



PARAGANGLIOMA OF THE ORGAN OF ZUCKERKANDL : REPORT OF A CASE

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Abstract

Tumors of the paraganglionic system will arise; these are termed paragangliomas, as they may be both physiologically and histopathologically similar to pheochromocytomas of the adrenal gland. Extra-adrenal paragangliomas make up approximately ten per cent of all paragangliomas, with those occurring in the organ of Zuckerkandl being the most common

We describe a case of paraganglioma in a 51-year-old woman with hypertension being present for 32 years. She suffered from abdominal mass, nausea, dizziness, emesis, headache, excessive sweating, palpitations, and back pain. An abdominal mass was diagnosed as a functional extra-abdominal paraganglioma by diagnostic imaging and biochemical tests. The mass was totally excised and her symptoms healed after the operation.

Symptoms of tumors of the organ of Zuckerkandl are secondary to synthesis and release of excess catecholamines. This is a rare case of functional paraganglioma arising from the organs of Zuckerkandl with successful surgical removal and amelioration of hypertension.

1. BACKGROUND

Paraganglia of the sympathoadrenal neuroendocrine system are distributed along the paravertebral and para-aortic axis from the base of the skull to the pelvic floor, and the largest compact collection of paraganglia in this system is the adrenal medulla. Tumors of the paraganglionic system will arise; these are termed paragangliomas, as they may be both physiologically and histopathologically similar to pheochromocytomas of the adrenal gland. Extra-adrenal paragangliomas make up approximately ten per cent of all paragangliomas, with those occurring in the organ of Zuckerkandl being the most common [1].

Keywords: Organ of Zuckerkandl, Paraganglioma

The incidence of functioning paragangliomas secreting catecholamines is estimated to occur in 0.001-0.01% of the general population and is believed to be responsible for less than 1% of adult hypertension which can be corrected surgically. Malignant versus non-malignant tumors are difficult to distinguish histologically and it seems that the only reliable indicators of malignancy are invasion of adjacent organs and distant metastases in areas where paraganglionic tissue does not occur normally [2]. In 10% of cases, extra-adrenal paragangliomas are inherited as part of the multiple endocrine neoplasia type 2 syndrome, von Hippel-Lindau disease, and rarely, neurofibromatosis type 1 [3]. These tumors have also been reported in multiple endocrine neoplasia type 1 patients [4].

2. CASE PRESENTATION

A 51-year-old woman with a history of paroxysmal hypertension and abdominal mass was admitted to Osmangazi University Hospital. She suffered from nausea, dizziness, emesis, headache, excessive sweating, palpitations, and back pain. Physical examination was relatively unremarkable except for palpable abdominal mass at the right of the midline, bilaterally minimal arteriolar narrowing on funduscopic examination. She has had hypertension since the age of 19. Her presenting blood pressure was 190/110 mmHg, blood urea nitrogen (52 mg/dl) and creatinin (2.1 mg/dl) levels were elevated.

She was admitted for investigation. Her blood pressure ranged from 250/120 mm Hg to 150/90 mm Hg and was elevated dramatically by abdominal compression. Abdominal ultrasonography and computerized axial tomography revealed a well-defined, right para-aortic mass, located on the right psoas major muscle that measured 90×70×50 mm in the greatest dimension (Figure 1). To confirm a diagnosis of extra-adrenal paraganglioma, a 24-hour urine collection revealed a vanilmandelic acid concentration of 21 mg (normal 0.7 to 6.8 mg), metanephrine concentration of 2790 mg (normal 0 to 135 mg), and catecholamine concentration of 300 µg (normal 0 to 1.50 µg). Plasma norepinephrine level was 4.1 ng/ml (normal 0.07 to 0.319 ng/ml). Plasma epinephrine level was within normal limits.

The patient was treated for several days preoperatively with phenoxybenzamine, which she tolerated well. Abdominal exploration was performed. A lobulated tumor that measured 90×70×50 mm was found anterolaterally to the distal abdominal aorta, superior to right psoas major muscle, and the right ureter passed behind it. The capsule was adherent to the distal abdominal aorta. The hydronephrosis of the right kidney and dilatation of the proximal part of the right ureter were also shown. The adrenals and all other organs in the abdomen were completely normal. The blood pressure decreased to 100/60 mm Hg when the tumor was removed and it remained stable. Several hypertensive spikes during the mobilization were managed with intravenous phentolamine.

Her postoperative course was uneventful. Histology confirmed the diagnosis of typical extra-adrenal paraganglioma with no capsular invasion. Macroscopic examination of the mass revealed it to be well capsulated and in regular shape. It

measured 90x70x50 mm. The tumor was elastic-soft and gray-white in color at the cut surface. Microscopic examination showed that the tumor cells were uniformly medium-sized, but several showed slight pleomorphism. Some of the tumor cells had granular eosinophilic cytoplasm and a lot of them had hyaline granules in the cytoplasm. There was no evidence of necrosis and hemorrhage in the tumoral tissue. For immunohistochemical analysis, keratin, vimentin, synaptophysin, chromogranin were used. The tumor cells demonstrated distinctly positive reaction for synaptophysin and chromogranin. Stainings for keratin and vimentin were negative (Figure 2).

3. DISCUSSION

Approximately 0.001-0.01% of adults harbor paragangliomas, which constitute a curable form of hypertension in 1% of hypertensive patients [2]. Of patients with paragangliomas discovered only at autopsy, 75% died suddenly from either myocardial infarction or a cerebrovascular catastrophe. Moreover, one third of the sudden deaths occurred during or immediately after minor operations which were unrelated paragangliomas [5]. Approximately 10% of paragangliomas arise in extraadrenal locations, may occur in any part of the body, including in the carotid body, in the heart, along the aorta, in the urinary bladder [6], and 50% of these are localized anterolaterally to the distal abdominal aorta between the origin of the inferior mesenteric artery and the aortic bifurcation, in the organ of Zuckerkandl [1]. In this case a paraganglioma arising from the organs of Zuckerkandl is presented. Symptoms of tumors of the organ of Zuckerkandl are secondary to synthesis and release of excess catecholamines which may be precipitated with change of posture, exertion, trauma, anesthesia, emotional stress, and sexual intercourse. Only 25-60% of extra-adrenal paragangliomas are functional, with clinical symptoms usually occurring in males aged 30-45 years who presented with paroxysmal hypertension or sustained hypertension, headache, excessive sweating, palpitations, abdominal pain, and anxiety attacks [6]. The symptoms manifested by the woman in the present case of nausea, dizziness, emesis, and back pain are typical of catecholamine storm, and she had paroxysmal hypertension, headache, excessive sweating, palpitations, and abdominal pain.

It has been reported that 10% of paragangliomas are malignant, 10% are bilateral or multiple. The incidence of malignancy in extra-adrenal paragangliomas (20-40%) is greater than that for adrenal paragangliomas (2-10%). Malignant versus benign tumors also are difficult to distinguish histologically as the only reliable indicators of malignancy are invasion of adjacent organs and distant metastases in areas where paraganglionic tissue is not found normally, that are the liver, lung, lymph nodes [2]. In our case the adrenals and all other organs in the abdomen were completely normal. There was no sign of invasion of adjacent organs and malignant spread. The clinical diagnosis of extra-adrenal paragangliomas is based on measuring excessive excretion of catecholamines and their metabolites in the urine or circulating blood [6]. Elevated levels of norepinephrine more commonly occur with extra-adrenal

paragangliomas [7]. Our patient had elevated urinary levels of catecholamines, vanilmandelic acid, or metanephrine, and elevated plasma levels of norepinephrine.

The localization of the paraganglioma is of paramount importance to accomplish the surgical removal. Angiography, computerized tomography [8], ¹³¹Iodine-labelled metaiodobenzylguanidine scintigraphy, magnetic resonance imaging [9] have been used for the diagnosis of extra-adrenal paragangliomas. Angiography is invasive, technically difficult, and may cause a hypertensive crisis. A less invasive technique is computerized tomography which greatly aid in accurate diagnosis with the associated risk of allergic reactions and potentially life-threatening hypertensive crisis [8]. Magnetic resonance imaging scanning provide confirmation that the visualized tumor is functioning and can aid in the localization of extra-adrenal or metastatic tumors [9]. Magnetic resonance imaging may be more specific because of its ability to distinguish various types of adrenal tumors on the basis of imaging characteristics, but computerized tomography scan has several advantages over magnetic resonance imaging. Computerized tomography scan is less expensive and more available than magnetic resonance imaging [10]. In our case abdominal ultrasonography and a computerized tomography was performed to localize a paraganglioma. The original computerized tomography, which scanned at only the adrenal bed, was interpreted as normal. A repeat scan evaluating the pelvis revealed a 9 cm tumor just proximal to the bifurcation of the aorta.

4.CONCLUSION

We have reported a case of paraganglioma arising from the organs of Zuckerkandl. We believe that this is a rare case of functional paraganglioma arising from the organs of Zuckerkandl with successful surgical removal and amelioration of hypertension.

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ZUCKERKANDL ORGANI PARAGANGLİOMASI: VAKA TAKDİMİ

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Retroperitoneal paragangliomalar; aorta çevresinde sempatik zincirle birlikte dağılmış olan özelleşmiş nöral krest hücrelerinden kaynaklanmaktadır. Bu makalede 32 yıllık hipertansiyon hikayesi olan 52 yaşında bir bayan hasta paraganglioma vakası olarak takdim edilmektedir. Hastada karında kitle, bulantı, kusma, baş ağrısı, baş dönmesi, aşırı terleme, çarpıntı ve sırt ağrısı gibi şikayetler mevcuttu. Yapılan radyolojik ve biyokimyasal incelemeler, karındaki kitlenin aktif fonksiyon gösteren ekstraabdominal bir paraganglioma olduğunu gösterdi. Kitlenin tamamı eksize edildi ve ameliyat sonrası hastanın semptomları normale döndü.

Anahtar Kelimeler: Paraganglioma, Zuckerkandl Organı

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Figures

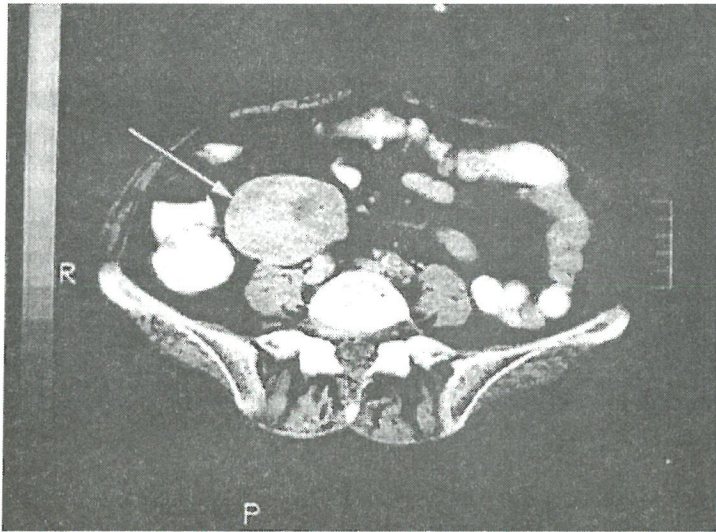


Figure 1
Computed tomography scan showing a 90x70 mm soft tissue mass just proximal to the bifurcation of the aorta.

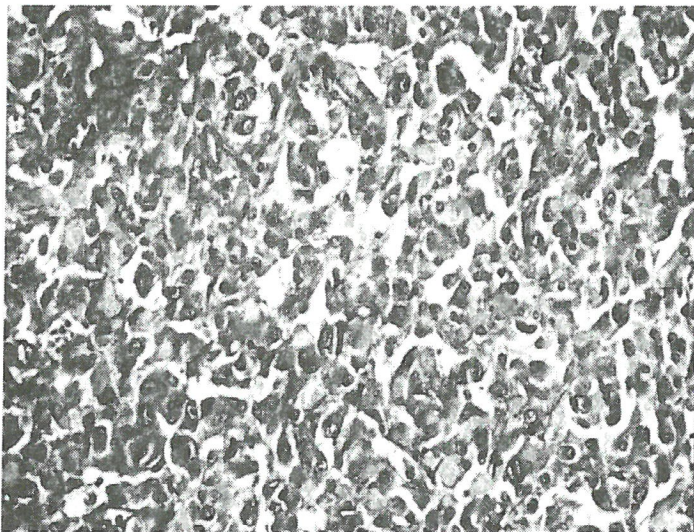


Figure 2
The tumor cells had pleomorphic nuclei and granular eosinophilic cytoplasm and some of them had hyaline granules in the cytoplasm (Hematoxylin and Eosin, $\times 200$)

