Annals of Hypertension

Case Report
Published: 27 Jun, 2018

A Case Report of a Giant Adrenal Pheochromocytoma

Margarita Gjata, Eneida Hoxha, Joniada Xhanaj, Ilir Gjermeni, Ledio Collaku and Mihal Tase*

Department of Internal Medicine, University Hospital Center Mother Teresa, Albania

Abstract

Pheochromocytoma is a rare catecholamine secreting tumor originating usually from adrenal medulla and produces signs and symptoms due excessive catecholamine secretion from tumor. A young female patient of 16 year age presented with paroxysmal attacks of hypertension causing, dizziness, blurring of vision and headache for last 5 months. Clinical suspicion of pheochromocytoma was confirmed by transabdominal USG and CT scan of abdomen. After having two weeks of preoperative preparation with alpha blocker and beta–blocker, open surgical removal of pheochromocytoma was done. Preoperative 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 causes of surgically correctable hypertension.

Keywords: 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 [1]. It was suggested that most doctors will meet only 1 patient with a pheochromocytoma in their working life time and a large general hospital will admit on an average 1 patient per year [2]. 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 16 year aged young female patient presenting with paroxysmal attacks of hypertension which was treated successfully by open surgical removal of pheochromocytoma in surgery unit-Mother Theresa Tirana Hospital.

Case Presentation

Mrs. Dorina, 16 years old teenager, height: 146 cm; weight: 46 kg, admitted into- Mother Theresa Tirana Hospital of with the complaints of paroxysmal attacks of hypertension, dizziness, blurring of vision and headache for last 5 months (Figure 1). Each attack persists for few minutes to half an hour and occurs irregularly once within two to three days to 3 to 4 times a day. On examination, patient had hirsutism and low stature, no menstrual cycle. Blood Pressure (BP) is high during paroxysmal attack (Systolic BP varies from 160 mmHg to 180 mmHg and diastolic BP varies from 100 mmHg to 120 mmHg). Complete blood count had polycythemia HB 17.4 g/dl and RBC 5.750000, 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. Abdominal USG shows right sided adrenal mass of 14.3 cm × 9.2 cm in diameter in superior pole with regular contours with calcifications.

Level or 24 hour urinary catecholamine (normetanephrine, metanephrine and vanilmandelic acid), and tumor markers (carcinoembryonic antigen, Carbohydrate Antigen [CA] 19.9, Ca 125 and beta-human chorionic gonadotropin) were normal. Cortisolemia 08:00 AM, TSH, fT4 were normal.

Sexual hormones FSH and LH were low: FSH 0.310 mIU/ml (1.20-9.00); LH 0.125 mIU/ml (1.1-12.6).

CT (Computed Tomography) scan of abdomen shows right sided adrenal mass of a size about 10.2 cm × 9.3 cm with calcifications and cystic degenerations, that shifts liver margin and superior

OPEN ACCESS

Correspondence:
Margarita Gjata, Department of Internal Medicine, University Hospital Center Mother Teresa, Tirana, Albania, E-mail: mresuli@yahoo.com

Received Date: *10 May 2018*
Accepted Date: *22 Jun 2018*
Published Date: *27 Jun 2018*

Citation:
Gjata M, Hoxha E, Xhanaj J, Gjermeni I, Collaku L, Tase M. A Case Report of a Giant Adrenal Pheochromocytoma. Ann Hypertens. 2018; 1(1): 1001.

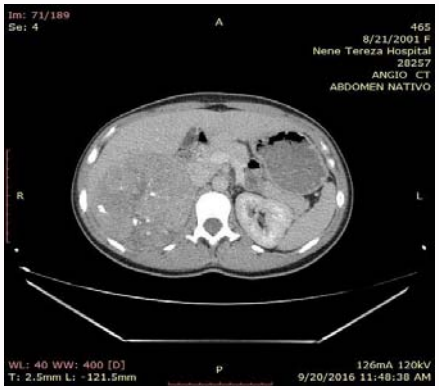

Figure 1: Angio CT abdomen native.

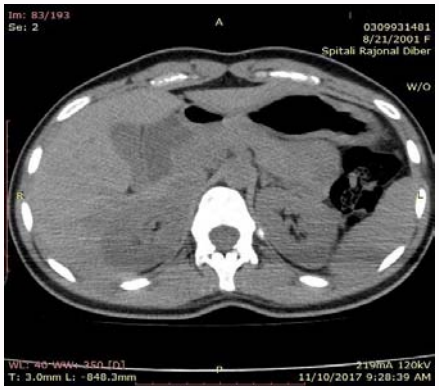

Figure 2: Axial CT with contrast showing superior.

pole of right kidney, but without clear confines with them (Figure 2).

ABPM before surgery

SBP 137 (110-164) mmHg, DBP 94 (67-126) mmHg, HR 70 (51-101) bpm, PP 43 (25-75) mmHg. Optimal 8%, Normal 11%, High normal 20%, Grade I 27%, Grade II 17%, Grade III 13%, Isolated systolic hypertension 4%.

Echocardiography

Diastolic LV internal dimension 49 mm; Systolic LV internal dimension TSVM 34 mm; Interventricular septum wall thickness 9 mm; Posterior wall thickness 9 mm; Left atrium dimension 33 mm; LV Ejection fraction 058%.

We planned for surgical removal of right adrenal gland (Rt. Adrenalectomy). Patient was prepared for surgery Anesthesiologist. Patient was given Doxazosin 2 mg (alpha blocker) for 10 days preoperatively. Patient was also given Metoprolol (beta blocker) 50 mg twice times daily starting from 11th day of preoperative preparation for 3 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 tumor was removed. Operative procedure was uneventful except marked fluctuation in BP. BP does not increased up to a level of 160/100 mmHg during handling of tumor. Postoperative recovery was uneventful and patient was discharged on 7th Post Operative Day. Blood pressure becomes normal from 3rd POD without any drug. ABPM after surgery SBP 127 (89-156) mmHg, DBP 85 (67-105) mmHg, HR 92 (59-132) bpm, PP 35 mmHg. Optimal 11%, Normal 20%, High normal 28%, Grade I 18%, Grade II 12%, Grade III 7%, Isolated systolic hypertension 4%.

Finally, Histopathology report further confirmed the adrenal tumor was pheochromocytoma.

Discussion

Pheochromocytomas occur in people of all races, although they are diagnosed less frequently in blacks and equal frequency in male and female [1-3]. 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 (NF1) and von Hippel-Lindau disease (VHL) [4,5].

Approximately 10% of pheochromocytomas are malignant [5]. 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 tumor. Catecholamine 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 [6]. 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 anesthesia and numerous drugs and agents (contrast media, tricyclic anti depressive drugs, metoclopramide and opiates) [7]. Typically, they worsen with time, occurring more frequently and becoming more severe as the tumor grows [5,8]. The five tumors most commonly associated with the overproduction of erythropoietin (i.e., paraneoplastic erythrocytosis) are hepatocellular carcinoma, renal cell carcinoma, hemangioblastoma, pheochromocytoma, and uterine myomata [9]. Giant pheochromocytomas (>7 cm in size) are rare entities with around 20 cases reported in the literature [10-15]. They do not present with the classical symptoms of pheochromocytomas [15]. Most patients present with vague discomfort while others may complain of a palpable abdominal mass. Operative surgery is the ideal management option [16]. There needs to be a multidisciplinary approach while managing such cases. Stringent preparation to combat crisis due to catecholamine surge (during tumor manipulation) and sudden decrease in peripheral vascular resistance (following lesional excision) need to be emphasized [17,18]. Presence of chromaffin cells in the extra-adrenal tissue is the only confirmative method of distinguishing the malignant variant from its benign counterpart [19]. Herein we highlight and discuss the management algorithm taken while managing one such case. 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 [20]. In comparison, a 24 hour urinary collection for catecholamines and metanephrines has a sensitivity of 87.5% and a specificity of 99.7% [21]. 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 [1-3]. 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 tumor [1-3]. Surgical resection of the tumor 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 [22]. 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 mg to 160 mg is achieved and the patient reports symptomatic postural hypotension [7]. Others alpha blocker such as Doxazosin, Prazosin and Terazosin are only rarely used because of their incomplete alpha blockade. Additional 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 [5,7,8]. Non cardioselective beta blockers, such as Propranolol or Nadolol are often used; however, cardioselective agents, such as Atenolol and Metoprolol, also may be used.

Conclusion

Pheochromocytoma is often called 10% tumor because 10 percent are bilateral, malignant, extra adrenal, multiple, familial and occur in children [23]. 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 [24].

References

1. Neumann HP. Pheochromocytoma. In: Fauci AS, Braunwald E, Kasper DL, Hauser SL, Longo DL, Jameson JL, et al, editors. Harrisons Principles of Internal medicine. 17th ed. New York: Mcgraw-Hill medical; 2008.
2. Karet FE, Brawn MJ. Pheochromocytoma: diagnosis and management. Postgrad Med J. 1994;70(823):326-8.
3. Yong L, Sheng-guo D, Zhen D, Xin M, Xin-yan S. Diagnosis and treatment of Pheochromocytoma in urinary bladder. J Zhejiang Univ Sci B. 2007;8(6):435-38.
4. Bentrem DJ, Pappas SG, Ahuja Y, Murayama KM, Angelos P. Contemporary surgical management of Pheocromocytoma. Am J Surg. 2002;184(6):621-5.
5. Bravo EL, Gifford RW jr. Pheochromocytoma. Endocrinol Metab Clin North Am. 1993;22(2):329-41.
6. Sheps SG, Jiang NS, klee GG, van Heerden JA. Recent developments in the diagnosis and treatment of Pheochromocytoma. Mayo Clin Proc. 1990;65(1):88-95.
7. Rothmund M. Adrenal glands and other endocrine disorders. In: Williams NS, Bulstrode CJK, O'Connell PR, editors. Bailey and love's short practice of surgery. 25th ed. London: Hodder Arnold; 2008.
8. Gifford RW, Kvale WF, Maher FT. clinical features, diagnosis and treatment of pheochromocytoma. A review of 76 cases. Mayo Clin Proc. 1964;39:281-302.
9. Drenou B, Le Tulzo Y, Caulet-Maugendre S, Le Guerrier A, Leclercq C, Guilhem I, et al. Pheochromocytoma and secondary erythrocytosis: role of tumour erythropoietin secretion. Nouv Rev Fr Hematol. 1995;37(3):197-9.
10. Sarveswaran V, Kumar S, Kumar A, Vamseedharan MK. A giant cystic pheochromocytoma mimicking liver abscess an unusual presentation - a case report. Clin Case Rep. 2015;3(1):64-8.
11. Wang HL, Sun BZ, Xu ZJ, Lei WF, Wang XS. Undiagnosed giant cystic pheochromocytoma: A case report. Oncol Lett. 2015;10(3):1444-6.
12. Korgali E, Dundar G, Gokce G, Kilicli F, Elagoz S, Ayan S, et al. Giant malignant pheochromocytoma with palpable rib metastases. Case Rep Urol. 2014;2014:354687.
13. Ambati D, Jana K, Domes T. Largest pheochromocytoma reported in Canada: A case study and literature review. Can Urol Assoc J. 2014; 8(5-6):E374-7.
14. Soufi M, Lahlou MK, Benamr S, Massrouri R, Mdaghri J, Essadel A, et al. Giant malignant cystic pheochromocytoma: a case report. Indian J Surg. 2012;74(6):504-6.
15. Li C, Chen Y, Wang W, Teng L. A case of clinically silent giant right pheochromocytoma and review of literature. Can Urol Assoc J. 2012;6(6):E267-9.
16. Staren ED, Prinz RA. Selection of patients with adrenal incidentalomas for operation. Surg Clin North Am. 1995;75(3):499-509.
17. Adenekan A, Faponle A, Badmus T. Anaesthetic management of giant phaeochromocytoma in a patient with chronic renal disease. J West Afr Coll Surg. 2011;1(2):112-22.
18. Antedomenico E, Wascher RA. A case of mistaken identity: giant cystic pheochromocytoma. Curr Surg. 2005;62(2):193-8.
19. Glodny B, Winde G, Herwig R, Meier A, Kühle C, Cromme S, et al. Clinical differences between benign and malignant pheochromocytomas. Endocr J. 2001;48(2):151-9.
20. Kudva YC, Sawka AM, Young WF jr. Clinical review 164: The laboratory diagnosis of adrenal pheochromocytoma: Mayo Clinic experience. J Clin Endocrinol Metab. 2003;88(10):4533-9.
21. De jong WH, Eisenhofer G, Post WJ, Muskiet FA, de Vries EG, Kema IP. Dietary influences on plasma and urinary metanephrines: Implications for diagnosis of catecholamine producing tumour. J Clin Endocrinol Metab. 2009;94(8):2841-9.
22. Wong C, Yu R. Preoperative preparation for pheochromocytoma resection: Physician survey and clinical practice. Exp clin Endocrinol Diabetes. 2010;118(7):400-4.
23. Duh QY, Liu C, Tyrrell B. Adrenals. In: Doherty GM, editor. Current Diagnosis and treatment: Surgery. 13th ed. New York: The Mcgraw-Hill companies; 2010.
24. Lo CY, Lam KY, Wat MS, Lam KS. Adrenal pheochromocytoma remains a frequently overlooked diagnosis. Am J Surg. 2000;179(3):212-5.