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Diagnostic and surgical challenges of a giant pheochromocytoma in a resource limited setting—A case report

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ABSTRACT

INTRODUCTION: Pheochromocytomas are catecholamine producing tumours which arise from chromaffin cells within the adrenal medulla. Patients with these tumours commonly present with a triad of headache, palpitations and hypertension.
CASE PRESENTATION: We present a case of a 37-year-old male patient who presented with dull left sided abdominal pain and discomfort for 6 weeks. A preoperative Computed tomography (CT) scan showed a huge left suprarenal tumour but urinary vanillylmandelic acid (VMA) were negative. The patient underwent an open surgical resection via an extraperitoneal approach without untoward intraoperative and postoperative events. Histopathological evaluation of the specimen showed a pheochromocytoma with a PASS score of 9. The successful management of the patient highlights the good results of team work despite the limitations of preoperative diagnosis.
DISCUSSION: Giant pheochromocytomas by definition are tumours more than 7 cm in size and are rare. They rarely secrete catecholamines and commonly present with vague abdominal symptoms. A computerized tomogram helps suggest the diagnosis whilst the biochemical workup for pheochromocytoma may be diagnostic. If the tumours are biochemically active, preoperative alpha-blockade is necessary and care must be taken at operation in handling the tumour. The surgical and anaesthetic team must be prepared to manage hypertensive crisis should it occur.
CONCLUSION: This case brings to the attention of clinicians the need to have a high index of suspicion of a giant pheochromocytoma in a patient presenting with vague abdominal symptoms whose CT scan shows a large retroperitoneal tumour, even in the absence of clinical symptoms and negative or absent biochemical workup.

1. Introduction

This work is reported in line with the Surgical Case Report Guidelines (SCARE) criteria [1]. Pheochromocytomas are catecholamine producing tumours arising from chromaffin cells within the adrenal medulla with a reported incidence of 0.1% in the general population [2,3]. The majority of pheochromocytomas are benign. Similar tumours arising from the sympathetic and parasympathetic chain are called paragangliomas and the majority of these tumours are non-secreting [4]. Pheochromocytoma commonly present with a triad of headache, palpitations and hypertension [5] due to the production of adrenaline, noradrenaline, dopamine and their metabolites. Pheochromocytoma can arise sporadically or as part of hereditary syndromes. Sporadic cases usually occur in the fourth and fifth decade of life and has equal gender distribution [6]. Hereditary pheochromocytoma occur in association with von Hippel-Lindau (VHL) syndrome, multiple endocrine neoplasia type 2 (MEN-2A/2B), neurofibromatosis type 1 (NF1), and hereditary pheochromocytoma-paraganglioma (due to mitochondrial succinate dehydrogenase gene mutations) [2,7,8].

Giant pheochromocytoma are by definition tumours more than 7 cm and are rare [9–11]. The majority of giant pheochromocytomas do not secrete catecholamines and hence do not present with classical symptoms of pheochromocytoma [11]. They present with vague abdominal symptoms. Computed tomography (CT) scan is the gold standard in the diagnosis of giant pheochromocytoma [10]. However there maybe diagnostic uncertainties in the determination of the organ of origin of these tumours. On CT scan they appear as large retroperitoneal tumours [9]. Biochemical evidence of elevated plasma free metanephrines and normetanephrines pro-

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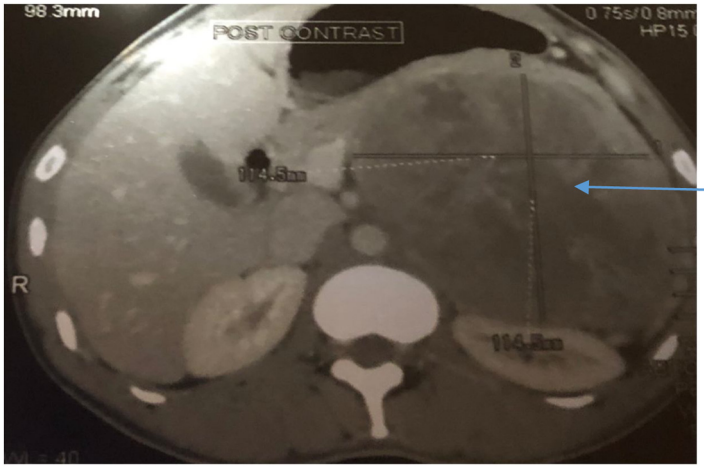

Fig. 1. Tumour (arrow) in transverse section.

vides high sensitivity in the diagnosis of pheochromocytoma [12]. Alpha blockade is not necessary as these tumours are biochemically inert. A multidisciplinary approach is advocated for, for successful management of phaeochromocytoma. Care must be taken when handling pheochromocytoma intraoperatively as release of catecholamines can trigger a hypertensive crisis. Early isolation of the venous drainage of the tumour reduces the risk of hypertensive crises [9].

The majority of pheochromocytomas including the giant category are benign. Histopathological morphological features will not distinguish benign tumours from malignant ones. This diagnosis of a malignant one is only confidently made by presence of metastases. The pheochromocytoma of adrenal origin scaled score (PASS) is used to try to predict malignant behavior [13]. Biologically aggressive tumours have been found to generally have a PASS score $\geq$4 [15]. However, this is not definitive, and is only suggestive of malignancy. Histopathological diagnosis of malignant pheochromocytoma is made if ectopic chromaffin cells are detected in the extra-adrenal sites [15].

2. Case report

A previously well 37-year-old male patient, presented to the general surgery outpatient department at a public and teaching hospital, complaining of dull left lumbar pain and abdominal discomfort which was intermittent for six weeks. He was referred to the outpatient clinic because of worsening discomfort and early satiety. He had no co-morbid conditions and there was no family history of malignancy. He did not smoke nor drink alcohol.

On examination he was well-looking, apyrexial, normotensive and not in any distress. On abdominal examination, there was left flank fullness on inspection and on palpation a mass approximately 14 cm was palpable in the left upper quadrant. The mass was non-tender, non-pulsatile and not expansile. Examination of other systems was normal.

A preoperative CT scan showed a large retroperitoneal tumour 14.3 cm by 12.3 cm arising from the left adrenal gland. The tumour was pushing the diaphragm superiorly, the stomach medially and the left kidney posteriorly as shown in Figs. 1 and 2. A diagnosis of a giant adrenal mass, possibly pheochromocytoma was made and the patient was admitted for biochemical and haematological workup as well as blood pressure monitoring. Wide consultation was made and this included Endocrinologists, Physicians, Radiologists, Pathologists, Laboratory scientists and Surgeons.

The biochemical and haematological results were normal as shown in Table 1.

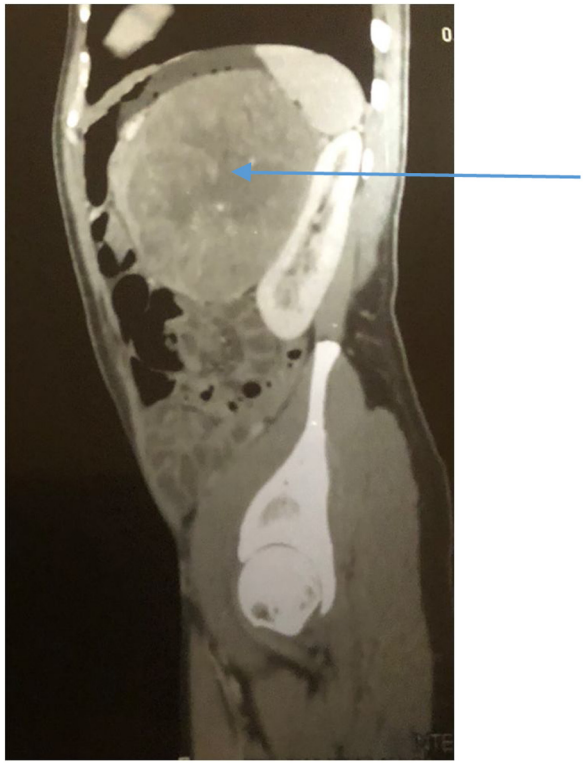

Fig. 2. Tumour (arrow) in sagittal section.

Table 1
Biochemical and Haematological results.

TEST	RESULT
Sodium	143mmol/l
Potassium	3.8 mmol/l
Urea	3.3 mmol/l
Creatinine	82 umol/l
White cell count	$7.2 \times 10\hat{9}$/l
Haemoglobin	12 g/dl
Platelets	$151 \times 10\hat{9}$/l

High performance liquid chromatography(HPLC) to measure serum metanephrines was not available at our institution at the time, so 24-hour urine collection for vanillylmandelic acid(VMA), homovanillic acid (HVA) and metanephrines was commenced. The following are the results on Table 2.