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Cardiac presentations mimicking acute coronary syndrome of a giant pheochromocytoma case

Akut koroner sendromu taklit eden dev feokromositoma olgusu

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ABSTRACT

Pheochromocytomas are benign tumors which originate from adrenal chromaffin cells. The classical triad of the pheochromocytoma includes headache, palpitations, and diaphoresis. Cardiac complications as arrhythmias, cardiomyopathy, ST elevated myocardial infarction, non-ST elevated myocardial infarction and myocardial infarction with non-obstructive coronary arteries (MINOCA) may be seen. Herein, we present a case of a 28-year-old male patient who admitted to an emergency department with chest pain and tachycardia one year ago. Coronary angiography was performed to diagnose a possible acute coronary syndrome. Urgent cardiac catheterization did not demonstrate an obstructive cardiac artery. Any intervention for revascularization was not needed. Nevertheless, the symptoms of the patient continued in the past year after this admission and he was admitted to the hospital a few times more with similar cardiac symptoms mimicking acute coronary syndrome. When the patient referred to our department, we determined that his plasma and urinary catecholamine levels were elevated. Magnetic resonance imaging (MRI) demonstrated a 167x70 mm sized heterogeneous mass including cystic components in the right adrenal gland, which pushes the right kidney towards inferior. After the pre-medication, a 170 mm in size 990-g weighted mass was successfully removed with open surgery. Histopathological findings confirmed the pheochromocytoma diagnosis. However, we presented a case of an exceptional giant pheochromocytoma mimicking acute coronary syndrome.

Keywords: Pheochromocytoma, acute coronary syndrome, giant adrenal mass

ÖZ

Feokromositomalar, adrenal kromaffin hücrelerinden kaynaklanan genellikle benign tümörlerdir. Feokromositomanın klasik triadı baş ağrısı, çarpıntı ve terlemeyi içerir. Aritmiler, kardiyomiyopati, ST segment elevasyonlu miyokard enfarktüsü, ST segment elevasyonu olmayan miyokard enfarktüsü ve obstrüktif olmayan koroner arterlerle miyokard enfarktüsü (MINOCA) gibi kardiyak komplikasyonlar görülebilir. Bir yıl önce göğüs ağrısı ve çarpıntı nedeni ile acil servise başvuran 28 yaşında erkek hastaya akut koroner sendrom ön tanısı ile koroner anjiyografi yapılmış. Koroner anjiyografide obstrüktif hastalık saptanmamış. Benzer şikayetlerinin devam etmesi üzerine tarafımıza refere edilen hastanın yapılan tetkiklerinde katekolamin düzeyleri yüksek saptandı. Manyetik rezonans görüntülemede sağ adrenal glandda 167x70 mm boyutlarında kistik heterojen kitle mevcuttu. Preoperatif medikasyon sonrasında 170 mm boyutunda ve 990 g ağırlığındaki kitle laparotomi ile başarılı bir şekilde çıkarıldı. Histopatolojik bulgular feokromositoma tanısını doğruladı. Bu bildiride genç bir hastada akut koroner sendromu taklit eden dev feokromositoma vakası sunulmaktadır.

Anahtar Kelimeler: Feokromositoma, akut koroner sendrom, dev adrenal kitle

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INTRODUCTION

Pheochromocytoma is a rare catecholamine secreting tumor which originates from adrenal chromaffin cells and affects 0.1-0.6% of hypertensive patients (1). The classical triad of the pheochromocytoma includes headache, palpitations, and diaphoresis. Cardiac complications as arrhythmias, cardiomyopathy, ST elevated myocardial infarction, non-ST elevated myocardial infarction and myocardial infarction with non-obstructive coronary arteries (MINOCA) may be seen (2). The average tumor size is approximately 4.9 cm (3). Giant pheochromocytomas are seen rarely and generally present silent, unlike the classic symptoms of catecholamine-releasing tumors. In many cases, they are incidentally discovered. The algorithm to diagnose a pheochromocytoma consists of the biochemical evaluation and imaging of a possible retroperitoneal mass (4). Herein, we presented a male patient with a giant pheochromocytoma, who manifests cardiac symptoms mimicking acute coronary syndrome before the diagnosis.

The abstract of this case report was presented as "poster presentation" in Endobridge 2019, 24-27 October 2019, Antalya, Turkey.

CLINICAL CASE

A 28-year old male admitted with uncontrolled hypertension, palpitations, and irritability with one year of symptom history. One year ago, he was admitted to the emergency department with chest pain and palpitation. Electrocardiogram demonstrated ST depressions in anterior derivations. Troponin I level was normal. Urgent cardiac catheterization showed slow coronary flow in the left anterior descending artery. An intervention for revascularization was not needed. Nevertheless, the symptoms of the patient continued for one more year after the first attack and he was admitted to the hospital a few times more with similar cardiac symptoms mimicking acute coronary syndrome. When the patient referred to our department, his blood pressure was 140/90 mmHg. Heart rate was 120 beats/minute and electrocardiogram showed sinus rhythm. The physical examination was normal. The patient was evaluated for a possible catecholamine-secreting tumor. We determined an elevation on 24 hours urinary catecholamine metabolites; metanephrine, 5587 µg/24 hours (50-250), normetanephrine, 9233 µg/24 hours (100-500), and dopamine, 728 µg/24 hours (65-400). Urinary adrenaline and noradrenaline levels were normal. Magnetic resonance imaging demonstrated a 167x70 mm sized heterogeneous mass including cystic components in the right adrenal gland, which pushes the right kidney to inferior (**Figure 1**). MIBG scan showed increased uptake in the right adrenal localized mass.

Four mg/day doxazosin treatment was initiated before surgery. 40 mg/day propranolol was added to achieve a much better tachycardia control. After ensuring the efficiency and tolerability of the therapy, the patient underwent to right adrenalectomy by open surgery. A 990 g weighted mass excised totally with its capsule (**Figure 2**). The histopathological evaluation confirmed pheochromocytoma. Capsule invasion was not seen and the Ki67 index was <5. The patient had an uneventful recovery period. Catecholamine levels measured 2 weeks after surgery decreased to normal. We continue to follow the patient as asymptomatic, normotensive, and with normal catecholamine levels.

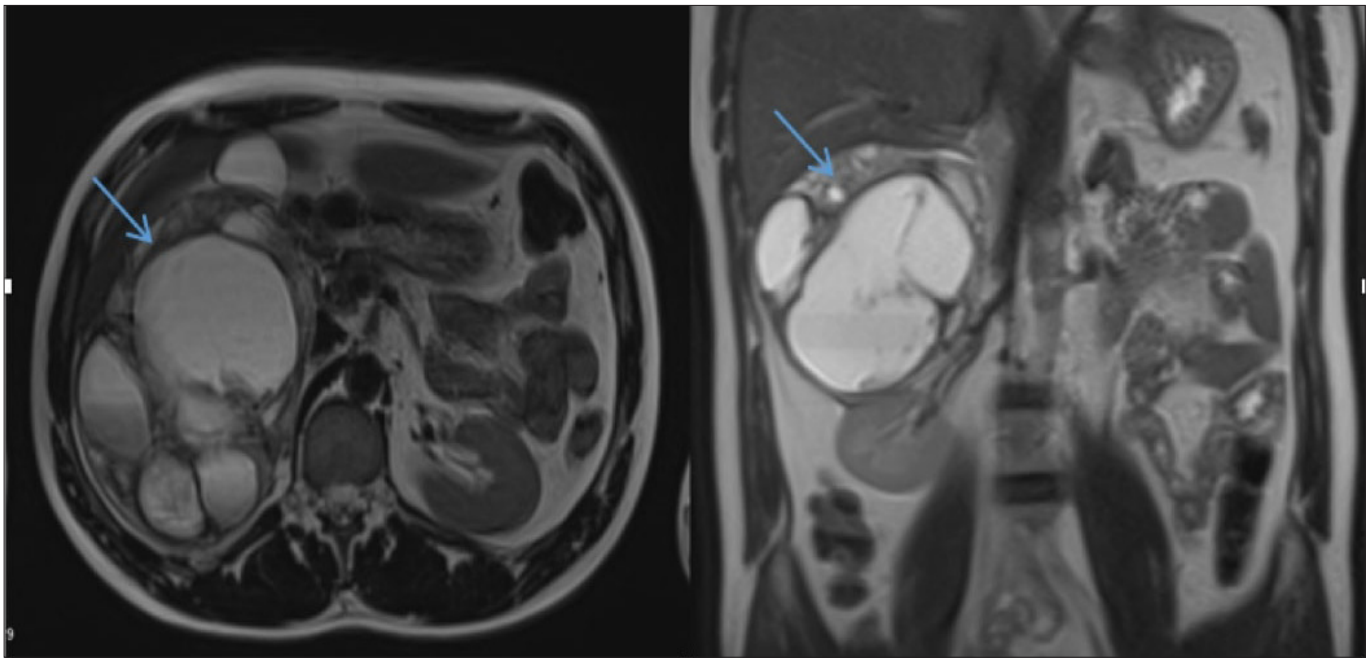

Figure 1. The heterogeneous mass 167x70 mm in sized including cystic components in the right adrenal gland

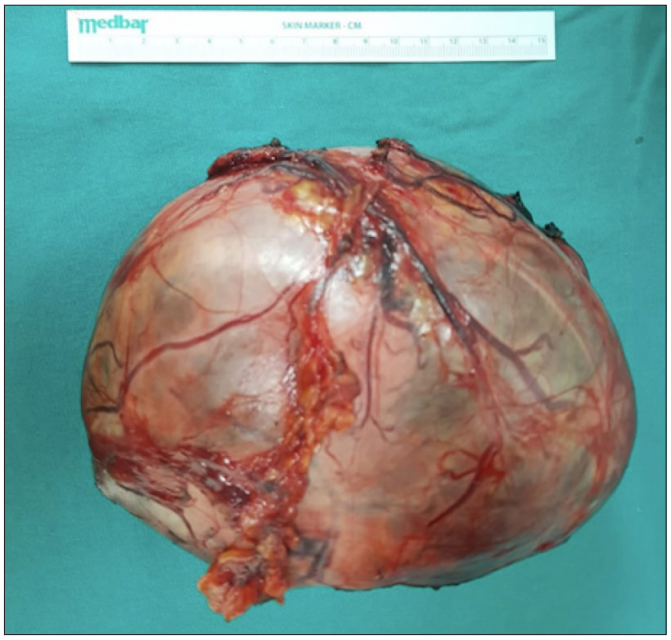

Figure 2. The macroscopic apperance of 17 cm-sized excised mass

DISCUSSION

Pheochromocytomas have rarely seen tumors originating from adrenal chromaffin cells, The estimated pheochromocytoma incidence is 0.8 per 100.000 people/year (5). Although most of them are benign tumors, 10 % of the cases can be malign (6). They appear in 4.-5. decades and the men-women ratio are seen equal (7). While most of these tumors being sporadic, they can also be seen as a component of hereditary syndromes such as Von Hippel-Lindau, neurofibromatosis type 1, MEN 2 (8). Even though classic symptoms are palpitations, sweating, headache, with the ever-growing usage of the imaging methods, they can be incidentally determined (9). Symptoms include tremor, dyspnea, pallor, generalized weakness, and panic attack-type symptoms as well as orthostatic hypotension, visual blurring, papilledema, weight loss, polyuria, polydipsia, constipation (10). Pheochromocytoma symptoms appear based on the excessive secretion of epinephrine, norepinephrine, and dopamine. The increased central symptomatic activity of the affected patients also contributes to this situation. They also cause laboratory abnormalities such as increased erythrocyte sedimentation rate, insulin resistance, hyperglycemia, leukocytosis, psychiatric disorders, and rarely, secondary erythrocytosis due to overproduction of erythropoietin (11). Circulating high catecholamine metabolites can lead to various cardiac or metabolic problems. While electrocardiogram is usually seen normal, there can also be some pathological findings as ST elevation, T inversions, abnormal Q waves (12). Diagnosing of the pheochromocytoma is usually based on the measurements of the urinary and plasma fractionated metanephrines and catecholamines levels and is followed by the localization of the tumor with abdominal and pelvic MRI or CT (13). If tumors are in size over 10 cm, MIBG or FDG-PET may perform to see whether there are extra-adrenal, multifocal or with any metastatic illnesses (14). Although 15% of pheochromocytomas are extra-adrenal, 95% of them can be abdominal or pelvic. As the size becomes larger, the possibility of malignancy increases. For tumors over 6 cm, the ratio of the benign-malign tumor is 1:8 (15). Despite the secretion of intensive vasoactive metabolites, they can also be asymptomatic and can be detected during an autopsy (16). Extensive necrosis of the adrenal gland, decreasing the production of catecholamines, and the retention of these hormones within the capsular mass after secretion may be the explanations of why these patients are asymptomatic. These factors may lead to delayed diagnosis and late detected tumors can be seen in larger sizes (4). Following the pheochromocytoma diagnosis, all patients should do the appropriate and necessary medical preparations for the resection. In a study evaluating 312 cases of pheochromocytoma, the average tumor size was found 4.5±2.9 (17). Another study analyzing 20 pheochromocytoma cases larger than 10 cm shows that the average age of the patients was 49. In this case series, while 8 patients were asymptomatic, 4 cases presented hypertension, and only 1 case presented chest pain (6). In a recent study of 34 pheochromocytoma cases over 10 cm, the average age was 49. While 31% of the cases presented asymptomatic, 21% had hypertension and back pain, %17 had hypertension and abdominal pain (4). In the report of Korgali et al. (18) a 63-year-old male presented similar symptoms mimicking acute coronary syndrome. Coronary angiography did not show obstructive coronary artery and further examination revealed a 20x17x9 cm pheochromocytoma. According to Uysal et al. (19) 37-year-old male presented hypertension and palpable mass in left upper quadrant. Further examination revelated a 18x8x13 cm malign pheochromocytoma with liver metastasis. Soufi at al. (20) report, a 17-year female patient presented with a 21x15 cm malign pheochromocytoma with liver metastasis while having no symptoms. Our case was 28-year-old and with 167x70 mm sized tumor. Cases under the age of 30 with benign masses in these sizes are seldom in literature. This is also the youngest giant pheochromocytoma case in Turkey among the reported cases with similar sizes. Although most of the cases over 10 cm are asymptomatic, our case was admitted to emergency service with chest pain, palpitation, hypertension symptoms which mimic acute coronary syndrome. In pheochromocytoma patients, electrocardiogram abnormalities appear due to excessively secreted catecholamine levels which stimulate myocardium (21). Exogenous epinephrine and norepinephrine are also cardiotoxic based on the dosage

(22). Pheochromocytoma had a relatively high incidence of cardiovascular complications. These complications include cardiac arrhythmia, ST and non-ST elevation MI, heart failure, hypertensive urgency, TIA, stroke, and subarachnoid hemorrhage. Rarely MINOCA may be seen (2).

Troponin levels may increase mildly while electrocardiogram has seen normal in many of the cases. However, clinical presentation and electrocardiogram findings can mimic acute coronary syndrome. In cases in which pheochromocytoma mimics acute coronary syndrome, coronary angiography does not show obstructive coronary artery. In our case, the patient was admitted to emergency service with typical chest pain and anterior ST depressions were observed in the electrocardiogram. Even though the patient was young, coronary angiography was performed to exclude a possible acute coronary syndrome, but any pathological finding was not revealed in angiography.

CONCLUSION

When physicians confront patient, who have uncontrolled hypertension or unexplained heart diseases, pheochromocytoma diagnosis should be kept in mind especially in younger patients.

ETHICAL DECLARATIONS

Informed Consent: Written informed consent was obtained from all participants who participated in this study.

Status of Peer-review: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

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Author Contributions: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

REFERENCES

1. Widimský J. Recent advances in the diagnosis and treatment of pheochromocytoma. Kidney Blood Press Res 2006; 29: 321–6.
2. Melson E, Amir S, Shepherd L, Kauser S, Freestone B, Kempegowda P. Myocardial Infarction with non-obstructed coronaries - atypical presentation of pheochromocytoma. Endocrinol Diabetes Metab Case Rep 2019; 2019.
3. Guerrero MA, Schreinemakers JMJ, Vriens MR, et al. Clinical spectrum of pheochromocytoma. J Am Coll Surg 2009; 209: 727–32.
4. Maharaj R, Parbhu S, Ramcharan W, et al. Giant cystic pheochromocytoma with low risk of malignancy: a case report and literature review. Case Rep Oncol Med 2017; 2017: 4638608.
5. Beard CM, Sheps SG, Kurland LT, Carney JA, Lie JT. Occurrence of pheochromocytoma in Rochester, Minnesota, 1950 through 1979. Mayo Clin Proc 1983; 58: 802–4.
6. Ambati D, Jana K, Domes T. Largest pheochromocytoma reported in Canada: A case study and literature review. Can Urol Assoc J 2014; 8: E374-7.
7. Reisch N, Peczkowska M, Januszewicz A, Neumann HPH. Pheochromocytoma: presentation, diagnosis and treatment. J Hypertens 2006; 24: 2331–9.
8. Goldstein RE, O'Neill JA, Holcomb GW, et al. Clinical experience over 48 years with pheochromocytoma. Ann Surg 1999; 229: 755–64.
9. Yu R, Nissen NN, Chopra P, Dhall D, Phillips E, Wei M. Diagnosis and treatment of pheochromocytoma in an academic hospital from 1997 to 2007. Am J Med 2009; 122: 85–95.
10. Bravo EL, Gifford RW. Pheochromocytoma. Endocrinol Metab Clin North Am 1993; 22: 329–41.
11. Bravo EL. Pheochromocytoma: new concepts and future trends. Kidney Int 1991; 40: 544–56.
12. Komamura K, Fukui M, Iwasaku T, Hirotani S, Masuyama T. Takotsubo cardiomyopathy: Pathophysiology, diagnosis and treatment. World J Cardiol 2014; 6: 602–9.
13. Lenders JWM, Duh Q-Y, Eisenhofer G, et al. Pheochromocytoma and paraganglioma: an endocrine society clinical practice guideline. J Clin Endocrinol Metab 2014; 99: 1915–42.
14. Eisenhofer G, Rivers G, Rosas AL, Quezado Z, Manger WM, Pacak K. Adverse drug reactions in patients with phaeochromocytoma: incidence, prevention and management. Drug Saf 2007; 30: 1031–62.
15. Alexandraki KI, Grossman AB. Adrenal incidentalomas: "the rule of four." Clin Med (Lond) 2008; 8: 201–4.
16. Arnaldi G, Boscaro M. Adrenal incidentaloma. Best Pract Res Clin Endocrinol Metab 2012; 26: 405–19.
17. Ichijo T, Ueshiba H, Nawata H, Yanase T. A nationwide survey of adrenal incidentalomas in Japan: the first report of clinical and epidemiological features. Endocr J 2020; 67: 141-52. doi: 10.1507/endocrj.EJ18-0486
18. Korgali E, Dundar G, Gokce G, et al. Giant malignant pheochromocytoma with palpable rib metastases. Case Rep Urol 2014; 2014: 354687.
19. Uysal E, Kırdak T, Gürer AO, İkidağ MA. Giant multicystic malignant pheochromocytoma. Turk J Surg 2015; 33: 296–8.
20. Soufi M, Lahlou MK, Benamr S, et al. Giant malignant cystic pheochromocytoma: a case report. Indian J Surg 2012; 74: 504–6.
21. Agarwal V, Kant G, Hans N, Messerli FH. Takotsubo-like cardiomyopathy in pheochromocytoma. Int J Cardiol 2011; 153: 241–8.
22. Paulin FL, Klein GJ, Gula LJ, Skanes AC, Yee R, Krahn AD. QT prolongation and monomorphic VT caused by pheochromocytoma. J Cardiovasc Electrophysiol 2009; 20: 931–4.