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Case Report

Case report of laparoscopic adrenalectomy for a giant pheochromocytoma: medical and surgical challenge

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ABSTRACT

Pheochromocytomas are rare adrenal tumors that can pose significant clinical challenges due to their potential for catecholamine release and hypertensive crises. We present a case of a large pheochromocytoma managed successfully through a laparoscopic adrenalectomy. A 66-year-old male presented with adrenal incidentaloma. Imaging revealed a sizable left adrenal mass (9.5 cm) consistent with a pheochromocytoma. Preoperative alpha- and beta-adrenergic blockade was initiated to optimize hemodynamic control. The patient underwent a laparoscopic adrenalectomy, with meticulous dissection and adrenal vein ligation to minimize catecholamine release during surgery. Post operatively, blood pressure and catecholamine levels normalized. Histopathological evaluation confirmed the diagnosis of pheochromocytoma with high pheochromocytoma of the adrenal gland scaled score (PASS) and grading of adrenal pheochromocytoma and paraganglioma (GAPP) scoring. Laparoscopic adrenalectomy proved to be a safe and effective approach for managing this large pheochromocytoma, resulting in improved blood pressure control and quality of life for the patient. This case underscores the importance of a multidisciplinary approach, including preoperative medical optimization and close postoperative monitoring, in the management of pheochromocytomas, and contributes to the growing body of evidence supporting laparoscopic techniques for these adrenal tumors.

Keywords: Giant pheochromocytoma, Catechiolamines, Laparoscopic adrenalectomy

INTRODUCTION

Pheochromocytomas are rare catecholamine-producing tumors arising from chromaffin cells in the adrenal medulla. While the majority of pheochromocytomas are relatively small in size, ranging from a few millimeters to a few centimeters, a distinct subset of cases presents with an atypical feature: giant pheochromocytomas.[1]

These tumors, characterized by an unusually large size, pose unique diagnostic and therapeutic challenges due to their potential for causing extensive local invasion, compression of adjacent structures, and a heightened risk of catecholamine-related complications.[2] Here we report a case of giant pheochromocytoma in a 56-year-old gentleman which was found during an evaluation for lower urinary tract symptoms. He was started on alpha blocker and referred to us. After adequate blockade for pheochromocytoma, he underwent laparoscopic left adrenalectomy (95×60×40 mm) and peri operative period was uneventful.

Despite advancements in imaging techniques and surgical approaches, the surgical removal of giant pheochromocytomas remains a formidable task, often requiring intricate planning and multidisciplinary collaboration. The potential for intra-operative hemodynamic instability and the risk of peri operative catecholamine surges further underscores the need for tailored peri operative management protocols.

CASE REPORT

We describe a case of 56-year-old gentleman who was referred to our department with a left adrenal mass. He had diabetes mellitus and hypertension for the last few years for which he was on insulin and multiple anti hypertensives. He had occasional sweating, palpitation and headache which was connected to hypoglycemia which was not documented. As he had lower urinary symptoms, he underwent ultrasound of abdomen and pelvis which showed a left adrenal mas. He had no other symptoms or symptoms due to complications of pheochromocytoma like myocardial infarction, congestive heart failure or cerebrovascular accident.

Table 1: Biochemical investigations.

S. Cortisol 8 AM	17.40 mcg/dl	(5-23)
Aldosterone renin ratio	3.5	(<30)
24-hour urine VMA	66.83 mg/24 hrs	(<13.6)
ECHO	LVEF 67% No RWMA, normal LVSF, LVDD	
S. creatinine	1.09 mg/dl	
Sodium	136 meq/dl	
Potassium	4.6 meq/dl	

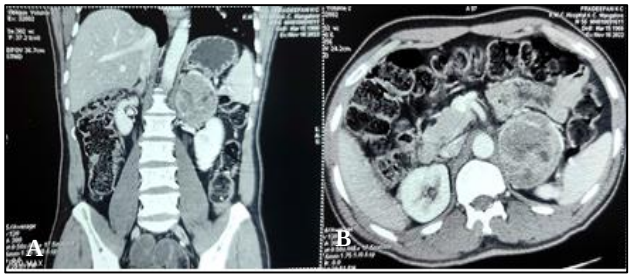

Figure 1 (A and B): Contrast enhance computerized tomography showed large well defined left adrenal mass.

He was further evaluated and found to have elevated 24-hour urine vanillyl mandelic acid (VMA): 66.83 mg/24 hours (Normal:<13.6) and started on non-selective alpha blocker and then referred to us for further management. His further hormonal evaluation showed normal reports including 8 AM cortisol-17.40 mcg/dl (Normal:5-23 mcg/dl) (Table 1). Contrast enhanced computerized tomography (CECT) of abdomen was done in adrenal protocol which showed well defined mass 65×60×74 mm, left adrenal gland (Figure 1). Right adrenal gland separately seen and is normal. Heterogeneous predominantly peripheral enhancement 162 HU which is retained in delayed phase and with few central necrotic non-enhancing areas. He was changed to selective alpha blocker from our department. It took 3 weeks to get adequate control with prazosin of 17.5 mg and metoprolol 100 mg in divided doses.

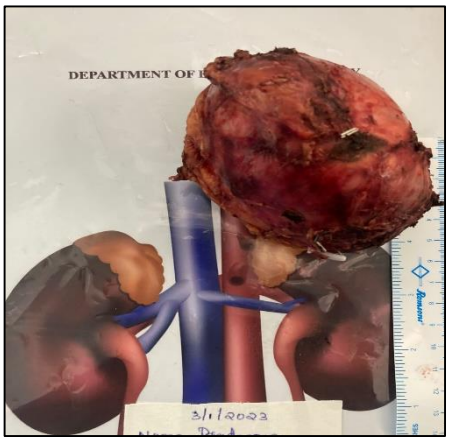

Figure 2: Specimen picture of left adrenalectomy.

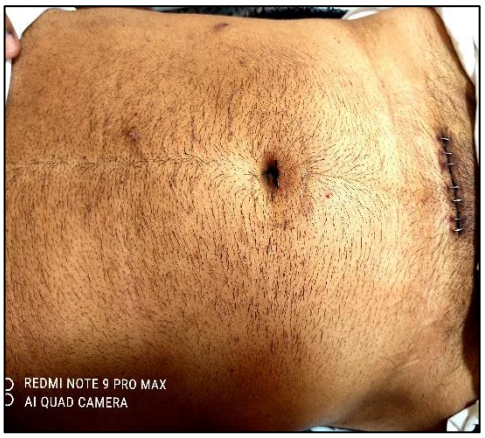

Figure 3: Post operative picture showing laparoscopic ports and pfannensteil incision.

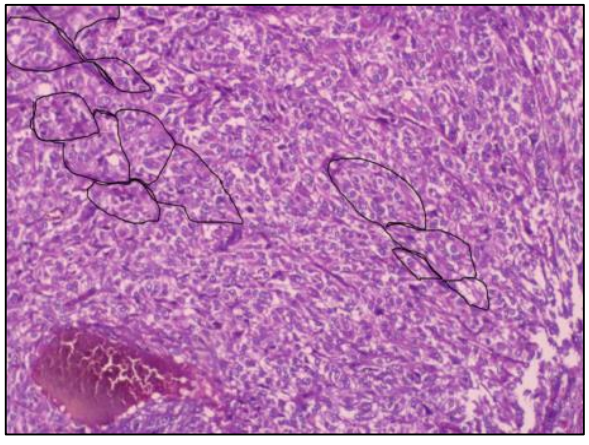

Figure 4: Zell Ballen appearance of pheochromocytoma.

He underwent laparoscopic trans peritoneal left adrenalectomy. Left adrenal tumor was large (95×60×40 mm) (weight:180 gm) and highly vascular in nature.

Multiple venous and arterial connections to the retro peritoneum was present. After clipping the adrenal vessels, Peritumoral dissection was done and specimen (Figure 2) removed through a separate Pfannenstiel incision in the supra pubic region (Figure 3). Highest blood pressure recorded in the peri procedure was 15/90 mmHg. Post operatively he had hypotension which was managed by ionotropes for 24 hours.

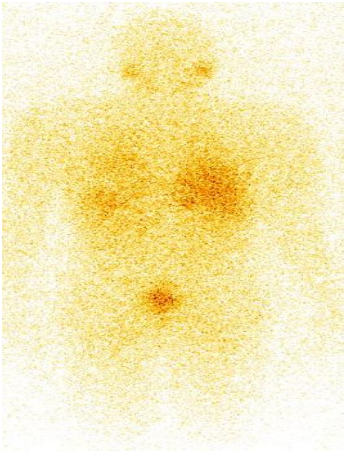

Figure 5: I 131MIBG showing no evidence of metastasis.

His sugars were closely monitored, looking for hypoglycemia. He was not on insulin or any hypertensives in the post operative period and discharged on post operative day 4. Histopathology reported as pheochromocytoma (Figure 4) with PASS score of 6 which suggests that this is an aggressive tumor. He was further investigated with I-metaiodobenzylguanidine (123 I-MIBG) scan (Figure 5) to rule out any metastasis which was normal. He was on close follow up for last one year and his plasma normetanephrines and metanephrines were within normal limits.

DISCUSSION

Pheochromocytomas are tumors arising from the chromaffin cells of adrenal medulla and these are neuroendocrine tumors which produces catecholamines.[3] Giant pheochromocytoma are tumors larger than 7 cm in size.[4] These giant tumors are rare in literature and characterized by an unusually large size, pose unique diagnostic and therapeutic challenges due to their potential for causing extensive local invasion, compression of adjacent structures, and a heightened risk of catecholamine-related complications. Majority of these tumors are lacking classical symptoms like headache, palpitation and diaphoresis and not producing catecholamines unlike our case.[5]

Tumour necrosis, abundant interstitial tissue compared to chromaffin cells and encapsulation of tumour by connective tissue are the causes of paucity of catecholamine production. Biochemical evidence of high plasma or urine metanephrines and normetanephrines have high sensitivity and specificity as compared to urine VMA.[6] As our patient had elevated urine VMA and was already started on alpha blockade from outside, normetanephrines or metanephrines are not done pre operatively as it can interfere with fractionated metanephrines value. CT scan is the first choice in imaging for adrenal lesions as compared to MRI as it gives better spacial resolution. On CT scan of abdomen, pheochromocytomas are heterogeneous in appearance with multiple cystic areas, hemorrhage, calcification, and necrotic areas within it. On arterial phase pheochromocytomas are enhanced due to its hyper vascular nature. The subsequent venous phase often shows washout, enhancing the lesion's detectability.[7] Pre operative management of pheochromocytoma is crucial to attain normal blood pressure, heart rate and restore effective circulating blood volume to avoid hemodynamic instability during peri operative period.[8]

In our case we used selective alpha blocker prazosin, which was gradually increased up to 17.5 mg/day over 3 weeks of time and added cardio selective betablocker (metoprolol XL) up to 100 mg/day to attain normal blood pressure and to avoid reflex tachycardia and post operative hypotension. Tumor size, blockade used and open adrenalectomy are associated with hemodynamic instability during peri operative period which was insignificant in our case due to adequate pre operative blockade.[9]

The definitive management of adrenal pheochromocytoma is surgical excision. Laparoscopic adrenalectomy is the best and safe procedure for large pheochromocytomas.[10] Laparoscopic adrenalectomy shows lower estimated blood loss, lower transfusion rate, lower hemodynamic instability, less postoperative complications, less Clavien-Dindo score 3 complications, shorter return to diet time, and shorter length of hospital stay. Most of the pheochromocytomas are benign, incidence of malignancy is less than 10%. Presence of chromaffin tissue in the extra adrenal tissue is the only pathognomonic feature of malignancy in pheochromocytoma. Predicting the malignant potential of a pheochromocytoma can be challenging, and various factors are considered by pathologists and medical professionals to assess the risk of malignancy.

Two commonly used scoring systems for assessing the malignant potential of pheochromocytomas are the PASS (pheochromocytoma of the adrenal gland scaled score) and the GAPP (gross appearance, age, pheochromocytoma of the adrenal gland scaled score, and proliferation index) score. PASS scoring system is a histological scoring system used to evaluate the appearance of pheochromocytoma under the microscope. It assigns scores based on various histological features such as cellularity, cell spindling, presence of necrosis, and mitotic figures. A higher PASS score suggests a higher likelihood of malignancy.[11] GAPP (gross appearance, age, pheochromocytoma of the adrenal gland scaled score, and proliferation index) score takes into

account not only histological features but also clinical and radiological findings. It considers factors such as the patient's age, the appearance of the tumor during surgery (gross appearance), the PASS score, and the proliferation index (Ki-67 labeling index). A higher GAPP score indicates a greater likelihood of malignancy.[12] It's important to note that no single scoring system is perfect, and the assessment of pheochromocytoma malignancy often involves a combination of clinical, radiological, and pathological factors. Additionally, molecular and genetic testing may also be used to help determine the tumor's malignant potential.[13]

The definitive diagnosis of malignant pheochromocytoma often requires histological examination and evaluation by an experienced pathologist. The treatment and management of pheochromocytoma can vary depending on its malignant potential, and decisions regarding surgery and follow-up care are made based on a comprehensive assessment of the individual case. In our case PASS score was high which was further evaluated with I 131 MIBG scan which was negative for metastasis. Post operatively patient experienced a favorable course with normalization of blood pressure and catecholamine levels. This case exemplifies the successful resolution of a challenging medical condition, ultimately improving the patient's quality of life including high blood pressure and diabetes mellitus

CONCLUSION

This case report not only underscores the effectiveness of laparoscopic adrenalectomy as a minimally invasive surgical option for large pheochromocytomas but also emphasizes the importance of a multidisciplinary approach involving endocrine surgeons, anesthesiologists and pathologists. Furthermore, it contributes to the growing body of evidence supporting the feasibility and safety of laparoscopic techniques in the management of adrenal tumors. While, our case demonstrates a positive outcome, the management of pheochromocytoma should always be tailored to the individual patient's characteristics and clinical presentation. Further research and collaborative efforts in this field can help refine treatment protocols and enhance patient care. This case report serves as a valuable addition to the existing medical literature, contributing to our understanding of the optimal management strategies for large pheochromocytomas.

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REFERENCES

1. Bogati K, Baniya S, Thapa S, Regmi UK, Karki N, Pokhrel M. A giant pheochromocytoma presenting in pheochromocytoma crisis: a case report. J Nepal Med Assoc. 2023;61(258):179.
2. Villarraga N, Palani G, Risk M, Sibley S. Unique presentation and perioperative management of a giant pheochromocytoma. JCEM Case Reports. 2023;1(3):65.
3. Antonio K, Valdez MMN, Mercado-Asis L, Taïeb D, Pacak K. Pheochromocytoma/paraganglioma: recent updates in genetics, biochemistry, immunohistochemistry, metabolomics, imaging and therapeutic options. Gland Surg. 2020;9(1):105-23.
4. Ambati D, Jana K, Domes T. Largest pheochromocytoma reported in Canada: A case study and literature review. Can Urol Assoc J. 2014;8(5-6):374-7.
5. Li C, Chen Y, Wang W, Teng L. A case of clinically silent giant right pheochromocytoma and review of literature. Can Urol Assoc J. 2012;6(6):267-9.
6. Garg MK, Kharb S, Brar KS, Gundgurthi A, Mittal R. Medical management of pheochromocytoma: Role of the endocrinologist. Indian J Endocrinol Metab. 2011;(4):329-36.
7. Čtvrtlík F, Koranda P, Schovánek J, Škarda J, Hartmann I, Tüdös Z. Current diagnostic imaging of pheochromocytomas and implications for therapeutic strategy. Exp Ther Med. 2018;15(4):3151-60.
8. Fang F, Ding L, He Q, Liu M. Preoperative management of pheochromocytoma and paraganglioma. Front Endocrinol (Lausanne). 2020;11:586795.
9. Kiernan CM, Du L, Chen X, Broome JT, Shi C, Peters MF, et al. Predictors of hemodynamic instability during surgery for pheochromocytoma. Ann Surg Oncol. 2014;21(12):3865-71.
10. Conzo G, Musella M, Corcione F, De Palma M, Ferraro F, Palazzo A, et al. Laparoscopic adrenalectomy, a safe procedure for pheochromocytoma. A retrospective review of clinical series. Int J Surg. 2013;11(2):152-6.
11. Thompson LD. Pheochromocytoma of the Adrenal gland Scaled Score (PASS) to separate benign from malignant neoplasms: a clinicopathologic and immunophenotypic study of 100 cases. Am J Surg Pathol. 2002;26(5):551-66.
12. Koh JM, Ahn SH, Kim H, Kim BJ, Sung TY, Kim YH, et al. Validation of pathological grading systems for predicting metastatic potential in pheochromocytoma and paraganglioma. PLoS One. 2017;12(11):187398.
13. Yamazaki Y, Gao X, Pecori A, Nakamura Y, Tezuka Y, Omata K, et al. Recent advances in histopathological and molecular diagnosis in pheochromocytoma and paraganglioma: challenges for predicting metastasis in individual patients. Front Endocrinol (Lausanne). 2020;11:587769.

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