

Case Report on Operative Management of Pheochromocytoma

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ABST	RACT					
Pheoc	hromocytomas are a type	of catecholamine	secreting tumors	with ra	are incidend	ce. The excess
catecholamine secretion, hallmark in these types of tumor cause fluctuations in blood pressure and make						
management quite risky if not adequately managed pre-operatively.						
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Vanill	ylmandelic acid.					
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INTRODUCTION

Pheochromocytoma (PCC) is a rare neuroectodermal tumor [1], with 80-85% derived from the chromaffin cells of the adrenal medulla [2]. The World Health Organization (WHO) defines pheochromocytoma as a tumor arising from catecholamine-producing chromaffin cells the adrenal medulla-an in intra adrenalparaganglioma. Closely related tumors of extra adrenal sympathetic (catecholamine producing) and parasympathetic (rarely catecholamine producing) paraganglia are classified as extra adrenal paraganglioma [3].

These are of rare type and the annual incidence of pheochromocytoma ranges from 2 to 8 cases per one million individuals [4]. The most common manifestations of this clinical spectrum include hypertension, headache, palpitations, episodic sweating, and feeling of doom [5].

This neuroendocrine tumor is associated with a most unpredictable and fluctuating clinical course during anesthesia and surgical intervention [5]. The main-stay in the management of pheochromocytoma surgeries is preoperative preparation which has improved perioperative outcome [1]. Preoperative medical management reduces both preoperative morbidity and mortality [6].

Preoperative preparation includes the use of alphaadrenergic antagonists, beta-adrenergic antagonists with or without other antihypertensive agents, fluid therapy as well as insulin therapy for hyperglycemia if required [7].

CASE REPORT

A 44-year-old male who had previously diagnosed (one year back outside hospital) with left-sided Pheochromocytoma was presented with complaints of left-sided abdominal pain, lasting from past one year and with a history of weight loss, intermittent sweating and palpitation.

In the past, he had a history of on and off yellow colored vomit during initial presentation. The patient was subjected to laparotomy procedure at the time of initial management, but was unsuccessful due to fail in following pre-operative management protocol. Presently he was admitted for further management.

On examination, vitals were found to be normal with a mild elevation of blood pressure (130/90mmHg). No palpable mass, no guarding, but mild generalized tenderness and presence of laparoscopic scar were observed. CVA was reported one year back and no history of cardiac illness, chest pain and palpitation were determined. His height was 160 cm and weighed 60 kg with a BMI of 23.4kg/m². He quit smoking 4 years back

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and is non-alcoholic. He was a known diabetic patient and on Glycinorm-M 80 and Voglibose 0.3mg.

His MRI abdomen showed large lobulated heterogeneously enhancing mass lesion in left para-aortic region, anterio-medial to the left kidney and abutting the medial limb of the left adrenal gland. Biopsy result confirmed pheochromocytoma (figure 1 [8]). Inferiorly the lesion extended up to the level of renal hilum displacing the renal vein inferiorly and partially encasing the renal artery on left side. No significant retroperitoneal lymph adenopathy was observed. Small-sized right kidney with few small cortical cysts were reported.



Figure 1:Pheochromocytoma

CT abdomen confronted with poorly marginated hypodense lesion with heterogenous enhancement arising from body to left adrenal gland measuring $6.4 \times 5.3 \times 7.1$ cm (AP × TR × CC) with surrounding fat stranding was noted (Figure 2 [9]). The lesions showed multiple foci of internal non-enhancing foci with signs of necrosis. No evidences suggestive of calcification/macroscopic fat were noted. The lesion was supplied by prominent left adrenal, left inferior phrenic and left renal arteries. The lesion was closely abutting the supra renal aorta; origins of celiac artery and SMA for the circumference of < 180 degrees with loss of fat plane were noted, however no obvious intra luminal thrombus/luminal narrowing was seen.

The lesion was seen indenting the left crura of diaphragm. The lesion was found partly encasing the left main renal artery, however no obvious intra luminal thrombus/luminal narrowing was seen. It also is abutting the left renal vein with no thrombosis seen.

Anteriorly it abuts the proximal body of pancreas and splenic vein.

Another lesion with similar imaging characteristics were seen in retrocaval region measuring 4.1×2.5 cm which showed thin sheet of peripheral calcification in its anterior aspect. This lesion was abutting the right renal artery. The right adrenal was normal.



Figure 2: Adrenal mass The patient urine analysis was performed (Table 1)

Test	Result		
Adrenaline in urine	0.97		
24 hr urine volume	2500ml		
Adrenaline level in urine 24 hr	2.43		
Nor-adrenaline urine	266		
Nor-adrenaline urine 24 hr	478.8 (normal < 90)		
VMA urine 24 hr	49.99 (normal < 13.6)		
Chromogranin A	392 (normal < 108)		

Table 1: Lab results

Blood reports show elevated metanephrine levels and urinary VMA (Vanillylmandelic acid). Endocrinologist and cardiologist opinion was noted and on behalf patient started with alpha blocker (Prazosin 2mgTID). The patient was hemodynamically stabilized and planned for surgery after optimal stabilization. On discharge, he was prescribed with Prazosin 2mg TID, Amaryl M1 (1/500 mg a.m) and Melmet SR (500 mg HS) and advised for salt restricted diet and diabetic diet.

After 10 days of medication, patient admitted again and planned for laparotomy + retroperitoneal tumor excision. Preoperative procedure was done as follows:

- Midnight starvation
- IVF NS100cc/hr
- HIV, HbsAg, blood grouping done
- Inj. Fragmin 2500Us/c
- Incentive spirometry
- Inj. Kephazon-S 2g IV

Patient after pre-operative procedure observed with episodes of pain and sweating. Urine nor-adrenaline was found to be greater than 3 times UL, BP (160/100 mmHg). Echo shows concentric LV hypertrophy, and if the patient BP is uncontrollable, suggestive of "pheo crisis", and should be managed with alpha blockers (Prazosin/ phenoxy benzamine), pre-operative with or without beta blockers. Surgery was postponed to 2-3 weeks later due to uncontrollable BP and withheld INJ. Fragmin. Patient BP was 120/90 and heart rate was

102bpm on the next day. Patient found to be comfortable and discharged with Prazosin the other day.

After ten days of alpha blocker therapy, patient was admitted again to hospital for surgical procedure. BP was monitored and measured 120/90 mmHg. Since BP was variable, he was advised with T.Aplar 10 mg TID and Propranalol 10 mg TDS and IVF NS 75cc/hr. On the next day, he showed rise in BP (220/110 mmHg) and added Alpha blocker therapy and IVF NS 75cc/hr. BP measured after an hour was variable 150/90 mmHg, and he was advised for Alprazolam 20mg TDS and Alpha blocker SOS. Patient BP was 200/110 mmHg on the consecutive day and Alprazolam was tapered to 10mg TID and alpha blocker 2mg QID, IVF NS 75 ml cc/hr.

Since BP was found stable, surgery was planned for a day after and pre-operative procedures performed as mentioned earlier.

On the day of surgery, epidural infusion (Ropivacaine 0.2% + Fentanyl) was administered and procedure done. Intra operatively, left adrenal mass (6 x 5 cm) was found adherent to left kidney, aorta, left haemidiaphragm, spleen, and inferior mesenteric vessels, which was dissected of these structures. The left-sided tumor found encroaching the renal vessels, from which it was dissected and tumor was excised. Right-sided extra adrenal para ganglion mass was noted, inferior to right renal hilus, posterior to IVC, adherent to right renal vein and IVC which was having dense adhesions assumed to be due to previous surgery. The adhesions were taken down and retro-caval dissection was done to remove the tumor in total. The right adrenal was separated from tumor and kept undisturbed. Intra-operatively few episodes of hypertension while dissecting the tumor were observed, which were managed appropriately.

Post operatively, patient was under monitoring of vitals for one day, managed with IV fluids, IV analgesics, antibiotics and PPI. DVT prophylaxis was done. He had transient paresis and numbness of right lower limb from ankle joint to groin in the immediate post-operative period, probably due to epidural infusion which subsided over the next few hours. Endocrinology and cardiology opinion was taken. Patient made an uneventful recovery.

Patient was eating and drinking normally, mobilizing well, wound was clean and BP was within normal limits. At discharge, the patient was voided well, advised to follow a proper diabetic diet, informed to contact hospital if any signs of sweating, palpitation and vomiting and discharged with T. Prazosin 2 mg TID, T. Amaryl 1/500 mg a.m and T. Melmet SR 500 mg HS.

DISCUSSION

Necessity of proper preoperative adrenoceptor blockade and other preventive procedures in pheochromocytoma patients are clearly describing in this case. Our patient was experiencing intermittent sweating and palpitation suggestive of catecholamines excess, together with difficulty to treat hypertension should arouse immediate doubt of pheochromocytoma [3]. The biochemical confirmation of the presence of pheochromocytoma should be based on measurements of metanephrines in plasma or urine. Blood report of this patient shows high increase in metanephrine and VMA levels, the former is precise as it is based on the constant adrenal activity of COMT, which converts catecholamines to metanephrines and the later exhibits high specificity due to the conversion of catecholamines to VMA, occurs through both the COMT and the MAO pathways [4].

After pre-operative procedure, patient had experienced elevated nor adrenaline and symptoms of pheochromocytoma and started on Alpha blocker Prazosin. These medications negate the effects of the excess hormones secreted by the pheochromocytoma, reducing the frequency and severity of dangerous blood pressure fluctuations intraoperatively and postoperatively [10]. Prazosin was preferred over Phenoxy benzamine in this case since they have conservation of alpha-2 effect and lower incidence of post-operative hypertension [5]. Added therapy with beta blockers is preferred in this case to counteract the tachycardia induced by nonselective alpha-blockade or due to vasodilatation induced increase in heart rate [6].

Excess catecholamine associates with decrease in intra vascular volume and blood pressure. As suggested by evidences [5], patient had normalized and expanded intravascular volume by infusing normal saline prior to surgical procedures to reduce post-operative hypotension. Reducing anxiety prior to induction is essential to prevent trepidation from causing catecholamine surges. Alprazolam has given prior to induction to reduce the hypertensive crisis likely to obtain [5].

CONCLUSION

The management of patients with pheochromocytoma begins from ample preoperative preparation, intraoperative monitoring and watchful follow-up during the postoperative period. Prognosisappears to be allied to tumor size, extent of uncontrolled HTN, and the existence of metastatic disease; it is good if the tumor is detected early to evade major complications linked to catecholamine excess.

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