

Undiagnosed pheochromocytoma in a patient with acute appendicitis: a challenging surgical emergency condition

Akut Apandisitli bir hastada tanımlanmamış feokromositoma: Zor acil cerrahi bir durum

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Abstract

Pheochromocytoma is a rare catecholamine secreting neuroendocrine tumor originating mostly from adrenal glands. A clinical challenge arises when a patient with a previously undiagnosed and untreated pheochromocytoma presents with a surgical emergency. A 24-years-old male presented to our emergency department with abdominal pain. Physical examination, laboratory tests and radiological findings were consistent with acute appendicitis. Prior surgery, his blood pressure raised to 240/130 mmHg, while heart rate was 125/min. Vital signs were stabilized by intravenous nitrates and beta blockers. Appendectomy was done without further complications. After surgery, high level of urine vanillyl mandelic acid and right adrenal mass in abdominal MRI that support the diagnosis of pheochromocytoma were found. Elective right adrenalectomy was made two months later and pathology revealed a pheochromocytoma. The postoperative period was uneventful and he has been followed for 18 months without any antihypertensive treatment.

Key words: Pheochromocytoma, surgery, emergency

Özet

Feokromositoma, çoğunlukla adrenal bezden kaynaklanan katekolamin salgılayan nadir bir nöroendokrin tümördür. Daha önceden bilinmeyen veya tedavi edilmemiş feokromositoma olguları acil cerrahi durumlarda klinik zorluklara neden olmaktadır. Karın ağrısı nedeniyle acil servise başvuran 24 yaşındaki erkek hastanın yapılan fizik muayenesinde, laboratuvar ve radyolojik tetkikleri akut apandisit ile uyumluydu. Acil cerrahi planlanan hastanın kan basıncı 240/130 mmHg, nabız sayısı 125/dakika idi. İntravenöz nitrat ve beta-blokörlerle vital bulguları stabilize edilen hastaya apendektomi yapıldı. Cerrahi sonrası, hipertansiyon etiyojisi araştırılan hastanın idrarda vanil mandolik asit düzeyi yüksek ve üst batın MR'da sağ adrenalde kitle saptandı. Apendektomiden 2 ay sonra, elektif olarak sağ adrenalectomi yapıldı. Postoperatif patolojisi feokromositoma olarak saptanan hasta 18 aydır sorunsuz olarak takip edilmektedir.

Anahtar sözcükler: Feokromositoma, cerrahi, acil

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Başvuru tarihi | Submitted on: 18.08.2013

Kabul tarihi | Accepted on: 25.11.2013

Introduction

Pheochromocytoma is a rare catecholamine secreting neuroendocrine tumor originating mostly from adrenal glands. Secretion of excessive amount of catecholamines such as noradrenaline, adrenaline and dopamine cause unbalanced sympathetic stimulation causing symptoms. Typical symptoms include episodes of hypertension, headache, palpitations and diaphoresis. It can also cause hypertensive emergency precipitating in acute stress requiring intense antihypertensive therapy¹. Undiagnosed pheochromocytoma may be problematic in some cases with failure of recognition of symptoms and delayed diagnosis can present with perioperative dilemma. Here, we present a patient without a prior history of hypertension who presented with acute appendicitis and experienced preoperative hypertensive crisis, successfully treated with intravenous antihypertensives.

Case report

A 24-year-old male presented to our emergency department with an acute onset of abdominal pain. He said that his pain is located periumbilically and started several hours ago with accompanying nausea and vomiting. He denied any medical conditions and medications.

Upon arrival, he was suffering from an acute abdominal pain. His vital signs were within normal limit. Physical examination revealed prominent tenderness with rebound in the right lower quadrant consistent with acute appendicitis. Cardiovascular and pulmonary examinations were normal. Blood chemistry results were within normal range while hemogram showed leukocytosis, 16.600/mm³ and neutrophily, 13.600/mm³. Posteroanterior upright thorax and plain X-ray of ab-

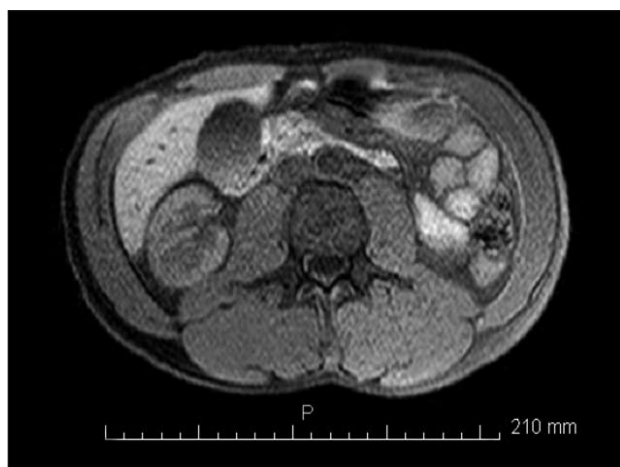


Figure 1. T1- weighted magnetic resonance of abdomen showing right adrenal adenoma

domen appeared normal. Acute appendicitis was suspected and abdominal ultrasound showed noncompressible appendix. Patient was consulted with general surgery department and appendectomy was planned. Before surgery, patient experienced palpitations and his blood pressure raised to 240/130 mm Hg, while heart rate was 125/min. Vital signs were stabilized to 140/90 mm Hg and 88/min with intravenous nitrates and beta blockers. Appendectomy was performed without any complications and patient was discharged after four days of hospitalization. In order to explain the malignant hypertensive situation in this young man, laboratory tests and radiological studies were performed. 24h urine collection disclosed vanillyl mandelic acid of 23 mg/24 H (3-9 mg/24 H) and abdominal MRI showed a right adrenal mass measuring 44x30 mm consistent with adrenal adenoma (**Figure 1**). Right adrenalectomy was made electively two months later and pathology revealed a pheochromocytoma. The postoperative period was uneventful and he has been followed for 6 months without any antihypertensive treatment.

Discussion

Pheochromocytoma is a rare but potentially lethal disease. The incidence of pheochromocytoma is about one in 1000-2000 patients and one in 50-1000 patients screened for hypertension². Pheochromocytoma may go unrecognized and up to a half may be found only at autopsy². There is a distinct group of patients with pheochromocytomas who remain normotensive despite active metabolites being secreted³. Sometimes high blood pressure may return to within the normal range because of myocardial damage. However, some patients with pheochromocytomas may present as surgical emergencies with hemorrhage and infarction of the tumor⁴.

Incidental presentation of pheochromocytoma represents usually a dramatic event, being a therapeutic challenge with a very difficult control of blood pressure and often carrying a tragic outcome. Absence of previous history of hypertensive crises or concomitant symptoms does not exclude pheochromocytoma in perioperative period. Perioperative diagnosis is difficult as hypertension and other symptoms can be linked to anxiety. A significant complication is being directly related to preoperative increase in systolic blood pressure³. Noxious stimuli such as venous catheterization, tracheal intubation, skin incision, anaesthetic drugs and palpation of the tumor or abdominal exploration will start the hypertension crisis by releasing catecholamine of the tumor. There are a few reports of intraoperative presentation of pheochromocytoma in the literature^{5,6}. Normally, elective surgery is a safe procedure with an operative mortality of 1,3% by the help of preoperative

management⁷. In the pre-operative period, pre-operative treatment with α -adrenergic blockers, such as phenoxybenzamine reduces adverse events in patients undergoing adrenalectomy. But mortality could be close to 80% when a patient diagnosed at the time of incidental surgery⁸. The hypertensive crisis should be immediately controlled. A α - and β -adrenergic blockers should be considered. It is essential that hypertension is controlled with a rapidly acting α -adrenergic blocker before instituting any β -adrenergic receptor blockade. Suppression of β -adrenoceptor-mediated cardiac sympathetic in the absence of adequate arteriolar dilatation may precipitate acute pulmonary edema⁹. We describe a young patient without any symptoms or history of pheochromocytoma experiencing hypertensive crisis before appendectomy. We believe that during anaesthesia induction, presence of acute stress have triggered rapid discharge of catecholamines and cause symptoms. We treated the hypertensive attack with intravenous nitrates and β -blockers. The stabilization of the patient take about 8 hours before the operation. As in our experience, a relatively simple operation such as appendectomy in a pheochromocytoma patient may be challenging where severe hypertension may jeopardize the life of the patient and the risk of the surgery overcome is greater than the nonoperative management of appendicitis¹⁰.

As a conclusion, severe hypertension observed in patients in acute stress like acute appendicitis must alert physicians for pheochromocytoma. Intense antihypertensive therapy is needed for stabilizing patients perioperatively.

References

1. Prejbisz A, Lenders JW, Eisenhofer G, Januszewicz A. Cardiovascular manifestations of pheochromocytoma. *J Hypertens* 2011;29(11):2049-2060.
2. Hume DM. Pheochromocytoma in the adult and in the child. *Am J Surg* 1960;99:458-496.
3. Plouin PF, Duclos JM, Soppelsa F, Boubllil G, Chatelier G. Factors associated with perioperative morbidity and mortality in patients with pheochromocytoma: analysis of 165 operations at a single center. *J Clin Endocrinol Metab* 2001;86:1480-1486.
4. Spencer E, Pycock C, Lytle J. Pheochromocytoma presenting as acute circulatory collapse and abdominal pain. *Intensive Care Med* 1993;19:356-357.
5. Sellevold OFM, Reder J, Stenseth R. Undiagnosed pheochromocytoma in the perioperative period. Case reports. *Acta Anaesthesiol Scand* 1985;29(5):474-479.
6. Voros DC, Smyrniotis B, Argyra E, Vadalouka A, Sifaka L, Papadimitriou J. Undiagnosed pheochromocytomas in the perioperative period. *Eur J Surg* 1996;162(12):985-987.
7. Sheps SG, Jiang NS, Klee GG, van Heerden JA. Recent development in the diagnosis and treatment of pheochromocytoma. *Mayo Clin Proc* 1990;65:88-95.
8. Stenstrom G, Haljamae H, Tisell LE. Influence of pre-operative treatment with phenoxybenzamine on the incidence of adverse cardiovascular reactions during anaesthesia and surgery for pheochromocytoma. *Acta Anaesthesiol Scand* 1985;29:797-803.
9. Prys-Roberts C. Pheochromocytoma recent progress in its management. *Br J Anaesth* 2000;85:44-57.
10. Tarant NS, Dacanay RG, Mecklenburg BW, Birmingham SD, Lujan E, Green R. Acute appendicitis in a patient with undiagnosed pheochromocytoma. *Anesth Analg* 2006;102(2):642-643.