Abstract:
Pheochromocytoma is a tumor of the catecholamine-producing cells of the adrenal medulla. These tumors produce excess catecholamine, which if missed or not properly treated, will almost prove fatal. Therefore prompt diagnosis and treatment is necessary. We present a case of bilateral pheochromocytoma in a young male who presented with complaints of sweating, headache, tachycardia and hypertension. Biochemical evaluation showed excess catecholamine production. Imaging studies showed bilateral adrenal mass. Patient underwent bilateral adrenalectomy after thorough preoperative preparation. Histopathology examination confirmed pheochromocytoma bilaterally.

Keyword: Pheochromocytoma, Adrenal, Hypertension

INTRODUCTION:
Pheochromocytoma is a rare catecholamine producing tumor arising from chromaffin cells of the adrenal gland. These tumors commonly presents in fourth or fifth decade, accounting for 0.5% of hypertension. Bilateral adrenal Pheochromocytoma is very rare condition occurring in 10% of the cases, most commonly associated with familial Pheochromocytoma. Bilateral disease occurring in a sporadic case is very rare. We present a case of bilateral pheochromocytoma in a young male without any family history.

CASE REPORT:
A 21 year old male presented with complaints of sweating and headache for past two years. There was no significant past and family history. On clinical examination he had tachycardia (108/minute) and hypertension (130/100 mm of Hg). Per abdomen examination was normal. Laboratory investigation showed normal Hemogram, renal function test and electrolytes. Ultra sonogram of the abdomen showed bilateral adrenal mass (right adrenal 4.8x3.8 cms, left adrenal 4.6x4.2 cms). Biochemical evaluation showed increased urine normetanephrines – 12551 micrograms/24 hours urine (normal range – 88 to 444) :1).
and increased serum normetanephrine -593pg/ml (normal range – 95 to 446pg/ml). Contrast enhanced computer tomography showed heterodense bilateral adrenal mass with right measured 4.8x 3.8 cms size, left adrenal mass measured 4.6x4.2 cms size and bilateral kidneys were normal (Figure 1).

Figure: 1- CECT Abdomen showing axial and coronal section of bilateral adrenal mass

His serum calcium was normal and computer tomography of brain was also normal. Serum cortisol and aldosterone levels were normal. Patient was started on alpha blocker phenoxybenzamine 10 mgs twice daily. His blood pressure came to normal (110/80 mm of Hg). As he developed tachycardia, metoprolol 25 mg was added. These drugs were given for three weeks. Patient was also infused with two liters of crystalloids daily for four days before surgery. Anesthetist was well informed of this case before posting for surgery. Abdomen was opened by chevron incision and bilateral adrenalectomy done transperitoneally. (Figure: 2).

Figure: 2 – Intraoperative picture of right adrenalectomy

No other mass seen along the aorta. Intraoperatively there is a transient fall of blood pressure (10 mm of Hg) after removal of tumor. Specimen sent for HPE. (Figure: 3)

Figure: 3 – Right and left adrenalectomy Specimen. 3 b – cut section.

Post operative period was uneventful and his blood pressure was normal without any drug. He was started on methylpredisolone initially, and then given oral prednisone as replacement therapy. Histopathology examination confirmed pheochromocytoma bilaterally. (Figure: 4)

Figure: 4 – HPE shows nests of tumor cells (zellballen) surrounded by a discontinuous layer of sustentacular cells and fibrovascular stroma
Sutures were removed on 14th post operative day. During discharge his blood pressure and pulse rate were normal. Patient was followed up after one month and was normotensive without any symptoms. His post operative Serum cortisol and aldosterone level were within normal limits.

**DISCUSSION:**
Pheochromocytomas are rare catecholamine producing tumors arising from chromaffin cells. It is estimated that pheochromocytoma is responsible for only approximately 0.5% of cases of hypertension. One to five percent of pheochromocytomas originate outside of the adrenal gland. These extra-adrenal pheochromocytomas are known as paragangliomas, because they arise from paraganglia, a network of chromaffin-producing neural crest tissue that anatomically parallels the sympathetic and parasympathetic ganglia. Pheochromocytoma has been called the “10% tumor”: 10% extra-adrenal, 10% familial, 10% bilateral, 10% pediatric and 10% malignant. Sporadic cases of pheochromocytoma are most often diagnosed in the fourth and fifth decades of life, while familial tumors tend to occur at a younger age. The clinical presentations of Pheochromocytomas are diverse and the tumor can mimic a variety of conditions, often resulting in erroneous and delayed diagnosis, therefore called as “great mimic”. Paroxysmal hypertension is the classic presenting symptom in patients with pheochromocytoma, but reported only in 30% to 50% of patients. The remainder of patients demonstrates persistently elevated blood pressure, and minorities are entirely normotensive. The triad of headache, episodic sudden perspiration, and tachycardia is a classic hallmark of pheochromocytoma. Other clinical symptoms were Palpitations, sweating, pallor, nausea, flushing, weight loss, tiredness, psychologic symptoms (anxiety, panic), orthostatic hypotension and hyperglycemia.

Bilateral pheochromocytoma is a rare tumor in a young patient without any family history or associated with hereditary syndromes like Von Hippel Lindau syndrome, MEN type 2 syndromes. Patient should be adequately prepared before surgery in order to prevent intraoperative rise in blood pressure while handling the mass. If patient develops tachycardia after alpha blocker, beta blocker can be given to prevent unopposed action of catecholamine on beta receptors. Intra vascular volume must be loaded preoperatively in order to prevent severe hypotension once mass is removed. Bilateral tumor was best approached transperitoneally by chevron incision. It gives the added advantage of inspecting for other masses in the paraganglionic area along the aorta. Post operatively after bilateral adrenalectomy patient must be supplemented with cortisol and if needed aldosterone. Patient should be warned of need for increased dose of cortisol in stressful situation like fever, trauma or emotional stress.

**CONCLUSION**
Bilateral Pheochromocytoma in a young male presenting as a sporadic case is a rare presentation. Prompt diagnosis, thorough preoperative evaluation and preparation are necessary to have good surgical outcome.

**REFERENCE**
3 A patient with bilateral pheochromocytoma as part of a VonHippel-Lindau (VHL) syndrome type 2C World Journal of Surgical Oncology 2007, 5:112