up for these tumours is essential. In addition, PGs can present greater difficulty in resection by appearing in unusual and surgically inaccessible locations, resulting in incomplete tumour resection or tumour spillage. While laparoscopic surgery has been advocated for the resection of small PGs,^{2,5} there are very few reports on the outcomes of minimally invasive excision of giant sized tumours (≥8cm). To our knowledge, there have only been three published cases of laparoscopically resected giant PGs,⁶⁻⁸ including one that was part of a case series evaluating robot assisted laparoscopic resection.⁸

Conclusions

Our report is the first to demonstrate microscopically complete excision of a PG via the laparoscopic route with a good

long-term outcome. The case reported here adds to the evidence that complete laparoscopic excision can be achieved for giant PGs with good perioperative and long-term outcomes when carried out by an experienced laparoscopic surgeon.

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