Abstract:
Pheochromocytoma and hyper-aldosteronism are endocrine causes of surgically correctable hypertension. Other causes include co-arctation of aorta and renal artery stenosis. Approach to the management of pheochromocytoma including per-operative preparation, intra-operative management and post-operative follow-up is unique and distinct from that of other causes of hypertension. A lady aged 45, presented to the OPD with history of dizziness and recurrent fainting attacks. On examination, she was found to be hypertensive. Analysis of a 24-hr urine sample revealed elevated levels of metanephrines. CECT of the abdomen revealed a left adrenal tumour, with an increased uptake on MIBG scintigraphy. She was diagnosed with a left adrenal pheochromocytoma. After pre-operative preparation with an alpha-blocker (Prazosin) and volume expansion, followed by beta-blockade, she was posted for surgery. A left adrenal pheochromocytoma was excised, which was followed by a sustained drop in blood pressure. The post-operative period was uneventful. She is currently on regular follow-up. Although Phenoxybenzamine is the gold standard, Prazosin was the alpha-blocker used for pre-operative preparation in this patient. It is preferable to employ a trans-peritoneal approach for all adrenal medullary tumours. This is because of the possibility of multiple functional adrenal medullary tumours, which need to be explored and excised in the event of sustained or resurgent hypertension after excision of an apparently solitary functional tumour. This will be facilitated by a trans-peritoneal approach.

Keyword: Pheochromocytoma, Prazosin, Phenoxybenzamine, Adrenalectomy, Propranolol

Introduction:
Causes of surgically correctable hypertension include adrenal tumours like pheochromocytoma, hyper-aldosteronism and other causes.
like co-arctation of the aorta and renal artery stenosis. Patients with endocrine causes, including pheochromocytoma and hyperaldosteronism, are managed in this department. Pheochromocytoma is a tumor of the chromaffin cells in the adrenal medulla, sympathetic paraganglia and parasympathetic paraganglia, which synthesizes, secretes and metabolizes catecholamines. The possibility of functional neuroendocrine tumours including pheochromocytomas must be considered in the initial evaluation of a patient with recently diagnosed hypertension. Such patients can have a stormy clinical course including hypertensive crises. However, removal of all functional neuroendocrine tumour tissue may result in cure. Approach to the management of pheochromocytoma including per-operative preparation, intra-operative management and post-operative follow-up is unique and distinct from that of other causes of hypertension. One such case is reported here.

Case report:
A lady aged 45 presented to the outpatient clinic of this department with history of dizziness, which she had been having for a period of six months. She had fainted on a couple of occasions. Having been examined and diagnosed with hypertension, she was put on antihypertensive therapy with 5 mg/day of Amlodipine. As part of the work-up for investigation of the cause of hypertension, an ultrasonogram of the abdomen was done, which revealed a mass supero-medial to the upper pole of the left kidney. On examination, her pulse rate was 94/min and the blood pressure was 150/90 mm of Hg. While there was no obvious mass on examination per abdomen, blood pressure recorded during deep palpation of the left lumbar region was 160/100 mm of Hg, suggestive of pheochromocytoma.
Metanephrine levels, estimated in a 24-hr urine sample, were elevated to 7.60 mg/24 hrs (upper limit of normal range - 1 mg/24 hrs). A CECT of the abdomen was done, which revealed a mass in the region of the left adrenal gland, measuring 10 cm x 6 cm x 4 cm (Figures 1, 2). There was no other mass lesion evident elsewhere in the abdomen and pelvis. MIBG scintigraphy, which was performed after blocking the thyroid with Lugol's iodine, revealed a hotspot corresponding to the left adrenal gland (Figure 3). A diagnosis of pheochromocytoma of the left adrenal gland was made. She was advised to take plenty of fluids and high-salt diet in order to expand the intravascular volume. She was started on alpha-blocker therapy with Prazosin (6 mg/day in three divided doses) for control of the hypertension. The intravascular volume was further augmented with transfusion of 3 units of whole blood. Response to alpha-blocker therapy was monitored by regular blood pressure recordings. After four weeks of alpha-blocker therapy, the blood pressure recorded was 110/80 mm of Hg. She was then started on Propranolol (20 mg/day). Six hours later, the
pulse recorded was 94/min and the blood pressure was 150/90 mm of Hg. Over the next four days, the pulse rate settled down to 72/min and the blood pressure to 120/70 mm of Hg. One week after starting her on Propranolol, the patient was posted for surgery. The adrenal mass was approached trans-peritoneally through a midline laparotomy. Blood pressure recorded at induction was 190/110 mm of Hg. A soft, vascular lesion, measuring 10 cm x 6 cm x 4 cm was found in the region of the left adrenal gland (Figure 4), which was excised (Figures 5, 6). This was followed by a drop in blood pressure to 100/60 mm of Hg, which was sustained over a period of 15 minutes. Haemostasis was ensured and the abdominal wound was closed. The diagnosis was confirmed on histopathological examination of the specimen (Figure 7). Her pulse rate in the post-operative period varied between 72/min and 82/min. Post-operative blood pressure ranged from 110/60 mm of Hg to 140/90 mm of Hg, without antihypertensive therapy. The patient was discharged on the 12th post-operative day. At the time of discharge, the pulse rate was 72/min and blood pressure was 120/70 mm of Hg. She has not been taking antihypertensive medication post-operatively. Four months after her surgery, the pulse recorded was 74/min and the blood pressure was 120/70 mm of Hg. She is currently on regular follow-up.

Discussion
According to reports around the world, adrenocortical adenoma causing Conn’s syndrome is the most common adrenal tumour causing hypertension. On the other hand, in this centre, which is a tertiary care centre with a dedicated department of endocrine surgery, 36 patients had pheochromocytoma while there was a single patient with hyper-aldosteronism over the past 3 years. Patients undergoing adrenalectomy for pheochromocytoma undergo preoperative preparation in order to control the blood pressure and expand the intravascular volume. Although Phenoxybenzamine is the gold standard, Prazosin was used in this patient for control of blood pressure. Phenoxybenzamine, being a non-competitive alpha blocker, causes blockade of alpha-1 as well as alpha-2 receptors. While the resultant vasodilatation is beneficial, Phenoxybenzamine causes reflex tachycardia and a profound fall in blood pressure in the post-operative period. If a selective alpha-1 blocker like Prazosin is used, these adverse effects are avoided. If Phenoxybenzamine is used in the pre-operative period, then it is preferable to stop Phenoxybenzamine and start the patient on selective alpha-1 blockers like Prazosin 48 hours before surgery. In the process of pre-operative preparation of the patient, Propranolol, which affectively controls tachycardia and prevents arrhythmias, is employed. If Propranolol is administered without adequate alpha blockade, it results in unopposed stimulation of the alpha receptors by the circulating catecholamines, which causes a paradoxical increase in blood pressure. To prevent this complication, beta blocker therapy is started only after adequate alpha blockade has been achieved. It is preferable to employ a trans-peritoneal approach for all adrenal medullary tumours. This is because of the possibility of multiple functional adrenal medullary tumours, which need to be explored and excised in the event of sustained or resurgent hypertension after excision of an apparently solitary functional tumour. This will be facilitated by a trans-peritoneal approach. Per-operative drop of blood pressure, if sustained for a period of 10 – 15
minutes, indicates that all functional tissue has been excised. This was observed in the case reported above. While the post-operative course has been uneventful till date in the present patient, such individuals need to be followed up for the rest of their lives due to the possibility of development of similar tumours later in life.

Bibliography: