

## A Not So Simple Retroperitoneal Mass-A Clue from Hypertensive Crisis

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### Abstract

Extra adrenal paragangliomas are tumours of chromaffin cells arising from the sympathetic and parasympathetic divisions of the autonomic nervous system. About 75% of them are located between the origin of the inferior mesenteric artery and bifurcation of the abdominal aorta known as the organ of Zuckerkandl. Their clinical presentations are varied and are mostly due to the excessive secretion of catecholamines by the tumour. The incidence of the classic triad of Headache, Palpitations and Diaphoresis is seen in as little as 24% of cases. A recent review reports hypertension to be paroxysmal in 48%, persistent in 29% and absent in 13 % of cases. About 49% of cases are incidentally diagnosed during abdominal imaging. These tumours may be confused with other common retroperitoneal neoplasms, especially when the patient is asymptomatic. We report a severe hypertensive crisis in such an asymptomatic patient, which was triggered on induction of anesthesia and manipulation of the tumour and its successful management in a patient with undiagnosed paraganglioma.

**Keywords:** Retroperitoneal tumour; Paraganglioma; Extra adrenal; Asymptomatic

### Case Report

A 28 years old lady was scheduled for resection of a retroperitoneal tumour. The tumour was incidentally diagnosed two years ago during a routine antenatal scan and the patient was advised to return for follow-up after delivery. She subsequently underwent caesarean section uneventfully under spinal anesthesia. Now she presented for the follow-up of the mass detected on her antenatal scan. A CT scan done revealed a 59x65x67 mm well defined soft tissue density mass lesion anterior to the right iliac vessels with an intense contrast enhancement suggestive of a retroperitoneal tumour. There was no history of hypertension, headache, perspiration, syncope, pain abdomen or pregnancy induced hypertension. Her vital parameters were normal during pre-anesthetic assessment with a blood pressure of 110/80 mmHg, pulse rate of 90/minute and all the preoperative blood investigations and ECG were within normal limits.

On the day of surgery, her base line blood pressure was 120/80 mmHg and pulse rate was 100/min. A wide bore intravenous access was secured and standard monitoring was established. An epidural catheter was secured and fixed at L<sub>2</sub>-L<sub>3</sub> inter-vertebral space. Anesthesia was induced with Propofol 100 mg and Fentanyl 100 mcg. During mask ventilation following administration of vecuronium 6 mg, the tumour was palpated per abdominally by the surgeon and instead of an expected fall following induction of anesthesia, there was a dramatic rise in blood pressure to 225/158 mmHg and HR to 120/min. Labetalol 5 mg and lidocaine 100 mg IV were given and anesthesia was deepened with sevoflurane and the trachea was intubated after the blood pressure reduced to 150/90 mmHg. Anesthesia was maintained with nitrous oxide in oxygen (50:50) and sevoflurane and intermittent boluses of vecuronium as required. Epidural was activated with 10 ml of 0.25% bupivacaine followed by continuous infusion at 8 ml/hr. An arterial line was soon secured. Over the next 30 minutes, her haemodynamic parameters remained within normal limits. During tumour manipulation the blood pressure again rose to 250/150 mmHg and the HR to 120/min. Despite deepening anesthesia with increasing concentrations of sevoflurane and the addition of dexmedetomidine infusion, the blood pressure remained high in the range of 180-200 systolic, 100-110 diastolic with a heart rate of 120-125/min. SpO<sub>2</sub>, EtCO<sub>2</sub>, airway pressure, temperature and ECG were

within normal limits. Peripheral pulses were feeble. At this juncture, a diagnosis of a secretory neuroendocrine tumour was entertained and in consultation with the surgeon it was decided to proceed with the surgery with additional measures to control the haemodynamic changes. It was extremely surprising that she underwent an LSCS uneventfully and her anesthetic record, which was summoned later, was a testimony to this. Central venous access was secured via the right internal jugular vein and labetalol in aliquots of 5 mg was given up to a total dose of 20 mg. Simultaneously a bolus of 6ml of 0.5% bupivacaine was administered through the epidural catheter. Following the above measures, the blood pressure and heart rate reduced to 160/90 mmHg and 100/min respectively. Blood glucose levels were checked and were found to be within normal limits. The tumour showed tortuous neovascularisation from the inferior mesenteric vessels. As the tumour could not be separated from the vascular supply of the sigmoid colon, sigmoidectomy and end to end anastomosis were performed and an ileostomy was fashioned. Tumour resection time was about 1 hour after which the blood pressure dropped to 60/30 mmHg. Epidural infusion was immediately discontinued and aggressive fluid resuscitation was instituted with 1L each of crystalloids and colloids to attain a CVP of 4-5 mmHg. Repeated doses of mephenteramine and phenylephrine in aliquots of 6mg and 100 mcg were given but the blood pressure showed little response. Then, noradrenaline 0.2 mcg/Kg/min infusion was started following which the blood pressure rose to 90/60 mmHg. The total estimated blood loss was about 300 ml. Following completion of surgery, the patient was electively ventilated for 1 hour, during which the haemodynamics remained stable with noradrenaline support. Residual neuromuscular blockade was reversed and the patient was extubated. An immediate echocardiogram ordered in the PACU showed moderate LV dysfunction with an EF of 0.4, following which dobutamine was added to the existing noradrenaline infusion. The inotropes

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were gradually tapered and discontinued on the second postoperative day. An echocardiogram done on the third post operative day revealed good LV function with an EF of 0.6. Blood glucose levels were normal throughout the post-operative course. On histological examination, the specimen showed thickly encapsulated endocrine tumour made up of large polypoidal cells with abundant granular cytoplasm and vesicular nuclei with rich arborising network of thin walled vessels adjacent to the clusters of tumour cells, which was in favour of a paraganglioma.

## Discussion

Paraganglioma was the name coined by Mascorro and Yates to describe a rare neuroendocrine neoplasmarising from neural crest tissue that develops into sympathetic and parasympathetic paraganglia throughout the body. Pheochromocytoma is the paraganglioma located in adrenal medulla. Extra adrenal paraganglia are located outside the adrenal gland. The system is vital in fetal development until the formation of the adrenal medulla, as a source of catecholamines. The function of chromaffin cells is similar to those in the adrenal medulla. About 70% of sympathetic paragangliomas are intraabdominal, usually found in the perinephric and paraaortic spaces [3,4]. The remaining 30% are located in the chest [3,4]. The organ of Zuckerkandl normally enlarges during pregnancy due to increased demand and probably this led to the detection of a retroperitoneal mass on the antenatal scan in our patient [5]. She was advised that the mass could be tackled following her delivery. The patient had no symptoms whatsoever throughout her pregnancy and indeed had an uneventful LSCS. Two years later she came for a follow up and was advised surgical excision.

About 75% of paragangliomas are sporadic and the remaining 25% are hereditary, but there was no suggestive family history in our patient to point to a hereditary cause [6-8]. The incidence of multicentricity for this tumor is approximately 10% of the total cases [9]. Approximately 10% of paragangliomas are clinically silent and detected incidentally at imaging study during evaluation of patients with unrelated symptoms [9].

Symptoms of catecholamine excess can be spontaneous or induced by strenuous physical exertion, trauma, labor, induction of general anesthesia and tracheal intubation, increased intra-abdominal pressure associated with cough, insufficient muscle relaxation, abdominal pressure changes due to the initiation of mechanical ventilation, creation of pneumoperitoneum, direct manipulation of tumour either during surgery or even during FNAC. In our case the initial rise in blood pressure could be due to abdominal pressure changes during induction of anesthesia and mask ventilation or due to per abdominal manipulation of the tumour by the surgeon following induction of anesthesia. The second and persistent rise in blood pressure was due to direct manipulation of the tumour by the surgeon. These intraoperative hypertensive responses are empirically attenuated by increasing the anesthetic depth and muscle relaxation, administration of vasodilators and  $\beta$ -blockers along with careful surgical handling of tumor tissue. Continuous monitoring of arterial pressure via an indwelling catheter is universally recommended as hypertensive crisis can lead to cardiac arrhythmias, myocardial infarction, stroke and death. Blood glucose levels when elevated are primarily due to stimulation of lipolysis and epinephrine induced glycogenolysis and gluconeogenesis in the liver [10].

As the majority of paragangliomas secrete predominantly the  $\alpha$ -agonist norepinephrine,  $\alpha$ -adrenergic antagonists are the mainstay of hypertensive control and preoperative pharmacological preparation is similar to that of pheochromocytoma.

Hypotension following tumour removal is definitively controlled within a few hours and there are anecdotal reports of patients requiring vasopressors for 48 hours. Blood glucose concentration should be monitored for at least 24 hours. Excessive rebound secretion of insulin may occur after removal of the tumor accounting for rare transient hypoglycemia reported up to 6 days postoperatively [10].

## Conclusion

Asymptomatic retroperitoneal tumours may be paragangliomas and although asymptomatic, can turn out to be functional on the operating table and surprise the anesthetist and surgeon alike. Hence complete biochemical workup to rule out paragangliomas and multiple tumours and metastasis is vital. Invasive haemodynamic monitoring is mandatory to promptly deal with haemodynamic perturbations from the secreting tumours. When hypervascular masses are seen in specific locations of the body, the possibility of paragangliomas should always be considered.

## References

1. Shinn HK, Jung JK, Park JK, Kim JH, Jung IY, et al. (2012) Hypertensive crisis during wide excision of gastrointestinal stromal cell tumor (GIST): Undiagnosed paraganglioma -A case report. *Korean J Anesthesiol* 62: 289-292.
2. Hee-Young Kim, Joo-Yun Kim, Hae-Kyu Kim, Seung-Hoon Baek (2013) Hypertensive Crisis during Removal of Retroperitoneal Mass in a Patient with Undiagnosed Paraganglioma, A Case Report. *Korean J Crit Care Med* 28: 64-66.
3. Pandurengan K, NatarajanR, Marudavanan R, Sowmya T, Raja Chidambaram K (2011) An Unusual Retroperitoneal Mass-Non Functioning Retroperitoneal Paraganglioma. *Internet Journal of Endocrinology* 6.
4. Sangster G, Do D, Previgliano C, Li B, La France D, et al. (2010) Case Report Primary Retroperitoneal Paraganglioma Simulating a Pancreatic Mass: A Case Report and Review of the Literature. *HPB Surg*.
5. Dossett LA, Rudzinski ER, Blevins LS, Chambers EP Jr (2007) Malignant pheochromocytoma of the organ of Zuckerkandl requiring aortic and vena caval reconstruction. *Endocr Pract* 13: 493-497.
6. Neumann HP, Bausch B, McWhinney SR, Bender BU, Gimm O, et al. (2002) Germ-line mutations in nonsyndromicpheochromocytoma. *N Engl J Med* 346: 1459-1466.
7. Amar L, Bertherat J, Baudin E, Ajzenberg C, Bressac-de Paillerets B, et al. (2005) Genetic testing in pheochromocytoma or functional paraganglioma. *J Clin Oncol* 23: 8812-8818.
8. Jiménez C, Cote G, Arnold A (2006) Review: Should patients with apparently sporadic pheochromocytomas or paragangliomas be screened for hereditary syndromes? *J Clin Endocrinol Metab* 91: 2851-2858.
9. Lee KY, Oh YW, Noh HJ, Lee YJ, Yong HS, et al. (2006) ExtraadrenalParagangliomas of the Body: Imaging Features. *AJR Am J Roentgenol* 187: 492-504.
10. Lentschener C, Gaujoux S, Tesniere A, Dousset B (2011) Point of controversy: perioperative care of patients undergoing pheochromocytoma removal-time for a reappraisal? *Eur J Endocrinol* 165: 365-373.