Unusual Presentation of Pheochromocytoma as CVA

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Abstract

Pheochromocytomas are catecholamine secreting enterochromaffin tumours causing paroxysmal hypertension. Our patient was a young boy who presented with cerebral haemorrhage. Sometimes diagnosing them can be difficult, due to overlapping with other conditions such as myocardial ischemia, baroreflex failure, migraine, panic disorder arrhythmias, hyperthyroidism, carcinoid tumours and stroke. This patient is of clinical interest as pheochromocytoma presented with life-threatening cerebro vascular attack.

Introduction

Pheochromocytomas are catecholamine secreting enterochromaffin tumours causing paroxysmal hypertension. The symptom triad is of headaches, diaphoresis and palpitations.

The cerebral manifestations of pheochromocytoma are uncommon [1]. Due to its variable clinical presentation, pheochromocytomas have been called "the masquerader". We describe a patient presenting with loss of consciousness in which the initial working diagnosis was cerebral haemorrhage due to hypertensive bleed. During the work-up, ultrasonography revealed an adrenal mass. MRI, urinary metanephrine and nor-metanephrine levels confirmed the diagnosis of pheochromocytoma.

Case Report

A 21-year old male presented to the emergency department in an unconscious state with a history of loss of consciousness for 2 hours. On evaluation, the patient had a Glasgow Coma Scale (GCS) score of 8; E2V2M4. The blood pressure was 200/140 mmHg on repeated recordings. Physical examination revealed plantar extension of the feet with exaggerated deep tendon reflexes and hypertonia. Non Contrast Computed Tomography (NCCT) of the head revealed a large cerebral haemorrhage in the left capsule-ganglionic region with intraventricular extension and mass effect (Figure 1).

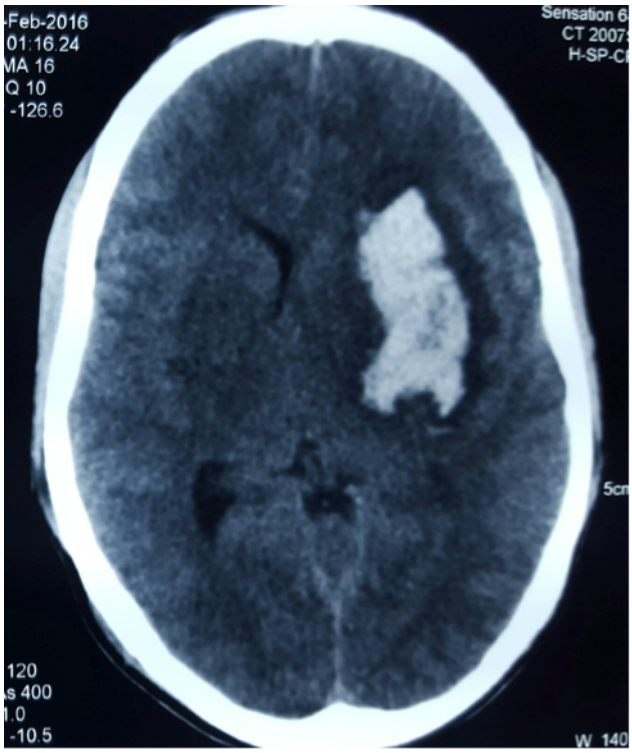

Figure 1. Large blood attenuation hyper density ~ 5 X 4.3 X 2.8 cms in left capsule- ganglionic region with intraventricular extension and mass effect.

The Patient was intubated and then managed on the lines of an Acute Cerebro vascular Attack (CVA). An ultrasound scan of the abdomen revealed a well-defined heterogeneous solid mass at the upper pole of right kidney, suspicious of an adrenal lesion. There was also left hydroureteronephrosis and a left renal calculus (10 mm in upper calyx). NCCT and MRI abdomen (figure 2) a showed soft tissue density lesion in the right adrenal, measuring 5.5 x 3.4 cms. After the initial management and confirmation of diagnosis, the patient was started on Prazosin (alpha blocker) 10 mg twice daily, telmisartan 40 mg and hydrochlorothiazide 12.5 mg twice daily. A week later, beta-blocker labetalol infusion was started along with Amlodipine 10 mg twice daily and clonidine

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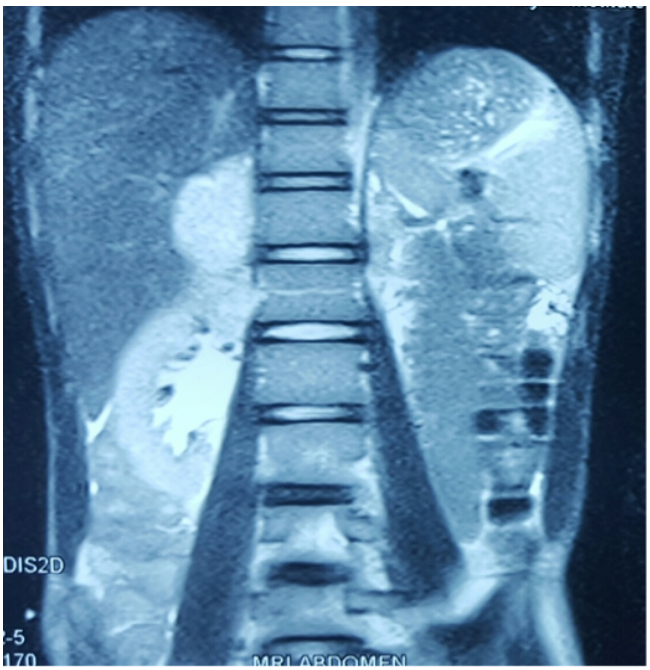

Figure 2. T2 weighted MRI delineating intensely enhancing well-defined right adrenal mass of 5.2 X 3.7 cms

0.2mg four times daily as the pressures were still uncontrolled. Urine metanephrine and nor-metanephrine levels were elevated to more than 10 times the normal limit, suggesting pheochromocytoma. After medical stabilization and adequate BP control, the patient underwent a right adrenalectomy.

The post-operative period was uneventful. He gradually regained consciousness and the GCS score was 15, with a residual right hemiparesis. The blood pressure gradually settled and was managed with oral anti-hypertensive Prazosin 5 mg once daily. The 24-hour urine metanephrine and nor-metanephrine levels were repeated after 2 weeks and 3 months post surgery, which were within normal limits. The patient is doing well at 8 months follow up and has right hemiparesis, which is gradually improving.

Discussion

The clinical variations of pheochromocytoma is well known and sometimes diagnosing them can be difficult, due to overlapping with other conditions such as myocardial ischemia, baroreflex failure, migraine, panic disorder arrhythmias, hyperthyroidism, carcinoid tumours and stroke [2,3]. The incidence of pheochromocytoma in the general population is 0.8 per 100 000. It is often not diagnosed accurately or managed by many internists, endocrinologist and nephrologists [4]. In a recent review Cook and Loriaux stated that hormone screening for adrenal tumours should be "tailor made" for the clinical context [5]. Our patient was a young man whose pheochromocytoma induced a cerebral haemorrhage, presenting as a stroke.

Cerebral haemorrhage secondary to pheochromocytoma is rare and the incidence is unknown. The condition is partially or completely reversible in some patients. First, hypertension alone can cause cerebral infarction in patients who have pheochromocytoma [6]. During hypertensive crises, the combination of cerebrovascular auto regulation failure and very high blood pressure can cause hypertensive encephalopathy [7]. Other possible mechanism may be endothelial dysfunction due to circulating toxins or chemotherapy agents. There may be cerebral infarction or haemorrhage due to compromise of the microcirculation by pressure from surrounding vasogenic edema [8].

Definitive management of a pheochromocytoma is surgical removal, which is curative in up to 90% cases. Our patient is of clinical interest as pheochromocytoma presented with a life-threatening cerebro vascular attack. This unusual case has been described to increase awareness of the life-threatening manifestations of pheochromocytoma. The clinician must have a high index of suspicion to diagnose and manage these cases.

References

1. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, et al. (1996) A reversible posterior leukoencephalopathy syndrome. *N Engl J Med* 334: 494-500. https://doi.org/10.1056/NEJM199602223340803
2. Manger WM (2006) Diagnosis and management of pheochromocytomas—recent advances and current concepts. *Kidney Int* 70 (suppl): S30–S35 https://doi.org/10.1038/sj.ki.5001974
3. 2 Mittendorf EA et al. (2007) Pheochromocytoma: advances in genetics, diagnosis, localization, and treatment. *Hematol Oncol Clin North Am* 21: 509–525 https://doi.org/10.1016/j.hoc.2007.04.012
4. Golden S, Robinson K, Saldanha I et al. Prevalence and incidence of endocrine and metabolic disorders in the United States: a comprehensive review. *J Clin Endocrinol Metab* 2009; 94: 1853–1879. https://doi.org/10.1210/jc.2008-2291
5. Cook DM, Loriaux DL. The incidental adrenal mass. *Am J Med* 1996;101:88-94 https://doi.org/10.1016/S0002-9343(96)00082-4
6. Lehmann FS, Weiss P, Ritz R, Harder F, Staub JJ. Revers- ible cerebral ischemia in patients with pheochromocytoma. *J Endocrinol Invest* 1999;22:212-4. https://doi.org/10.1007/BF03343544
7. Goswami R, Tandon N, Singh B, Kochupillai N. Adrenal tumour, congestive heart failure and hemiparesis in an 18- year-old male. A clinical-pathological conference. *Int J Car- diol* 1995;49:233-8. https://doi.org/10.1016/0167-5273(95)02302-D
8. McKinney AM, Sarikaya B, Gustafson C, Truwit CL (2012) Detection of microhemorrhage in posterior reversible encephalopathy syndrome using susceptibility-weighted imaging. *AJNR Am J Neuroradiol* 33: 896-903. https://doi.org/10.3174/ajnr.A2886

- **Key Points**
- Though there is the symptom triad of headaches, diaphoresis and palpitations for pheochromocytoma, rarely cerebral ischaemia and stroke symptoms can also be the presenting features in patients who have pheochromocytoma .
- Diagnosis of pheochromocytoma can be difficult at times, as the symptoms often overlap with other conditions such as hyperthyroidism, carcinoid tumors, myocardial ischaemia etc.
- Cerebral haemorrhage is partially or completely reversible in patients of pheochromocytoma.