Case Report
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Could Retroperitoneal Ganglioneuroma be a Dopamine Secreting Ganglioneuroma?

Retroperitoneal Ganglionöroma Dopamin Salgılayan Bir Ganglionöroma Olabilir mi?

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Abstract

Ganglioneuromas are rarely occurring benign tumors characterized by hyperplasia of mature ganglia and satellite cells. They are well-differentiated, slow growing, and autonomous nervous system neoplasms, which are usually asymptomatic and do not release any hormones.

A male patient aged 26 years was evaluated for secondary hypertension six months ago. Ultrasonography of the abdomen revealed a mass lesion around the right kidney. An analysis of the 24-hour urine sample of the patient revealed the following parameters: 5-HİAA=3.9 mg/day (2-7), metanephrine=56.3 µg/day (52-341), and normetanephrine=146.1 µg/day (88-444). The computed tomography scan of the abdomen showed a retroperitoneal mass of 10 cm in size, containing minute calcified foci in the right retroperitoneal region. The mass was excised through general surgery and was classified as ganglioneuroma. The blood pressure of the patient returned to normal level after surgery, and he needed no further antihypertensive treatment. Besides, the metanephrine and normetanephrine levels in the 24-hour urine were also observed to be normal as in the preoperative period.

Retroperitoneal masses can actually be ganglioneuromas and an accurate diagnosis can be achieved only through postoperative histopathological evaluation. After the operation, blood pressure of the patient returned to normal. This suggests that retroperitoneal ganglioneuroma could possibly secrete dopamine, epinephrine, or norepinephrine.

Keywords: Retroperitoneal ganglioneuroma; hypertension; catecholamine

Özet

Ganglionöromalar; matür ganglion ve satellit hücrelerin hiperplazisi ile karakterize, nadir görülen iyi huylu tümörlerdir. İyi diferansiye, yavaş büyüyen ve genellikle asemptomatik, hormon üretmeyen otonom sinir sistemi neoplazmlarıdır.

Yirmi altı yaşındaki erkek hasta, altı ay önce sekonder hipertansiyonu olması nedeni ile değerlendirildi. Batın ultrasonografisinde sağ böbrek yakınlarında kitle lezyonu belirlendi. Hastanın 24 saatlik idrar örneğinde: 5-HİAA=3,9 mg/gün (2-7), metanefrin=56,3 µg/gün (52-341) ve normetanefrin=146,1 µg/gün (88-444) saptandı. Batın bilgisayarlı tomografide, sağ retroperitoneal alanda kalsifiye odak içeren 10 cm büyüklüğünde retroperitoneal kitle izlendi. Genel cerrahi tarafından kitle eksize edildi ve ganglionöroma tanısı aldı. Cerrahiden sonra hastanın tansiyonu normale geldi ve antihipertansif tedaviye ihtiyacı kalmadı. Yirmi dört saatlik idrar metanefrin ve normetanefrin düzeyleri de operasyon öncesi dönemde olduğu gibi normal saptandı.

Retroperitoneal kitleler gerçekten ganglionöroma olabilmekte ve doğru tanı ancak postoperatif histopatolojik değerlendirme ile ortaya çıkabilmektedir. Operasyondan sonra hastanın kan basıncı normale dönmüştür. Bu durum; retroperitoneal ganglionöromanın dopamin, epinefrin veya norepinefrin salgılayabileceğini düşündürmektedir.

Anahtar kelimeler: Retroperitoneal ganglionöroma; hipertansiyon; katekolamin

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Introduction

Ganglioneuromas are rarely occurring benign tumors characterized by hyperplasia of mature ganglia and satellite cells. They are well-differentiated, slow growing, and autonomous nervous system neoplasms, which are usually asymptomatic and do not release any hormones (1). In the majority of the cases, they are detected incidentally. Histopathological examination is mandatory for differentiation of ganglioneuromas from malignant neuroendocrine tumors such as pheochromocytoma and neuroblastoma. Ganglioneuromas may develop on skull base, neck, posterior mediastinum, retroperitoneum, and adrenal gland through the sympathetic chain (2). They are particularly prevalent in the posterior mediastinum and retroperitoneal region (3). Sixty percent of the cases occur before the age of 20 years. The treatment of these cases is the complete excision of the tumor (2). Our aim in this study was to present a patient with the rarely occurring ganglioneuroma in the retroperitoneal region.

Case Report

A 26-year-old male patient was evaluated for symptoms of secondary hypertension six months ago. Doppler ultrasonography (USG) of the renal artery was done to determine the etiology of early onset of hypertension, and stenosis was not detected. USG revealed a mass lesion around the right kidney. A 24-hour urine sample of the patient was subjected to analysis and the following parameters were recorded: 5-hydroxyindole acetic acid (5-HIAA)= 3.9 mg/day (2-7), metanephrine= 56.3 µg/day (52-341), and normetanephrine= 146.1 µg/day (88-444). The positron emission tomography-computed tomography (PET-CT) revealed a mass lesion in the right adrenal region with high retention of fluorine-18 (F-18) fluorodeoxyglucose (FDG). The 131I/123I-Metaiodobenzylguanidine (MIBG) scintigraphy, carried out to check for pheochromocytoma, showed no retention. In the abdominal CT examination, a retroperitoneal mass of approximately 10 cm in size, containing min calcified foci, was observed in the right retroperitoneal region (Figure 1). The mass was excised through general surgery and diagnosed as ganglioneuroma (Figure 2). The histological examination showed spindle-shaped Schwann cells arranged in small fascicles, clusters of ganglial cells, and fibrous tissue. The ganglial cells had abundant cytoplasm, large eccentric nuclei, and one or more prominent nucleoli. There was no evidence of neuroblastoma. During the period after surgery, the patient's blood pressure returned to normal levels and he needed no antihypertensive treatment. In addition, the metanephrine and normetanephrine levels in the 24-hour urine were observed to be normal as levels in the preoperative period.

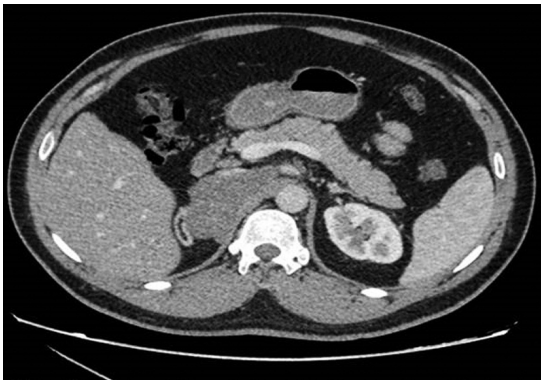

Figure 1: Abdominal CT arterial phase.

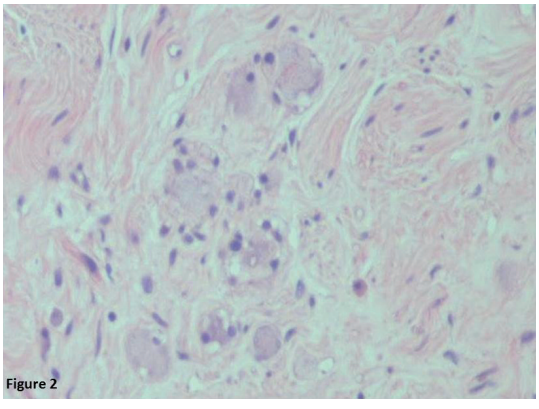

Figure 2: Ganglioneuroma: Histopathological examination (HE, x20).

Discussion

Ganglioneuroma originates from neural crest cells comprising sympathetic ganglion and adrenal gland cells. Ganglial cells are composite structures containing fibrous tissue and Schwann cells. Ganglioneuroma is a benign tumor observed three times more frequently in women than in men. Furthermore, it is more prevalent in people under the age of 20. Besides, it is more commonly detected in the mediastinum and retroperitoneal regions. In our case, the patient was a 26-year old male, and

the tumor was located in the retroperitoneum. Ganglioneuromas are usually asymptomatic and release no hormones. Because of a slow growth rate, it is usually diagnosed in the late adolescent period. The diagnosis can be made by monitoring the pressure exerted by the tumor on the surrounding tissues and through the mass effect. If the tumor grows retroperitoneally, abdominal pain and distention could be the main symptoms (4). In our case, abdominal pain was absent although the mass was enormous. The tumors generally do not release any hormones, but when they do, they release vasoactive intestinal peptide, androgen hormone, or catecholamine, which cause hypertension, sweating, diarrhea, virilism, and hypokalemia (3). Although hypertension was historically present in our case, the 24-hour urine levels of metanephrine and normetanephrine were normal. Besides, the potassium levels were normal. Interestingly, the blood pressure of the patient was normal after surgical removal of the ganglioneuroma, which suggested that it could possibly secrete dopamine. However, our efforts at measuring the levels were unsuccessful. Erem et al. (5) had reported a dopamine secreting adrenal ganglioneuroma in a patient. In addition, Yoshida et al. (6) reported a patient diagnosed with retroperitoneal ganglioneuroma who had a history of hypertension. Ganglioneuromas are radiologically well-located solid masses with contoured lobules and may involve partial calcification in some cases (7). Radiological monitoring methods such as USG, CT, and magnetic resonance imaging (MRI) are helpful in evaluating the size and composition of the mass and its relation to adjacent tissues. Previous studies have reported that preoperative diagnosis of ganglioneuroma is usually difficult and the diagnosis is made through histopathological examination (2). In our case, the diagnosis was also made through histopathological examination of the completely excised tumor. Ganglioneuroma is classified as a neurogenic tumor. The histopathological diagnosis for neurogenic tumors relies on the presence of mature ganglion cells. Unlike neuroblastoma, immature cells, atypical mitotic structure, or necrosis are absent in ganglioneuroma (5). Ganglioneuromas can be cured by excision and no relapse has been reported (8). In contrast, metastases of matured neuroblastomas are encountered in the lymph node adjacent to the tumor mass or in the regions away from the tumor. In our case, there was no evidence of metastasis before and after the operation. In conclusion, preoperative diagnosis of retroperitoneal ganglioneuromas is difficult, as it might be radiologically similar to the other tumors.

Conclusion

It is important to remember that retroperitoneal masses can actually be ganglioneuromas, and an accurate diagnosis can be achieved only through postoperative histopathological examination. In case of suspected abnormal clinical parameters such as hypertension, all the catecholamines should be measured before the operation to prevent a possible hypertensive crisis.

An inform consent form was obtained from the patients.

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Author Contributions

Concept: Fusun Saygılı; Design: Nilufer Ozdemir Kutbay; Data Collection or Processing: Yesim Ertan; Analysis or Interpretation: Baris Eker; Literature Search: Banu Sarer Yurekli; Writing: Hatice Ozisik.

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