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AN ATYPICAL MANIFESTATION OF PHEOCHROMOCYTOMA CRISIS: ACUTE DELIRIUM

Agarwal M¹, Kant R², Sharma D³, Madan S³

¹Junior Resident, ²Additional Professor, ³Senior Resident, Department of Internal Medicine and Radiodiagnosis, AIIMS, Rishikesh

Abstract

Pheochromocytoma is a rare neuroendocrine tumor presenting with a classical triad of symptoms. In 10% of the cases the presentation can be atypical, and the diagnosis is often missed for several years. Most common neurological manifestations are haemorrhage, seizures, etc., but pheochromocytoma crisis presenting as delirium has been infrequently reported. We present a case of a middle-aged female who presented with hypertensive emergency and acute confusional state. Other causes of delirium were ruled out in the patient and a timely diagnosis of pheochromocytoma was made with CT abdomen showed a large right suprarenal mass which was confirmed on 123I-MIBG scan and supported by elevated plasma metanephrines. She responded to alpha and beta blockers following which her blood pressure spikes were controlled and subsequently her sensorium improved significantly. Patient underwent surgical resection of the tumor and was followed up after 2 weeks with her blood pressure controlled without any medications.

Key Words: Pheochromocytoma, Delirium, Hypertension

Introduction

Pheochromocytoma is a rare catecholamine-secreting neuroendocrine tumour with an estimated annual incidence of 0.8 per 100,000 person-years¹ and is a rare cause of endocrine hypertension, representing < 0.2% of the total cases in the general population. Although pheochromocytomas may occur at any age but they are commonest in the fourth to fifth decade with equal male and female preponderance.²

Pheochromocytoma has a classic triad of symptoms consisting of episodic headache, diaphoresis, and

tachycardia³ but most do not present with this. It has atypical presentations in 9%–10% of cases. Even with classical presentations, the diagnosis is often missed for several years because the clinical manifestations are diverse and may mimic other disorders. Prompt diagnosis and treatment cures the condition but delay in diagnosis can result in serious cardiovascular and neurological complications. Most common neurological manifestations are haemorrhage, seizures, etc. However, pheochromocytoma crisis presenting as delirium has been reported in only a handful of cases. We present a case of a middle-aged female with hypertension who presented with hypertensive emergency and acute confusional state.

Case presentation

A 55-year-old female with history of hypertension and diabetes, presented with altered behavior in the form of agitation, refusal to food and acute confusional state for the last 2 days. There

Corresponding Author

Dr.Ravi kant, M.D. (Med. BHU) FICP, FICN, DFID (Diabetes),
Department of internal Medicine
AIIMS Rishikesh, Uttarakhand, India Mob.9870717859

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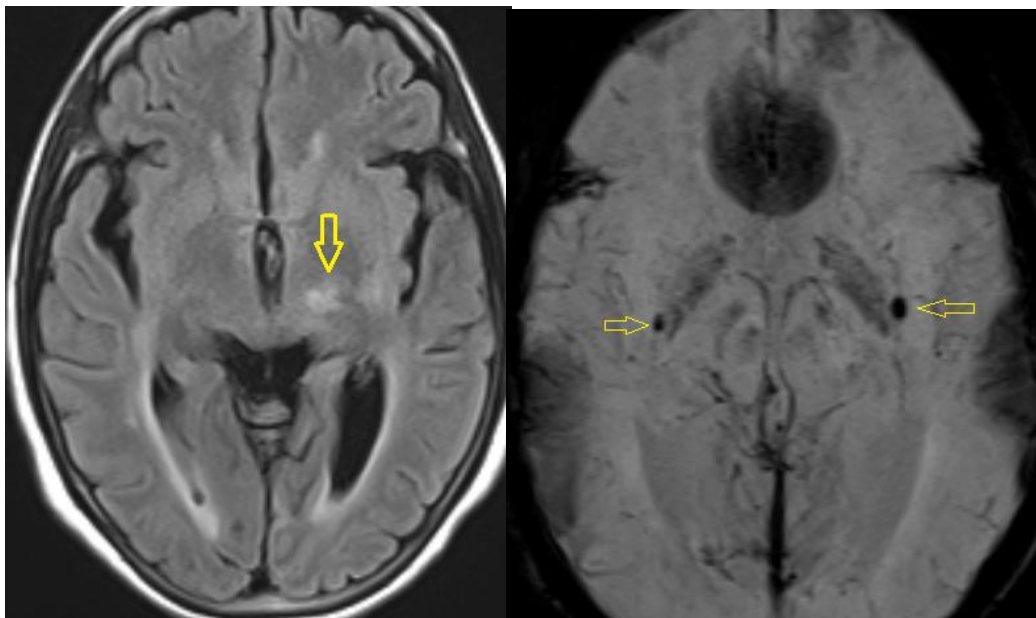
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was no history of fever, headache, seizures, or loss of consciousness in the past. No history of substance abuse. Patient's past medical history was significant for an episode of ischemic stroke 7 years ago followed by complete recovery. Patient was on regular oral hypoglycaemic agents and antihypertensive drugs. She has had 3 hospital visits in last 1 year for intractable headaches, however detailed evaluation was not done.

Upon presentation to the Emergency, the patient was conscious, but confused, and not following verbal commands. Her Glasgow Coma Scale (GCS) was 13/15 (E4V4M5), Pulse rate- 130/min, BP- 230/130 mm Hg and axillary temperature was 98.2 F. Examination showed bilaterally equal and reactive pupils and systemic examination was unremarkable. Fundus examination revealed non-proliferative diabetic retinopathy. Electrocardiogram showed findings of left ventricular hypertrophy. Rest of the blood investigations including workup for

endocrine hypertension like serum electrolytes, thyroid function tests and early morning fasting serum cortisol were within normal range.

A random blood glucose obtained by finger prick method was 67 mg/dl. Keeping a possibility of hypoglycaemic encephalopathy intravenous dextrose was administered. Despite correction of hypoglycaemia there was no improvement in sensorium of the patient. Considering a likelihood of hypertensive encephalopathy, we started her on intravenous labetalol along with oral angiotensin receptor blocker and later added a diuretic. We monitored the blood pressure closely with repeated readings showing episodic spikes in blood pressure with no significant decline. Contrast imaging (MRI) of the brain (Image 1) ruled out acute cerebrovascular accident and posterior reversible encephalopathy syndrome (PRES). Electroencephalography (EEG) was normal.



(A)

(B)

(Image 1) A- Axial T2W FLAIR image at the level of thalami shows small chronic infarct in left thalamus (arrow). B- Axial MIP susceptibility weighted MRI image shows foci of microbleed in bilateral basal ganglia (arrows).

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Ultrasound scan of the abdomen showed a large (around 11 x 9 cm) well-encapsulated right suprarenal heterogeneous lesion with vascularity. With a suspicion of pheochromocytoma we sent fasting plasma metanephrines and normetanephrines which were- 80.8 pg/ml and 1890 pg/ml respectively (reference range below 195 pg/ml). CECT abdomen showed a well-defined heterogeneously enhancing

right suprarenal mass (image 2) with absolute and relative washout values of 50 and 25 respectively suggestive of likely pheochromocytoma. Patient was started on alpha-blocker (prazosin) for optimal blood pressure control. Over the next 24 hours with the normalization of blood pressure spikes, her sensorium gradually improved, and she was comprehending normally.

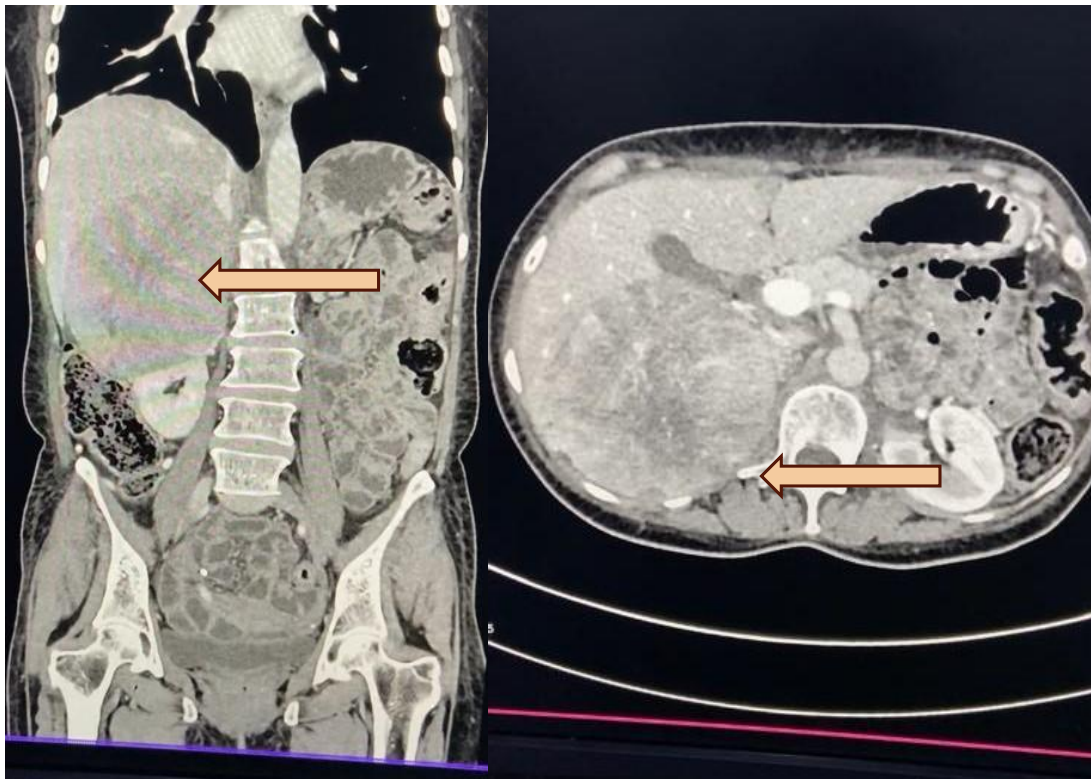


Image 2: CECT abdomen showing a well-defined hyper enhancing right suprarenal mass (arrow)

The patient was planned for surgical excision of the tumour. A preoperative metaiodobenzylguanidine (I-131 MIBG) scan was performed which confirmed the diagnosis of pheochromocytoma (Image 3). Preoperative optimization of blood pressure was done using alpha and beta blockade. The right adrenal mass was excised (image 4). During the intraoperative period, the patient had hypotension along with significant hypoglycaemia, requiring inotropes which were gradually tapered off over

the next 48 hours. She was discharged with normal blood pressure and no residual focal neurologic deficit. The patient was followed up after 2 weeks, her sugar levels and blood pressure controlled without any medications.

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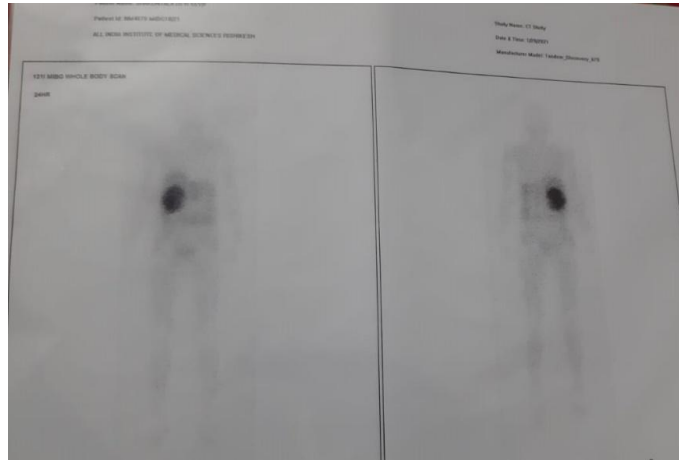


Image 3: I-131 scan showing MIBG avid soft tissue density mass lesion arising from upper pole of right kidney. Two images are of the same patient from front and back.



Image 4: a large well circumscribed mass 12x 10 cm right suprarenal mass

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Discussion

Pheochromocytoma can present with diverse clinical manifestations, hence also known as a 'Great Masquerader'. [4] Neurological symptoms including thunderclap headache, seizures, strokes have been reported during acute pheochromocytoma crisis. These symptoms are mainly due to cerebral ischemia caused due to catecholamine surge during the crisis. Pheochromocytoma crisis manifesting as acute delirium state has been very rarely reported in literature as most of the cases reported stroke as the main neurological complication, which was ruled out in our case.

In a study by Anderson et al, out of 28 patients who presented with pheochromocytoma crisis, 39% were associated with neurological symptoms. Severe headache was the most common neurological symptom (10 patients). Two patients sustained an ischaemic stroke. Other complications were seizures (five patients), intracerebral haemorrhage and cortical blindness. Only 3 patients had a clinical picture of delirium. In all the cases blood pressure was significantly elevated during the acute crisis (range 170/90–290/130 mm Hg) which was consistent with our clinical finding. [5]

Pheochromocytoma is provisionally diagnosed by elevated 24-h urine catecholamine metabolites levels. Radiologically, a CT scan with adrenal protocol is carried out to locate the adrenal tumour. If it is undetected by conventional imaging or if metastasis must be ruled out, a secondary imaging modality is used, which includes I-MIBG scan as done in our patient.

Surgical excision remains the treatment of choice. [6] If the patient presents in crisis, surgery must be deferred until the patient is hemodynamically stable which is achieved by adequate alpha and beta blockade. Our patient was operated 1 week after the presentation and was rendered normotensive thereafter.

It is important for clinicians to diagnose pheochromocytomas at an early stage to prevent catastrophic complications. Not always do patients present with the classic triad of symptoms and the presentations can be atypical. Sometimes the presentation is only uncontrolled hypertension

and fluctuating blood sugar levels with episodic worsening of symptoms attributing to excess catecholamines during the crisis. With advancement in diagnostics, many atypical presentations of pheochromocytoma are being reported in literature, however the key is to extract detailed history with prudent investigations to rule out other causes of delirium and approach the case with a clinical suspicion of a pheochromocytoma in such settings. Our patient responded both hemodynamically and clinically with appropriate alpha and beta blockade. With the definitive treatment being surgical, meticulous preoperative assessment is important as intraoperative crisis can be fatal and life threatening.

Conclusions:

Rare presentation of Pheochromocytoma crisis can be acute delirium, we have to carefully evaluate it according to the clinical conditions.

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