RESEARCHERID
THOMSON REUTERS
Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 5 (Issue 1): January 2018 ISSN: 2394-9414
DOI- 10.5281/zenodo.1158603 Impact Factor- 4.174

HEMORRHAGIC STROKE SECONDARY TO PHEOCHROMOCYTOMA - A RARE CASE REPORT

Dr. Vinod Kumar[1], Dr. Darpanarayan Hazra[*2], Dr. Shahid Mahdi[3] & Dr. Shubhash Chandra[4]
[1,*2,3&4] Cardiac Intensive Care, BLK Super Specialty Hospital

Keywords:
Pheochromocytoma, ICH and Pheochromocytoma, Refractory hypertension in ICH.

Abstract

Pheochromocytoma is a tumour in the adrenal medulla and sympathetic Para ganglia, which synthesizes and secretes catecholamines. High circulating levels of catecholamines can lead to severe hypertension and can have devastating effects on multiple body systems (cardiovascular, cerebrovascular, etc.). Pheochromocytoma can be a cause for refractory hypertension in a case of hemorrhagic stroke. We hereby report a case of 39 years old male patient who was diagnosed to have hemorrhagic stroke secondary to Pheochromocytoma. Although surgical treatment represents the only modality of ultimate cure, pharmacological pre-operative treatment remains the mainstay of successful outcome.

Introduction

Pheochromocytoma has been called many names – from a friendly "great mimicker" to a treacherous "cold-blooded killer" (the associated hypermetabolic state would actually make it rather "warm-blooded"), which mostly translates into our fear of missing it, because of highly feasible fatal complications. In addition to being a great mimicker, Pheochromocytoma represents one of the most dazzling clinical paradoxes, where signs and symptoms vary to hardly understandable degrees in seemingly comparable clinical settings.

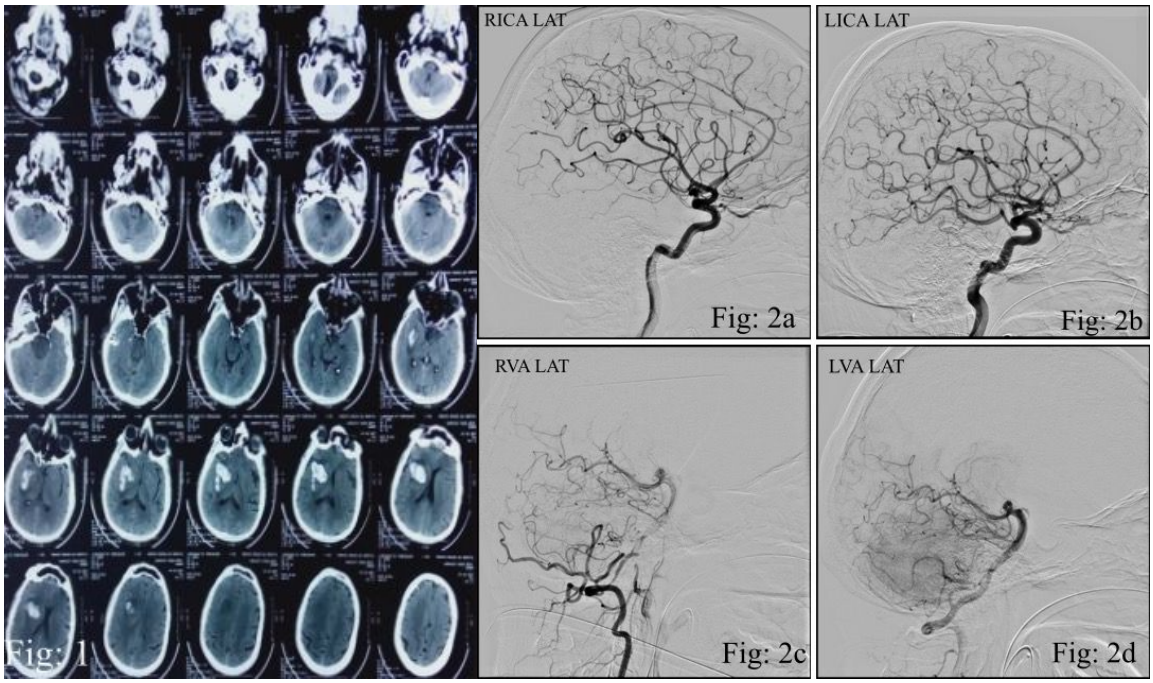
Diagnostic clues are usually provided by additional symptoms attributed to sympathetic activation, such as diaphoresis, palpitation, apprehension, and/or anxiety. The diagnosis is established by the demonstration of increased excretion of catecholamines or catecholamine metabolites, and can usually be verified by analysis of a single 24-hour urine sample collected when the patient is hypertensive or symptomatic.

Case report

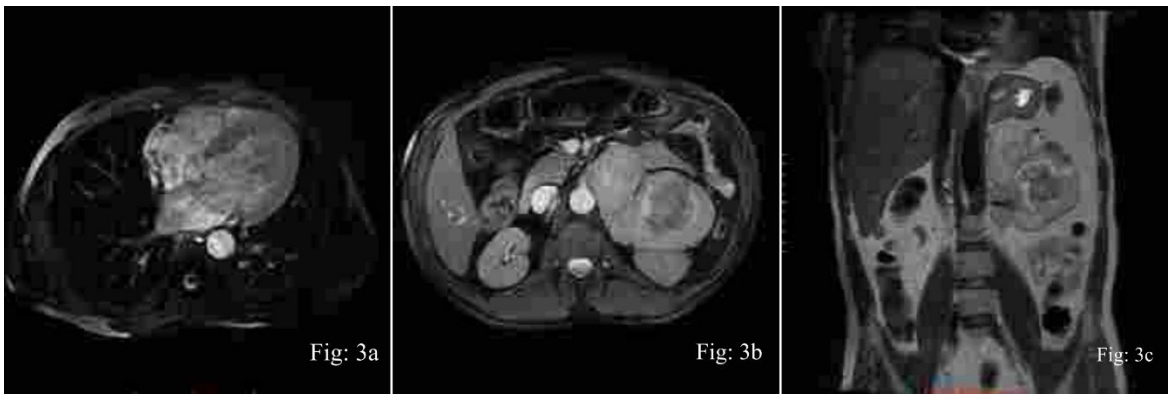
A 39 years old hypertensive gentleman, presented to emergency room with complaints of sudden onset left sided weakness and slurring of speech. He had no history of trauma or fall, seizures, vomiting or similar episode in the past. He had a three year past history of intermittent (mild-severe) headache in the morning which is relieved by taking antihypertensive medications, and frequent episodes of sweating & palpitation during strenuous activities. General examination revealed a drowsy patient with heart rate (62/min), B.P (190/90 mmHg), R.R (26/min) and room air oxygen saturation was 98%. Neurological examination revealed GCS – E3V3M5 with left sided hemiplegia (power-0/5), planter's extensor with preserved limb movements on the right side. Pupils were bilateral equal and reacting to light. Other systemic examination was within normal limits. NCCT brain (Fig: 1) revealed Intracerebral Hemorrhage in right Ganglio-capsular area. He was immediately started on anti-hypertensives, anti-epileptics, osmotic diuretics and other supportive care. Hypertension persisted inspite of initiating the patient on three anti-hypertensive drugs (Nitro-glycerine, alpha blocker and Calcium channel blockers). DSA Brain (Fig: 2a-2d) ruled out any A-V malformation or ruptured aneurysm. In view of refractory hypertension and correlating with his clinical presentation, provisional diagnosis of Pheochromocytoma was made. Lab investigations reported Metanephrine :84.47 µg/g creatinine (20.00-150.00), Normetanerphine :277716.86 µg/g creatinine (70.00-335.00), VMA (Vanilly Mandelic acid) 26.98 mg/g creatinine (<5.50). MR abdomen (3a-3c) was suggestive of a neoplastic adrenal mass. The patient has been put on Alfa blocker since admission, hence Beta blockers and Na Nitroprusside were subsequently added to the treatment regime, to which he responded well. After medical stabilization, total resection of the tumour was achieved via retroperitoneal approach with left subcostal incision by the department of

RESEARCHERID
THOMSON REUTERS
Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 5 (Issue 1): January 2018 ISSN: 2394-9414
DOI- 10.5281/zenodo.1158603 Impact Factor- 4.174

Surgical Gastroenterology. Intraoperative manipulation of the mass caused sudden hypotension which was managed with IV fluids and ionotropic supports. Postoperatively, the patient improved rapidly and & inotropic supports were tapered off the next day of surgery. Histopathological examination confirmed the diagnosis of Pheochromocytoma. He was managed conservatively for the Intracerebral Hemorrhage with oral medications along with primary supportive care. He was discharged on the 11th postoperative day on single anti-hypertensive and anti-epileptic drugs, with a yearly follow up plan of urinary Vanilly Mandelic acid (VMA) and Metanephrine levels.

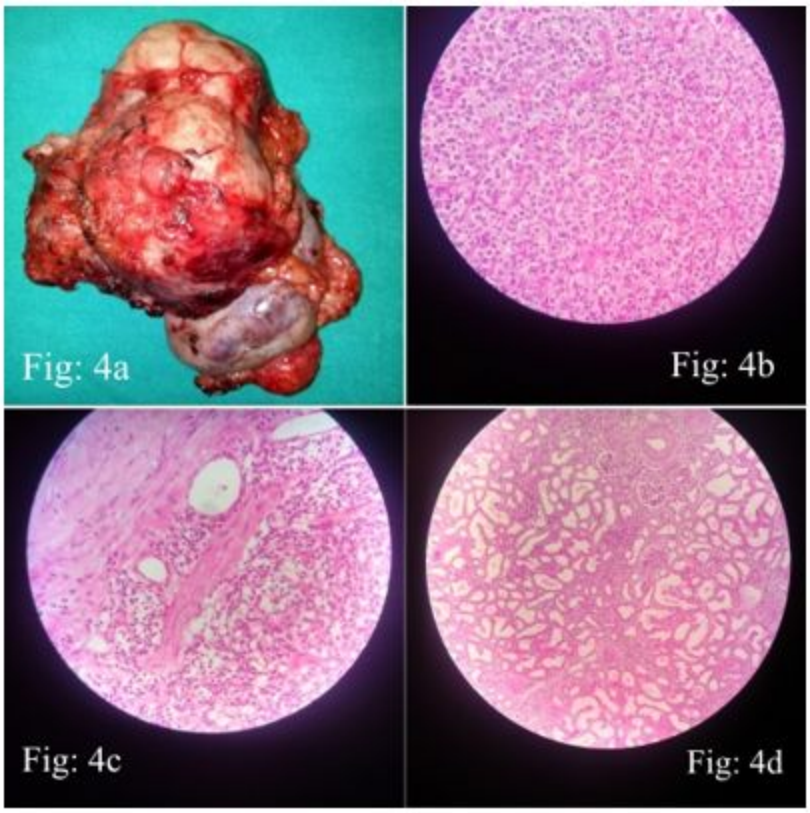


(Fig: 1 NCCT Brain - Intracerebral Hemorrhage in right Ganglio-capsular region extending in Fronto-Temporal lobe with mild surrounding edema and midline shift, Fig: 2a-2d Normal DSA Brain study)



(Fig: 3a-3c- MR abdomen revealing a large well defined lobulated altered signal intensity lesion in left lumbar region with left adrenal gland not separately visualized, showing areas of gradient blooming-which was suggestive of a probable neoplastic adrenal mass)

RESEARCHERID
THOMSON REUTERS
Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 5 (Issue 1): January 2018 ISSN: 2394-9414
DOI- 10.5281/zenodo.1158603 Impact Factor- 4.174



(Fig: 4a-d -External surface of adrenal is lobulated, capsulated with focal breach in capsule. Cut section showed a yellow tan lobulated tumour with areas of necrosis and hemorrhage. Tumour was invading into hilar soft tissue. Kidney appeared unremarkable externally and on cut surface. Immunohistochemistry revealed Synaptophysin, Chromogranin: CD56- Positive in tumour cells. CK: Negative in tumour cells. Ki67 Proliferative index is about 6-7%. Pathological staging: T3Nx Mx.)

Discussion

Pheochromocytoma is a functioning tumour of adrenal medulla, derived from catecholamine producing chromaffin cells. It is known as the "ten percent tumour" as ten percent of tumours are inherited, ten percent are extra adrenal, ten percent are malignant, ten percent are bilateral and ten percent occurs in children. Essentials of diagnosis for Pheochromocytoma are frequently sustained hypertension, with or without paroxysms, episodic headache, excessive sweating, palpitation, visual blurring, postural tachycardia, hypotension, elevated urinary catecholamine or their metabolites, hyper metabolism and hyperglycemia (1-4). Our patient had paroxysmal hypertension, headache, sweating and dizziness which was never thoroughly evaluated. Cerebral infarction and intracerebral hemorrhage are well known, but are rare complications of Pheochromocytoma.

Pheochromocytoma as a secondary cause of hypertension must always be suspected. It is probably reasonable to work up a young hypertensive patient who does not respond to appropriate therapeutic trial or has paroxysmal episodes, rapidly progressive or resistant hypertension, severe hypertension, hypertension associated with systemic manifestations (sweating, pallor, etc.), as well as postural reactions (hypotension or tachycardia) (4-9).

Diagnosis is established by 24 hours urinary VMA levels. MRI is preferred for localization of tumour, because contrast media used in CT scan can provoke paroxysms. 123I MIBG (Meta-Iodo-Benzyl-Guanidine) scan will identify 90% of primary tumours and is essential for the detection of multiple extra adrenal tumours and metastasis. Complete surgical excision is the goal of treatment, but should only be carried out as an elective procedure.

RESEARCHERID
THOMSON REUTERS
Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 5 (Issue 1): January 2018 ISSN: 2394-9414
DOI- 10.5281/zenodo.1158603 Impact Factor- 4.174

Preoperatively it is vital to control blood pressure because anesthetic induction or minimal manipulation of adrenal or extra adrenal tumours during surgery may cause dangerous fluctuations in blood pressure. The agent of choice would be long acting alpha-adrenergic blocker, Phenoxy-benzamine. Beta blockade with propranolol is instituted 3-4 days prior to surgery. Beta blockade should not be used without prior alpha blockade, as unopposed vasoconstriction can lead to potentially catastrophic hypertension with adrenergic blockade and meticulous intraoperative blood pressure control. Hypertension persists in 10% to 35% of patients after successful resection of Pheochromocytoma. However, the hypertension is relatively easy to control requiring minimal medications. Patients should be followed up for persistent hypertension apart from the possibility of tumour recurrence even after resection of apparently benign tumours. Post-operative intensive care monitoring should be done as hypovolemia and hypoglycemia may occur (7-13).

Conclusion

Intracerebral hemorrhage is a serious and an unexpected complication of Pheochromocytoma that is very rarely seen in young adults. A balance between adequate preoperative medical blockade and expedition of surgery to minimize the risk of further perioperative stroke is needed. Early diagnosis, multi-disciplinary treatment approach is essential. Our case is an addition to the literature of the clinical and radiological features and treatment modalities in such rare cases.

Conflict of interest: The authors declare that there is no conflict of interests regarding the publication of this paper.

Acknowledgement

Dr. Vikas Gupta (Director & HOD in Neurosurgery and Interventional Neuro-Radiology/ Endovascular Neurosurgery), Dr. Vivek Yadav (Consultant Neuro and Spine surgery), Dr. Deep Goel (Sr. Consultant & Director in Minimal Access, Bariatric & Surgical Gastroenterology), Dr. Neeraj Bhalla (Director and Senior consultant-Cardiology)

References

1. Yeo H, Roman S. Pheochromocytoma and functional paraganglioma. Curr Opin Oncol 2005;17:13-18
2. Zuber SM, Kantorovich V, Pacak K (2011) Hypertension in 8. pheochromocytoma: characteristics and treatment. Endocrinol Metab Clin North Am 40: 295-311.
3. Garg MK, Kharb S, Brar KS, Gundgurthi A, Mittal R (2011) Medical management of pheochromocytoma: Role of the endocrinologist. Indian J Endocrinol Metab 15 Suppl 4: S329-336.
4. Rothmund M (2008) Adrenal glands and other endocrine disorders. In: Williams NS, Bulstrode CJK, O'Connell PR, editors. Bailey and Love's short practice of surgery. (25thedn) UK: Edward Arnold Ltd, 813-14.
5. Duh QY, Liu C, Tyrrell JB, Adrenals. In: Doherty GM, Current Diagnosis and treatment: surgery. (13thedn) USA: McGraw Hill companies, 737-50.
6. Meaney E, Alva F, Moguel R, Meaney A, Alva J, Webel R. Formula and nomogram for the sphygmomanometric calculation of the mean arterial pressure. Heart. 2000;84:64.
7. Alfakih K, Walters K, Jones T, Ridgway J, Hall AS, Sivananthan M. New gender-specific partition values for ECG criteria of left ventricular hy- pertrophy: recalibration against cardiac MRI. Hypertension. 2004;44: 175–179.
8. Gong Y, Hua Y, Keep RF, Hoff JT, Xi G. Intracerebral hemorrhage: effects of aging on brain edema and neurological deficits. Stroke. 2004; 35:2571–2575.
9. Lee JC, Cho GS, Choi BO, Kim HC, Kim YS, Kim WK. Intracerebral hemorrhage-induced brain injury is aggravated in senescence-accelerated prone mice. Stroke. 2002;33:1190–1195.
10. Kothari RU, Brott T, Broderick JP. The ABCs of measuring intracerebral hemorrhage volumes. Stroke. 1996;27:1304–1305.

Volume 5 (Issue 1): January 2018 ISSN: 2394-9414
DOI- 10.5281/zenodo.1158603 Impact Factor- 4.174

11. Meyer JS, Rogers RL, Mortel KF. Prospective analysis of long-term control of mild hypertension on cerebral blood flow. Stroke. 1985;16: 985–990
12. Kawai K, Kimura S, Miyamoto J, Sekiguchi T, Kano S, Ogawa Y, et al : A case of multiple extra-adrenal pheochromocytomas. Endocrinol Jpn 26 : 693-696, 1979
13. Lee JG, Choi JH, Lee SB, Lee IC, Bae SD : Clinical analysis of hypertensive intracranial hemorrhage. J Korean Neurosurg 22 : 484- 451, 1993