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CASE STUDY

PHEOCHROMOCYTOMA: AN UNUSUAL PRESNTATION

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ABSTRACT

Pheochromocytoma is a rare, catecholamine secreting neuroendocrine tumour of adrenal medulla representing nearly 5% of adrenal incidentalomas. We present a case of an 80 year old male complaining of recurrent urinary tract infection diagnosed as pheochromocytoma on further investigation.

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INTRODUCTION

Pheochromocytoma is a rare, potentially fatal tumours presenting with a spectrum of non-specific symptomatology. The tumour is of a very special kind owing to its nature of duplicity. Diagnosis of pheochromocytoma is frequently overlooked in the elderly due to paucity of classical symptoms of sympathetic overactivity, and features of catecholamine excess being confounded by the effects of ageing, comorbidities, and medications, hence, it may pose a challenge to diagnose them.

Case Presentation

An 80 year old man came with complains of recurrent urinary tract infections(UTI) since 6 months with on and off urinary incontinence and left lumbar pain since 6 months associated with swelling. Patient was a known case of hypertension (HTN) and type 2 diabetes mellitus (DM) was newly diagnosed. There were no complaints of increased blood pressure or labile hypertension but concomitant urinary tract infection was present.

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Patient was sexually inactive. No other significant findings were found. On physical examination his blood pressure was 110/70 mmHg, sugars were under control, 24 hour urinary metanepherine levels was grossly elevated along with a high serum metanephrine to creatinine ratio suggesting elevated levels of metabolities of catecholamines. VMA levels were also high. Urine cytology suggestive of no malignant cells and pus culture showed no growth. On histopatholgy an encapsulated cystic tumour with residual compressed tumour in the wall was present. Cells were arranged in nests, containing an amphophilic cytoplasm and malignant changes. PET scan showed low grade FDG uptake and noted a solid cystic left adrenal mass measuring 9 x 8.2 x 8.7cms. After preparing with prazosin and metoprolol pre-operatively for 5 days with adequate oral and intravenous hydration and phentolamine drip during surgery, a left sided adrenelectomy was performed. The post operative period was identified in marked reduction of symptoms of diabetes mellitus and hypertension. And further no complains of urinary tract burning along with decrease in aggressive behaviour pattern.

DISCUSSION

Pheochromocytoma, neuroendocrine tumour of adrenal medulla has an incidence of 1 per 2 million population and

0.1% in people suffering from hypertension. Classically it is closely associated to three syndromes - von Hippel Lindau (VHL) syndrome, multiple endocrine neoplasia type 2(MEN 2) and neurofibromatosis type 1 (NF1) – now nearly 10 genes have been identified related to its mutation. 40% presents with a triad of diaphoresis, paraxosymal headache and tachycardia (Reisch, 2006) and few may develop complications due to hypertensive crisis resulting in fatal outcomes. Hence, a multidisciplinary approach including a team of surgeons, anaesthetist and endocrinologist is necessary to improve patient outcome (https://www.researchgate.net/publication/49747599_Pheochromocytoma_presenting_as_recurrent_urinary_tract_infections_A_case_report), as the tumour could be life threatening due to the catecholamine storm after the resection and the malignant potential of large tumours. The term paraganglioma and pheochromocytoma have a fine line of distinction, proposing that tumour with adrenergic phenotype has a relatively low rate of malignancy and a predilection of paraganglioma is associated to succinate dehydrogenase mitochondrial enzyme mutation (Tischler, 2006 and Henri, 2009).

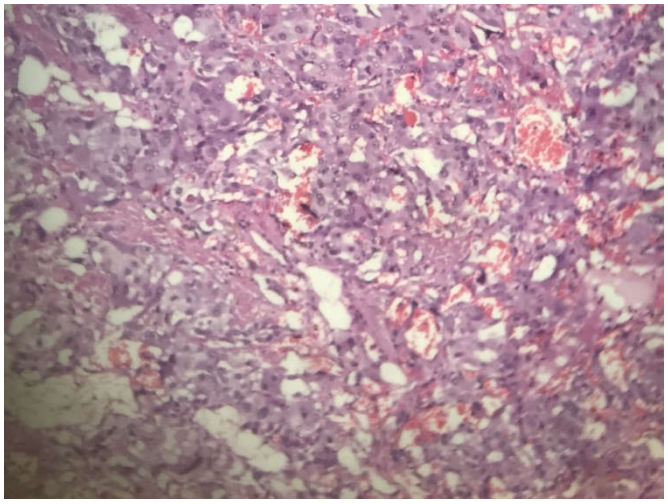

Figure 1. Histopatholgy slide of specimen with H and E staining

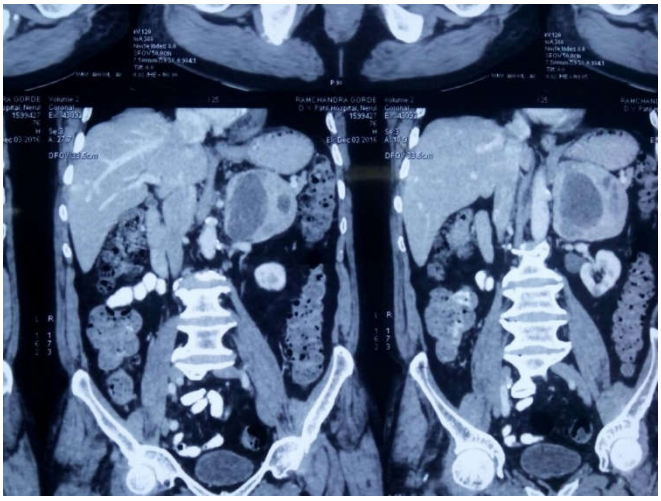

Figure 2. PET scan image 1

The mainstay of treatment is surgical removal of the tumour with expertise in the procedure of laproscopic adrenelectomy being the gold standard treatment of choice (Bjorn Edwin, 2001 and Kuriansky, 1999). However in our case we began with a laproscopic approach but due to large tumour size, we had to do an open left sided adrenelectomy. Pre-operative preparation with alpha blockers helped in minimizing the hypertension complications.

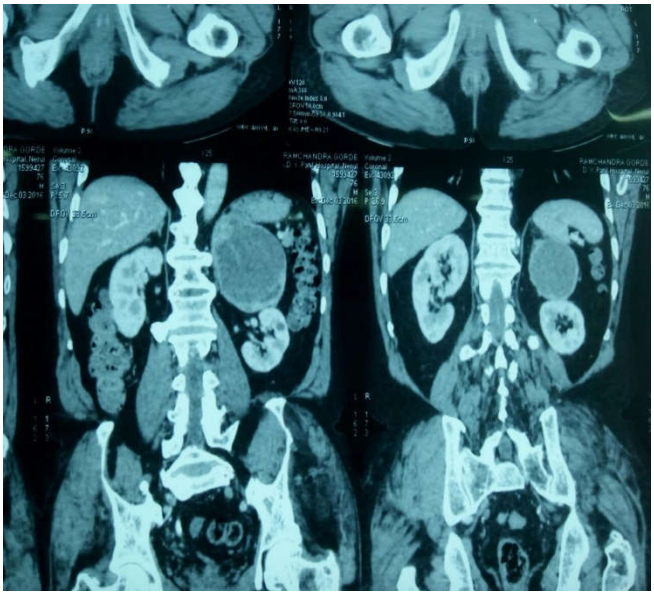

Figure 3. PET scan image 2

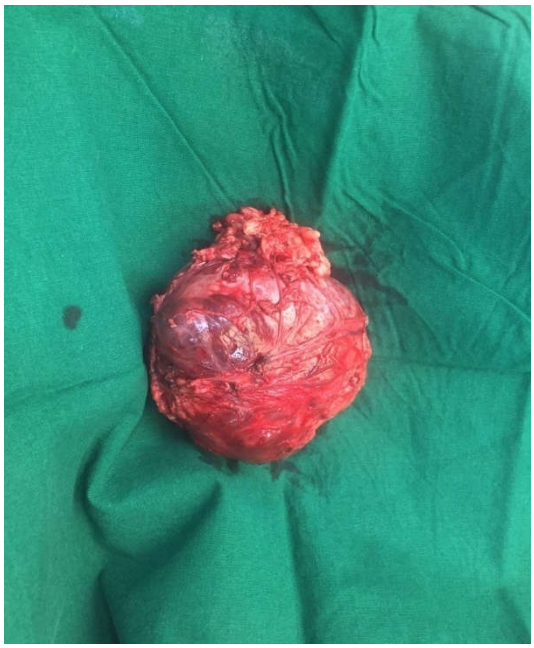

Figure 4. Gross specimen of tumour after resection

A suspicious eye should always be open when dealing with various endocrinal pathology as these tumours are an incidental diagnosis (Katharyn, 2008; http://www.merckmanuals.com/professional/endocrine-and-metabolic-disorders/adrenal-disorders/pheochromocytoma). and its familial association to each other is known. Our patient came with peculiar complaints of urinary incontinence and UTI and a series of investigation and prompt history led to our diagnosis. Excessive adrenergic stimulation due to release of catecholamines resulted in elevated blood pressure and hyperglycaemia which normalized after tumour resection. Therefore, the key to best patient outcome is early diagnosis and prompt screening of various genetic mutations in patients (Karel Pacak and Sunil J. Wimalawansa, 2015).

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