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ADRENAL PHEOCHROMOCYTOMA PRESENTING WITH SYNCOPE IN AN ELDERLY PATIENT: A CASE REPORT

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Contribution

NP concieved the idea and designed the case report. SAMB collected picture and wrote the report . Final review was made by NHB. All authors contributed to submitted case report.

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ABSTRACT

Pheochromocytoma is a rare catecholamine-producing neuroendocrine tumor who often presented with palpitation, headache, hypertension, acute abdominal pain and chest pain but we report a case of pheochromocytoma presented with syncope.

Key Words: Pheochromocytoma, Adrenal, syncope, Elderly

INTRODUCTION

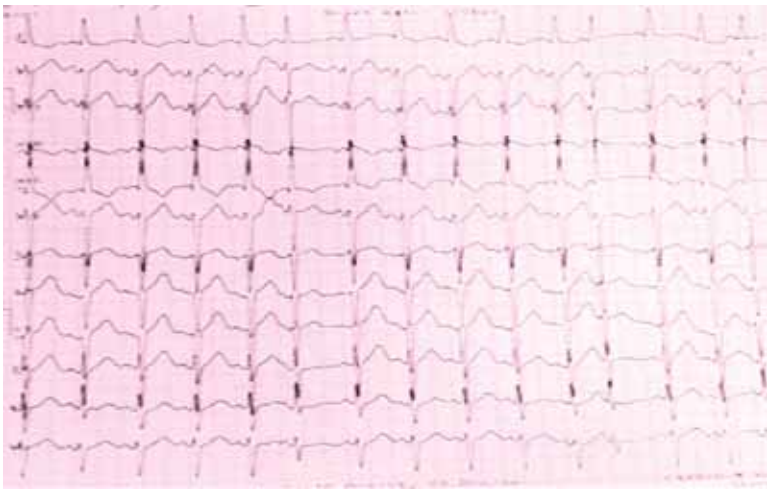
Pheochromocytoma is a rare catecholamine-producing neuroendocrine tumor that arise from chromaffin cells of the adrenal medulla or extra-adrenal para ganglia with an estimated incidence of less than 0.1% in the general population.[1,2] Spontaneous and massive necrosis within the tumor is associated with acute and severe clinical presentation of massive catecholamine release, including acute abdominal pain , chest pain, or even shock.[1]

It has been found that the clinical presentation of pheochromocytoma is usually due to the direct activities of catecholamines and is highly variable.[1] Interestingly, there were also typical clinical manifestations includes episodic attacks of headache, palpitation, sweating and hypertension.[3] Hypertension is often paroxysmal, and usually presented with sudden attacks of alternating hypertension and hypotension. Although typical clinical presentations of pheochromocytoma is palpitation, headache and hypertension, a patient with pheochromocytoma presenting as recurrent syncope due to hypotension have been reported in this study. This patient's hypotension was mainly postural and extremely severe. Syncope due to hypotension is unusual in this disease and only a few cases have been reported.

CASE REPORT

The case reported here was 87 y/o man with history of anterior STEMI 3 months ago, after that suffered from resistant hypertension (un controlled hypertension despite used of 5 anti hypertensive classes of drugs), palpitation , hypokalemia and episodic abdominal pain. The case was admitted to hospital due to sever transient fluctuation of blood pressure (systolic and diastolic blood pressure between (260 – 60) and (140 – 40) mm Hg, respectively) and recurrent syncope in sitting position. In his thorough assessment of clinical, we observed the orthostatic hypotension during syncopal events. The evidence of in electrocardiogram of the case had been clearly seen sinus tachycardia despite use of high dose beta blocker (Figure 1).

Figure 1: Patient's Electrocardiogram



Laboratory tests: There have been few Para clinical investigations into the case, metanephrine and normetanephrine (urine 24 hrs) was 1189 [<350] and 2347 [< 600] micg /24 hr, respectively. And free cortisol was 147.9 micg / 24 hr [50 – 190]. An analysis of Abdominopelvic sonography showed Cystic-solid mass with diameter of 62*60*52 mm in left adrenal gland.

Also, Abdominopelvic CT scan without contrast showed mass lesion with heterogeneous density and well defined rim without calcification in left adrenal gland with approximate diameter of 70*55*66 mm (Figure 2,3). It is also noting that liver, spleen, pancreas and kidneys was normal in size and density.

Figure 2: CT Scan of Left Adrenal Mass

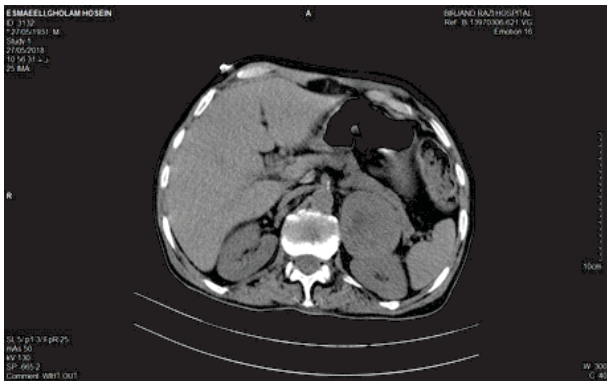
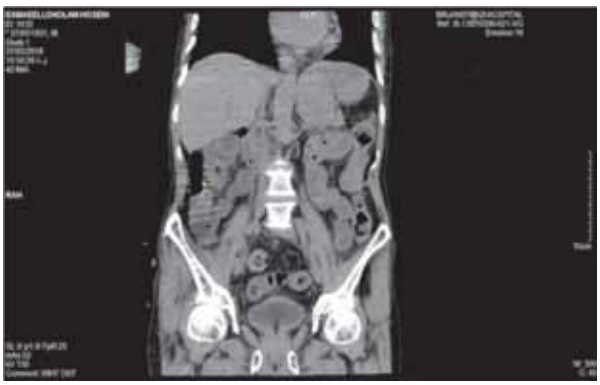


Figure 3: CT Scan of Left Adrenal Mass



Spiral lung and mediastinal CT scan without contrast showed mediastinal mass with heterogeneous density and diameter of 20*21*37 mm in subcarina. In addition, calcified hilar lymph node was seen (adenopathy). It was not possible to investigate the Iodin 131 scan for evaluations of concurrent paraganglioma because it was do not perform in our center.

According to laboratory tests, left adrenal mass, mediastinal mass, fluctuation in blood pressure and orthostatic hypotension in this patient, pheochromocytoma was recognized and treated with phenoxybenzamin (10 mg TDS). After control of orthostatic hypotension and fluctuation in blood pressure, surgical consultation was done to remove laparoscopic lumps of adrenal mass. It seems high risk and not possible that these results are due to old age, recent myocardial infarction, surgical procedures and lack of permission by patient fellows. Then the patient after control of symptoms discharged.

DISCUSSION

Characteristic clinical presentation of pheochromocytoma include paroxysmal headache, diaphoresis, palpitation, anxiety and hypertension.[1] Symptoms related to pheochromocytoma also include nausea, vomiting, chest and abdominal pain.[2] Noticeable point relates to this issue is that signs and symptoms are often paroxysmal because of episodic secretion of catecholamines by pheochromocytoma.[2]

As was pointed out in the paper, recurrent and large amount of catecholamine secretin from infarct zone of tumor in to the circulation accelerated the progression of infarction and cause repeated attacks of intermittent hypotension and hypertension with headache, palpitation and chest pain and leading to massive necrosis with in the pheochromocytoma.[1] Patients with massive necrosis of adrenal pheochromocytoma often presented with abdominal pain.[4] By implementation the necrosis process within the tumor plasma levels of catecholamines turn into normal condition and is created remission in symptoms. Episodic abdominal pain in the patient due to tumor necrosis and episodic secretion of catechol amines was justifiable. It is noteworthy that β adrenal receptors stimulated with catecholamine excess cause sinus tachycardia or even serious ventricular tachycardia.

In recent years, Li and colleagues reported cases with pheochromocytoma and acute myocardial infarction.[4] More than half of patients with pheochromocytoma crises presenting AMI and ACS had few significant coronary artery disease. As respects increased catecholamine levels lead to hemodynamic compromise of the myocardium, accelerated cell fibrosis and death. And therefore, In patients with pheochromocytoma crises presenting AMI with significant coronary artery disease, catecholamine excess may worsen the myocardial infarction. The mechanism of myocardial infarction and dysfuction in pheochromocytoma patients is due to coronary artery vasospasm or direct toxic effects of catecholamines. Catecholamines cause increases in left ventricular work load with stimulation of ventricular hypertrophy due to hypertension and increased in heart rate. Changes in coronary arteries include thickness of media that compromise blood supply of myocardium cause coronary artery vasospasm. Anti-ischemic therapy with B blocker before a adrenoreceptore blockade in this patients can be harmful.[5]

It is interesting to note that the onset of symptoms after acute myocardial infarction has also occurred in the case of this study. One of the issues emerging from this finding relate specifically to primary excess in catecholamine levels due to adrenal tumor which causes acute myocardial infarction.

The evidence of the case presented thus far supports the idea that sinus tachycardia related with excess in catecholamine levels. In our patient high dose metoprolol administered after myocardial infarction because of persistent sinus tachycardia, and after that exacerbate the fluctuation in blood pressure, which can be due to blockade of B adrenoreceptores before blockade of a andernoreceptores. Researchers list a lot of reasons why hypotension is due to an excess of catecholamine. These are hypovolemia, alternating secretion of catechol amines, proportion of epinephrine to norepinephrine in the secretion, insufficiency in peripheral response to catecholamines, adrenocortical insufficiency and baroreflex impairment. pheochromocytoma causes depletion in circulatory blood volume Periodic secretion of catechol amines cause hypotension , because persistent hypercatecholaminemia can desensitization of blood vessels to catechol amines and sudden drop in norepinephrine levels causes reduction in vascular tone.[6,7] In this patient, the mentioned mechanisms do not ascend because hypotension in the case of this study is orthostatic.

A classification study of this diseases by Ma reports that 8% - 12.5% of pheochromocytomasare classed as malignant and approximately 15% - 26% of that are metastatic. The larger size of the tumor (>6 cm) , the further excretion of dopamine, and the greater local invasion of tumor are arguments in favor of the malignancy.[8] In our case report, tumor size in abdominal CT more than 6 mm, which can be suggestive of malignancy. As well as subcarinal mass in spiral lung CT could suggest metastasis lesion from the primary tumor.

Preoperative diagnosis is often based on clinical presentation and measurement of catecholamines and their metabolites in blood and urine. As this case clearly demonstrates signs, symptoms and laboratory finding in favor of pheochromocytoma. The most important basis in the treatment of malignant pheochromocytoma is surgically resection of primary tumor and to prevent recurrence and metastasis and also treat hypertensive symptoms by catecholamine blockade agents.[8] Also, according to the current guidelines nonselective alpha blockers such as phenoxybenzamine is a gold standard for preparation of pheochromocytoma patients before surgery. Unfortunately in our patient feasibility of biopsy and confirm diagnosis and certain cure there was not available because of his old age, clinical condition and dissatisfaction of his family members for surgery. This patient finally treated with phenoxybenzamine, volume expansion and discharged after partial control of symptoms. alternating hypertension and orthostatic hypotension and syncope.

CONCLUSION

The purpose of the current study was to introduce the case report for pheochromocytomawith rare manifestation. Malignant pheochromocytoma is very rare and aggressive neoplasms in the elderly, and there are often many other comorbidities worsening the survival like our patient.Typical clinical manifestation of

pheochromocytoma is headache, diaphoresis, palpitation, anxiety and hypertension, but one of the most significant current study is the topical clinical presentation in our patientthere was labile blood pressure and syncopal episodes due to orthostatic hypotension that can be draw attention to the rare clinical presentations of pheochromocytoma. This case reveals the need for further clinical andparaclinicalinvestigation in patients with pheochromocytoma.

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