

## Complete excision of large invasive retroperitoneal paraganglioma mislabelled as hypertrophic obstructive cardiomyopathy

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### Abstract

Paraganglioma originates from chromaffin cells of adrenal medulla and autonomic paraganglia, which are derived from the neural crest cells. Paragangliomas are half as common as pheochromocytomas with 69% occurring in head and neck, 22% in abdomen and pelvis and 10% in the thorax. About 70% paragangliomas are sporadic, 30% are hereditary, having identifiable germline mutations of Succinate Dehydrogenase enzyme (SDH).

**Keywords:** Invasive Retroperitoneal Paraganglioma Hypertrophic Cardiomyopathy.

### Introduction

Pheochromocytoma and Paraganglioma originates from chromaffin cells of adrenal medulla and autonomic paraganglia, which are both derived from the neural crest cells. About 70% paragangliomas are sporadic, 30% are hereditary, having germline mutations of Succinate Dehydrogenase enzyme (SDH). Different paragangliomas have different presentations based on location and ability to secrete hormones. Head and neck paragangliomas constitute 70% of them; originate in parasympathetic tissue and invariably are non-secretory. Secretory paragangliomas originate from sympathetic nervous system and secrete catecholamines. They usually present as sustained or paroxysmal hypertension.<sup>1</sup> Reversible dilated or hypertrophic cardiomyopathy (sometimes mimicking features of HOCM), can also be a feature of this disease.<sup>2</sup>

We report an unusual case of invasive paraganglioma presenting with severe episodic hypertension; mislabelled as HOCM on the basis of echocardiographic findings, which led to a delay in establishing the correct diagnosis.

### Case Report

A 13 years old male, hypertensive for 3 years, diagnosed

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### Pre-Operative Picture :



### Post-Operative picture :

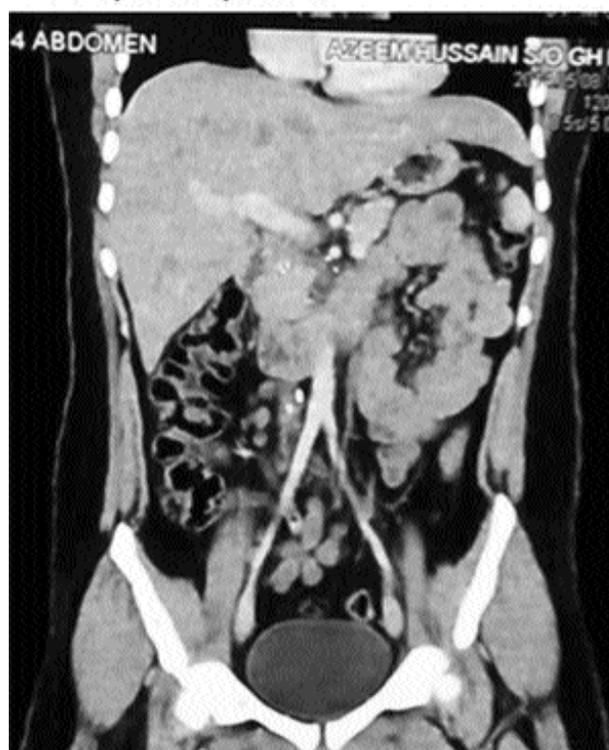


Figure: Pre- and post-operative view.

as HOCM for 1 year; presented to the hospital ER in February 2013, with complaints of altered consciousness and blurring of vision for 2 hours. His blood pressure was 220/110 mmHg and there was grade III ejection systolic murmur at left sternal border, as well as grade II pansystolic murmur at mitral area, radiating to axilla. Glasgow Coma Scale (GCS) was 5/15. A diagnosis of hypertensive encephalopathy was made. There were multiple previous ER visits with similar symptoms in other hospitals. There was no family history of hypertension.

Earlier an abdominal ultrasound done for vague abdominal pain, had revealed a retroperitoneal mass in para-aortic region. A percutaneous biopsy of this mass was done quite inappropriately, without any evaluation or awareness of hypertension, because of its episodic occurrence. Biopsy showed a neoplastic lesion arranged in nests of polygonal cells, surrounded by a rim of sustentacular cells. These polygonal cells had pleomorphic, rounded nuclei with coarse chromatin. Chromogranin A stain was positive in polygonal cells, S-100 was positive in flattened sustentacular cells on immune histochemistry. All these features were consistent with Paraganglioma. Moreover, an echo led to diagnosis of HOCM in a Cardiac Centre subsequently. The abdominal mass and biopsy were ignored by the patient's family.

During this admission hypertension was persistent in nature with acute elevations occurring especially in the morning hours and by a change of posture, in particular when he sat up or was in the supine position (Figure-1). Acute blood pressure elevations were also triggered by abdominal palpation. Due to persistent high blood pressure with episodic crises, patient had frequent bursts of arrhythmias, especially supraventricular tachycardia, requiring emergency management. As a result of these crises and the difficulty in finding a surgical team to operate, the patient remained admitted for 3 months.

During crises blood pressure was controlled with additional intravenous isosorbide di nitrate infusion at 1-2mg/hr. The same strategy was used in the peri-operative period. Oral alpha blocker (Prazocin 2mg x 6 hourly) and beta blocker (Propranolol 40mg x 8hourly) were used as background anti hypertensives.

Workup for secondary hypertension revealed elevated urinary VMA levels of 68.3 mg/24 hours (n <11.6 mg/24 hours). Urinary and Plasma metanephrine and nor metanephrine levels which have a higher diagnostic yield, are not available. Ultrasound and CT-scan of abdomen confirmed a large, retroperitoneal soft tissue mass, measuring 6.4 x 3.5 cm, involving Right ureter and Right renal vein, invading into the Inferior Vena Cava (IVC) with

tumour thrombus terminating just before IVC insertion into right atrium (Figure-1).

His thyroid function tests, parathyroid hormone (PTH), Calcitonin, Ultrasound neck and Renal Doppler were normal. Echo showed Moderate asymmetrically hypertrophied LV and interventricular septum (non-obstructive).

Due to the large tumour size, its invasion into IVC and its extent up to right atrium, it was a difficult decision for surgeons to operate on the patient, in view of very high risks involved. Multiple surgeons were approached and the case was considered in many tumour board meetings of different hospitals. All were of the view that tumour is unresectable, until a liver transplant surgeon was located in another city after 8 months of hard work, who after high risk consent attempted this surgery on 13th September 2013.

Complete excision of the tumour was done, which was involving Right ureter, Right Renal vein and IVC, with IVC tumour thrombus from the entrance into Right atrium up to Right renal vein and hepatic vein. It was an 8 hour long surgery, during which acute blood pressure elevations were managed with intravenous isosorbide di nitrate infusion. Intra and peri-operative course of surgery remained uneventful, with discharge of the patient on pain killers only, on 9th post-operative day.

Six months post-operative, patient remains well and has resumed his normal day to day activities. He remains normotensive and off all medications. His post-operative urinary VMA level also normalized (4mg/hr). Repeat echocardiographic findings showed regression of changes with only mild asymmetrically hypertrophied left ventricle.

## Discussion

Extra-adrenal paragangliomas account for 10 to 15% of all adult paragangliomas with an incidence rate of 2-8 cases per million. They are roughly half as common as pheochromocytomas. Approximately 69% paragangliomas originate in head and neck, 10% in thorax and 22% in abdomen and pelvis.<sup>1</sup> Abdominal paragangliomas are mostly retroperitoneal in location, accounting for 85% of all extra adrenal paragangliomas and all are usually secretory.

Different paragangliomas have different clinical presentations based on location and ability to secrete hormones. Clinical expression of phaeochromocytoma may involve numerous cardiovascular manifestations, but usually present as sustained or paroxysmal hypertension. Reversible dilated or hypertrophic cardiomyopathy

(sometimes mimicking features of HOCM) are well established cardiac manifestations of pheochromocytomas.<sup>2</sup>

Very few cases with echocardiographic findings mimicking hypertrophic obstructive cardiomyopathy associated with paraganglioma have been reported.<sup>3,4</sup> This case was initially mislabelled as HOCM based on echocardiographic findings alone. He presented to various hospitals with acute hypertension and encephalopathy, the investigation of which eventually led to the establishment of the current diagnosis.

Treatment options in paraganglioma vary with the extent of disease. Laproscopic surgery is preferred, but large and invasive tumours require an open procedure, as in our case. Manipulation of the tumour can cause a significant haemodynamic response and rise in both systolic and diastolic blood pressures. Combined alpha and beta blockers are used for perioperative management of high blood pressures. Acute hypertensive crises are usually managed with Sodium nitroprusside (SNP), nitroglycerine, nicardipine and phentolamine.<sup>5</sup> This patient was managed with intravenous isosorbide di nitrate infusion in hypertensive crises. With all the possible measures of blood pressure control during acute crises, this patient did not experience any devastating permanent neurological and cardiovascular sequelae such as myocardial infarction and cerebrovascular accident.

Complete resolution of symptoms, normalization of blood

pressures with partial regression of asymmetrical hypertrophy of left ventricle and obstruction has been documented in other cases<sup>4</sup> as well as in this case. Patient continues to be normotensive after tumour removal, and echocardiographic findings have improved, unlike HOCM.

### Conclusion

This case report together with few others in literature indicates that HOCM like features can be induced by this tumour, with complete or partial resolution of these features following successful tumour removal. This is in contrast to classic HOCM which is a lifelong irreversible disease.

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