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Case Report

A rare case of symptomatic bilateral pheochromocytoma

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ABSTRACT

Pheochromocytoma with reported incidence of 2-8 per million, is a rare and special tumor with its own unique clinical and pathological features, originating from the amazingly intelligent neuroendocrine cells of chromaffin cells of the adrenal medulla. It may behave as 'great masquerader' and be sweet and predictable as a child, while at times act as a 'treacherous murderer' crashing and tearing everything in its path with fierce rage, the main cornerstone of the disease being surgical excision. Here, we report a rare case of middle aged female presenting with hypertensive crisis which necessitates intensive care unit (ICU) management where she responded well to anti-hypertensives and radiological investigations were suggestive of bilateral pheochromocytoma. The course of treatment consisted of array of investigations to ascertain the diagnosis before embarking on bilateral adrenalectomy keeping in mind the consequences of absence of adrenal and subsequent possibility of Nelsons syndrome, which we were able to treat successfully. Here we want to highlight the consideration of bilateral adrenal tumors as a differential diagnosis for suprarenal growths, subsequent surgical course and post-operative medical treatment highlighting the importance of mineralocorticoids and glucocorticoids for day to day functioning.

Keywords: Adrenal pheochromocytoma, Bilateral pheochromocytoma, Hypertension and pheochromocytoma, Nelsons syndrome

INTRODUCTION

The globally increased blood pressure is the silent killer affecting all age groups hampering day today lives of people. Pheochromocytomas are a rare cause of secondary hypertension.They form 0.2-0.6% of such cases.[1]

These tumors are known to clinically manifest by classical combination of headache, palpitations and sweating. Other symptoms like abdominal pain, nausea, vomiting, shortness of breath and chest pain are known to occur. There might be associated episodes of sudden cardiac arrest due to myocardial infraction or can have an episode of cerebrovascular accident which results in sudden deaths at times, these events can be precipitated by an range of stimuli including defecation, exercise or micturition. These all clinical myriads of symptoms are attributed to increased release of catecholamines otherwise termed as pheochromocytoma crisis. While the crisis may occur as part of the natural disease process, it can also be precipitated suddenly by general anesthesia, direct trauma to the tumor, or with the administration of metoclopramide.[2]

Pheochromocytoma originatefrom the chromaffin cells of the adrenal medulla and secrete excess catecholamines (norepinephrine, epinephrine and dopamine). They are essentially benign tumors, with only 2-15% being malignant. These forms part of MEN 1 syndrome, MEN2 syndrome, MEN2A and MEN2B, Neurofibromatosis type 1 syndrome.

The malignant type of pheochromocytoma is only diagnosed in the presence of extra-chromaffin spread or

evidence of invasion in to surrounding structures or distant metastases. The prognosis of the tumors is unpredictable at the initial presentation.[3]

Surgical excision in the form of adrenalectomy is the main line of treatment whenever possible even in cases with advanced disease surgical debulking helps to prolong the survival. Chemotherapy helps in controlling symptoms and survival in patients with metastatic disease.[4]

CASE REPORT

We report a 25-year-old female, who presented with pain in abdomen, headache, palpitations and a blood pressure of 210/120 mmHg on admission. She also had history of similar complaints since 3 months and multiple hospital visits. Physical examination elicited lump palpable in left lumbar of size 6×5 cm, firm, ballotable, globular, tender and not moving with respiration. An abdominal ultrasound revealed lobulated heterogenous lesions in B/L suprarenal region causing mass effect in form of displacement of kidneys.

Contrast enhanced computed tomography (CECT) abdomen and pelvis revealed large lobulated heterogenous solid cystic lesion in B/L adrenal gland with findings suggestive of pheochromocytoma.

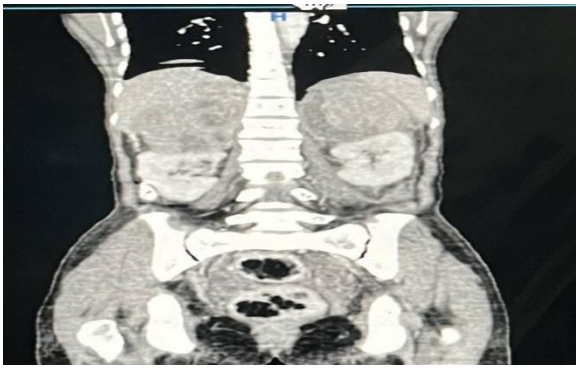

Figure 1: Coronal section of contrast enhanced abdomen pelvis showing bilaterally enlarged suprarenal glands.

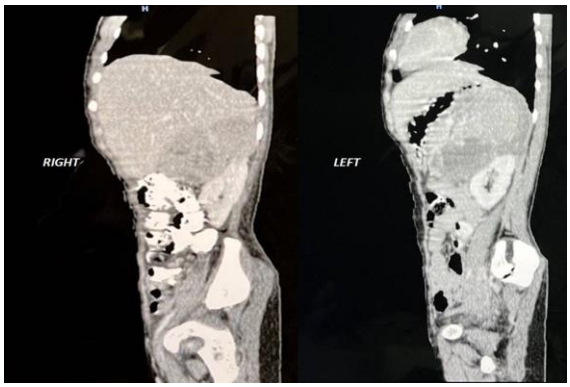

Figure 2: Sagittal section films confirming enlarged right and left suprarenal glands.

After admission patient underwent series of investigations such as VMA, cortisol, meta-nephrin, TFT to rule out any thyroid, parathyroid and cardiac disorders. Patient was initially managed medically for anemia, abdominal pain and continuously raised blood pressure with nifedipine and phenoxybenzamine and subsequently posted for surgery. Intra-op findings were consistent with right lump in close relationship with inferior liver surface, renal pelvis, inferior vena cava.

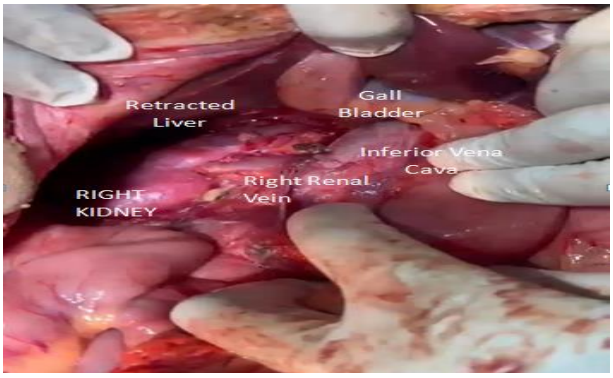

Figure 3: Intra-op surgical field right side.

Left lump in close relation with pancreas with maintained margins.

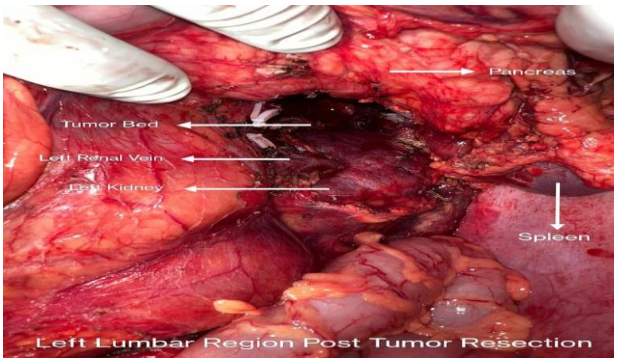

Figure 4: Coronal section of contrast enhanced abdomen pelvis showing bilaterally enlarged suprarenal glands.

Patient underwent bilateral adrenalectomy.

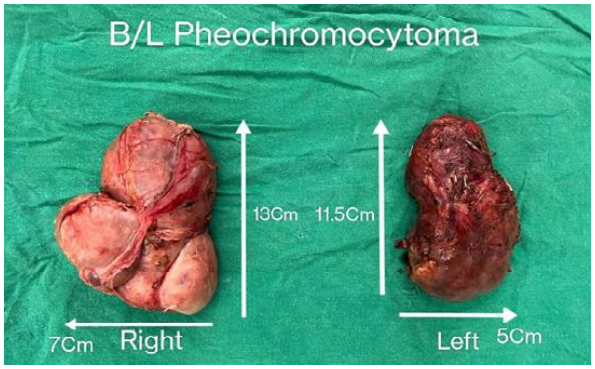

Figure 5: Post-operative excised specimen.

A pathological examination showed right and left suprarenal mass with histological findings consistent of pheochromocytoma and negative for adrenocortical adenoma, sarcoma, capsular invasion. Histopathological evaluation-Nested (Zellballen), trabecular or solid arrangement showing nests outlined with sustentacular cells.

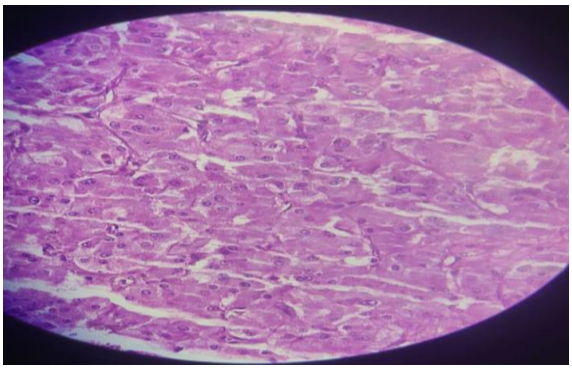

Figure 6: Histopathology showing nsted (Zellballen), trabecular or solid arrangement showing Nests outlined with sustentacular cells.

Post operatively on day 5 patient started developing sweating, loose stools, and fall in blood pressure and was initially managed conservatively on IV fluids and antibiotics. Patient was placed in continuous BP and BSL charting which revealed fall in blood pressure and hypoglycemia. Patient was then started on fludrocortisone 0.1 mg and prednisolone 5 mg tablets. However, due to reoccurrence of symptoms after few days, doses of the drugs were increased and continuous maintenance therapy was given to which the patient responded well.

Patient was discharged on continuous oral intake of fludrocortisone and glucocorticoids.

DISCUSSION

The paroxysmal symptoms caused due to pheochromocytoma which usually present as an acute medical emergency in the casualty are attributable to the catecholamines released by the tumours- dopamine, epinephrine and norepinephrine which are the major cause of cerebrovascular and cardiac complications.[5] Not that it always presents with the classical triad of symptoms, the clinical presentation is most commonly in the form of an adrenal incidentaloma, with only 4% of adrenal masses found known to be pheochromocytomas, most common being benign adenomas.[6] Thus, bilateral pheochromocytomas are a very rare phenomenon.

In the present case, combining mass and the presenting complaints, the diagnosis of pheochromocytoma is more likely. For fear of providing incomplete treatment and for risk of remittance, surgery is the treatment of choice for these tumours. It has been proven curable for more than 90 percent of the patients. As much as the surgical part is crucial, the preoperative preparation is of utmost importance in reducing the morbidity and mortality.[7]

To minimize hemodynamic instability during tumor manipulation, alpha blockade is recommended for pheochromocytoma resection.[8] The most common elective drug being used is phenoxybenzamine. In 20-70% patients hypotension an develop as a complication postoperatively.

Surgery causes depletion of glycogen stores, and after tumour removal there may occur sudden catecholamine withdrawal, which in turn leads to rebound hyperinsulinemia and severe hypoglycemia postoperatively suggestive of Nelsons syndrome which can be tackled by monitoring blood pressure and Blood sugar levels.[8]

CONCLUSION

Patients with undiagnosed pheochromocytoma may present in extremis to an emergency department with pheochromocytoma crisis and coexisting metabolic, thromboembolic, or surgically emergent events. These patients ultimately require coordination with complex critical care and optimization for definitive surgical management, which begins in the emergency department.

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