

## Case Report

### A Case Report of Adrenal Pheochromocytoma.

SK Biswas<sup>1</sup>, MJ Alam<sup>2</sup>, MM Rahman<sup>3</sup>, MM Rahman<sup>4</sup>, L Sanjowal<sup>5</sup>, MMSU Islam<sup>6</sup>.

#### Abstract

Pheochromocytoma is a rare catecholamine secreting tumour originating usually from adrenal medulla and produces signs and symptoms due excessive catecholamine secretion from tumour. A young female patient of 21 year age presented with paroxysmal attacks of hypertension causing palpitation, dizziness, blurring of vision and headache for last 06 months. Clinical suspicion of pheochromocytoma was confirmed by 24 hour urinary catecholamine level, transabdominal USG and CT scan of abdomen. After having two weeks of preoperative preparation with phenoxybenzamine and propranolol, open surgical removal of pheochromocytoma was done. Perioperative fluctuation of BP was well managed by IV fluid overload, intravenous phentolamine, intravenous esmolol and intravenous ephedrine. Postoperative recovery was uneventful and BP regains to normal range from 1st postoperative day. Pheochromocytoma is a rare cause of hypertension. If the diagnosis of pheochromocytoma is overlooked, the consequences could be disastrous, even fatal; however, if a pheochromocytoma is identified, it is potentially curable, as being one of the cause of surgically correctable hypertension.

Key Words : Pheochromocytoma, catecholamine, hypertension, alpha blocker.

#### Introduction

Pheochromocytoma is a rare tumor originating from catecholamine secreting chromaffin cells that are derived from the ectodermic neural system and mostly situated within the adrenal medulla<sup>1</sup>. It was suggested

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1. Dr. Swapan Kumar Biswas, FCPS (Surgery), MRCS (Edin), Asst. Professor, Dept. of Surgery, FMC, Faridpur.
  2. Dr. Md. Jahangir Alam, DA, Asst. Professor, Dept. of Anaesthesiology, FMC, Faridpur.
  3. Dr. Md. Mizanur Rahman, FCPS (Surgery), Senior Consultant, Dept. of Surgery, FMCH, Faridpur.
  4. Dr. Md. Mahafuzur Rahman, MBBS, Asst. Registrar, Dept. of Surgery, FMCH, Faridpur.
  5. Dr. Lipika Sanjowal, DA, MCPS (Anaesthesiology), Asst. Professor, Dept. of Anaesthesiology, Diabetic Association Medical College, Faridpur.
  6. Dr. M. M. Shahin-Ul-Islam, FCPS (Medicine), MD (Gastroenterology), Asst. Professor, Dept. of Gastroenterology, FMC, Faridpur.

#### Address of Correspondence:

Dr. Swapan Kumar Biswas, FCPS (Surgery), MRCS (Edin), Asst. Professor, Dept. of Surgery, FMC, Faridpur. Phone: 088-01712120828. Email: swapan\_kb@yahoo.com

that most doctors will meet only 1 patient with a pheochromocytoma in their working lifetime and a large general hospital will admit on an average 1 patient per year<sup>2</sup>. Because of excessive catecholamine secretion, pheochromocytoma may precipitate life-threatening hypertension or cardiac arrhythmias. Pheochromocytoma is fascinating and challenging to clinicians because it combines lethal potential if untreated with possible long term cure in the majority if diagnosed and treated surgically. We present a case report of adrenal pheochromocytoma of 21 year aged young female patient presenting with paroxysmal attacks of hypertension which was treated successfully by open surgical removal of pheochromocytoma in surgery unit-II of Faridpur Medical College Hospital.

#### Case Report

Mrs. Afroza, 21 years old housewife admitted into FSW-II of FMCH with the complaints of paroxysmal attacks of palpitation, dizziness, blurring of vision and headache for last 06 months. Each attack persists for few minutes to half an hour and occurs irregularly once within two to three days to 2 to 3 times a day. On examination, patient had no abnormal physical findings except Blood Pressure (BP) is high during paroxysmal attack (Systolic BP varies from 140 to 210 mmHg and diastolic BP varies from 100 to 140 mmHg). Complete

Blood Count, Random Blood Sugar, Blood urea, Chest X-rays and ECG reports were within normal limit. We made a provisional diagnosis of secondary hypertension due to adrenal pheochromocytoma but for confirming this diagnosis we need to do biochemical investigations i.e. serum catecholamine level or 24 hour urinary catecholamine level which was not locally available. Abdominal USG shows right sided adrenal mass of 8 cm in diameter. Later 24 hour urinary catecholamine and metanephrine level were done at BIRDEM, Dhaka which confirmed biochemical diagnosis of pheochromocytoma. CT (Computed Tomography) scan of abdomen shows right sided adrenal mass of a size about 6cm x 10 cm.

We planned for surgical removal of right adrenal gland (Rt. Adrenalectomy). Patient was prepared for surgery with collaboration with Cardiologist and Anesthesiologist. Patient was given Phenoxybenzamine (alpha blocker) for 10 days preoperatively, starting with 10 mg 12 hourly and gradually increasing up to 30 mg 12 hourly and BP downs to normal level (Systolic BP 110 to 80 mmHg and Diastolic BP 70 to 50 mmHg). Patient was also given Propranolol (beta blocker) 10 mg three times daily starting from 11th day of preoperative preparation for 5 days. Operation was done on 16th day of preoperative preparation and last dose of drugs were given on the morning of the day of surgery.

Open right adrenalectomy was done by right subcostal incision. Adrenal vein was ligated first and tumour was removed. Operative procedure was uneventful except marked fluctuation in BP. BP increased up to a level of 260/150 mmHg during handling of tumour and fell to a non recordable BP following ligation of adrenal vein. Our experienced anaesthesiologist team well managed the situation using preoperative IV fluid overload, intravenous phentolamine (Rapid acting alpha adrenergic antagonist) and Intravenous esmolol (Rapid acting beta adrenergic antagonist) during operative mobilization and removal of tumour. A sudden fall of blood pressure following ligation of adrenal vein was managed by rapid infusion of large volumes of fluid and intravenous ephedrine.

Postoperative recovery was uneventful and patient was discharged on 8th Post Operative Day. Blood pressure becomes normal (Systolic 110 to 90 mmHg and diastolic 80 to 60 mmHg) from 1st POD without any drug. Finally, Histopathology report further confirmed the adrenal tumour was pheochromocytoma.



Fig. 1: Photograph of patient, Mrs. Afroza 21 Years.



Fig. 2: USG showing right adrenal mass



Fig. 3: CT scan showing right adrenal mass



Fig. 3: Operative specimen of pheochromocytoma

## Discussion

Pheochromocytoma is a rare neoplasm, which are derived from cells of the chromaffin tissue and mostly situated within adrenal medulla. Only approximately 15% Pheochromocytoma develops from extra-adrenal chromaffin tissue which lies in the paraganglion

chromaffin tissue of the sympathetic nervous system extending from base of skull to the urinary bladder. Common locations of extra-adrenal Pheochromocytomas include the organ of Zuckerkandl (close to origin of the inferior mesenteric artery), urinary bladder wall, heart, mediastinum and carotid and glomus jugulare bodies<sup>3</sup>.

Pheochromocytomas occur in people of all races, although they are diagnosed less frequently in blacks and equal frequency in male and female. Pheochromocytomas may occur in persons of any age. The peak incidence, however, is between the third and the fifth decades of life. Approximately 10% occur in children. The majority of cases are sporadic, with only 16% having a history of associated endocrine disorder such as Multiple Endocrine Neoplasia type II (MEN IIA and IIB), Neurofibromatosis 1 (NF 1) and von Hippel-Lindau disease (VHL)<sup>4,5</sup>. Approximately 10% of pheochromocytomas are malignant<sup>5</sup>. Direct invasion of surrounding tissue or the presence of metastases determines malignancy. Unfortunately, no reliable clinical, biochemical or histological features distinguish a malignant from a benign pheochromocytoma.

The clinical manifestations of a pheochromocytoma results from excessive catecholamine secretion by tumour. Catecholamines typically secreted, either intermittently or continuously, includes norepinephrine and epinephrine; rarely dopamine is secreted. The biological effects of catecholamines are well known. Catecholamine secretion in pheochromocytoma is not regulated in the same manner as in healthy adrenal tissue. Relative catecholamine levels also differ in pheochromocytoma. Most pheochromocytomas secretes norepinephrine predominantly, where as secretions from normal adrenal medulla are composed of 85% epinephrine<sup>6</sup>. The classic history of a patient with pheochromocytoma includes spells (Paroxysms) characterized by headaches, palpitations and diaphoresis in association with severe hypertension. The spells may vary in occurrence from monthly to several times per day and the duration may vary from seconds to hours. Paroxysms may be precipitated by physical training, induction of general anaesthesia and numerous drugs and agents (contrast media, tricyclic antidepressive drugs, metoclopramide and opiates)<sup>7</sup>. Typically, they worsen with time, occurring more frequently and becoming more severe as the tumour grows<sup>5,8</sup>.

The first step in the diagnosis of a pheochromocytoma is the biochemical confirmation of catecholamine excess. Plasma metanephrine testing has the highest sensitivity (96%) for detecting a pheochromocytoma, but it has a lower specificity<sup>9</sup>. In comparison, a 24 hour

urinary collection for catecholamines and metanephrines has a sensitivity of 87.5% and a specificity of 99.7%<sup>10</sup>. The biochemical diagnosis is followed by the localization of the pheochromocytoma and/ or metastases. Magnetic Resonance Imaging (MRI) is preferred over Computed Tomography (CT) scanning because contrast media used for CT scans can provoke paroxysms. In addition MRI has a reported sensitivity of up to 100% in detecting adrenal pheochromocytoma. <sup>123</sup>I-MIBG (Iodine-131 labeled metaiodobenzylguanide) scanning is reserved for cases in which a pheochromocytoma is confirmed biochemically but CT scan or MRI does not show a tumour.

Surgical resection of the tumour is the treatment of choice and usually results in cure of hypertension. Careful preoperative preparation requires with combined alpha and beta blockade to control blood pressure and to prevent intraoperative hypertensive crisis<sup>11</sup>. Alpha-adrenergic blockade, in particular, is required to control blood pressure and prevent a hypertensive crisis. Phenoxybenzamine is the preferred alpha blocker in preparation for surgery. A dose of 20 mg of phenoxybenzamine initially, should be increased daily by 10 mg until a daily dose of 100-160 mg is achieved and the patient reports symptomatic postural hypotension<sup>7</sup>. Others alpha blocker such as Doxazosin, Prazosin and Terazosin are only rarely used because of their incomplete alpha blockade. Additional beta ( $\beta$ ) blocked is required if tachycardia or arrhythmias develop; this should not be introduced until the patient is alpha blocked because unopposed alpha adrenergic receptor stimulation can precipitate hypertensive crisis<sup>5,7,8</sup>. Non cardioselective beta blockers, such as Propranolol or Nadolol are often used; however, cardioselective agents, such as Atenolol and Metoprolol, also may be used.

An experienced anaesthesiologist and an experienced surgeon are crucial to the success of operation. During surgery for pheochromocytoma there is a risk of hypertensive crisis due to excessive catecholamine release with handling of tumour or hypotensive crisis following ligation of adrenal vein which cannot be completely precluded even with preoperative alpha blockade<sup>4</sup>. Central venous catheter, invasive arterial monitoring and cardiac monitoring are essential. Preoperative fluid overload is advisable to occupy the sudden expansion of the vascular bed when the tumour is removed. During surgery intravenous infusions of alpha blocking drug (Intravenous phentolamine, a rapid acting alpha adrenergic antagonist) and beta blocking drug (Intravenous esmolol, a rapid acting beta

adrenergic antagonist) are given, if required, as determined by the blood pressure, pulse rate and central venous pressure. Sodium nitroprusside, a direct peripheral vasodilator is also used for management of intraoperative hypertension. A sudden fall of blood pressure is usual when venous drainage of the tumour has been ligated, at this point rapid infusion of large volumes of fluid and intravenous ephedrine may be necessary as vascular compartment expands.

Surgical management has progressed through the years. Prior to introduction of laparoscopic adrenalectomy, thoracoabdominal approach was utilized at some centers, more commonly the midline abdominal and flank approaches have been used<sup>12</sup>. Since the first laparoscopic adrenalectomy for Pheochromocytoma was done in 1992, it has been performed in numerous centers with excellent success over past decade<sup>13-15</sup>. Laparoscopic adrenalectomy is comparable to open approach, and should be considered preferentially in patients with tumour less than 6 cm. For larger or extraadrenal tumour an open approach is favoured<sup>4,16,17</sup>. Biochemical cure should be confirmed by assay of 24 hour urinary catecholamine 2-3 weeks after surgery and the lifelong urinary catecholamine measurement should be performed to identify recurrent or metachronous pheochromocytoma.

## Conclusion

Pheochromocytoma is often called '10% tumour' because 10 percent are bilateral, malignant, extra-adrenal, multiple, familial and occur in children<sup>18</sup>. Pheochromocytoma is one of the few causes of hypertension that can be treated surgically. Although it is the causative factor of hypertension in about 0.1% to 0.6% of the hypertensive population, detection is mandatory, not only for the potential cure of the hypertension but also to avoid the potentially lethal effects of the unrecognized tumor<sup>19</sup>.

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