Benign Pelvic Retroperitoneal Paraganglioma: A Very Rare Localization: Case Report

Benign Pelvik Retroperitoneal Paraganglioma: Çok Nadir Bir Yerleşim

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ABSTRACT Paragangliomas are rare neoplasms originating from the paraganglial tissues composed of extra-adrenal chromaffin cells and 10% are localized to the retroperitoneal area. These tumors may be functional or non-functional due to the catecholamine secretion. Approximately 1/3 of retroperitoneal paragangliomas are malignant. The primary treatment of these tumors is complete surgical resection. Although there are risks of metastasis and recurrence for malignant paragangliomas; the metastasis, malignant transformation and recurrence potentials for benign paragangliomas are not known yet. Therefore, benign paragangliomas should be followed-up for long time as malignant paragangliomas. Herein, we present a 75 year-old patient with a benign, abscessed retroperitoneal paraganglioma in the retroperitoneal area of vesico-uterine space mimicking adnexal mass.

Key Words: Extra-adrenal paraganglioma; retroperitoneal space; pelvic neoplasms; abscess

ÖZET Paragangliomalar, ekstraadrenal kromaffin hücrelerin oluşturduğu paraganglial dokudan gelişen nadir tümörlerdir ve %10'u retroperitoneal alana lokalizedir. Katekolamin salınımına bağlı fonksiyonel veya non-fonksiyonel olabilirler. Retroperitoneal paragangliomaların yaklaşık 1/3'i maligndir. Bu tümörlerin esas tedavisi komplet cerrahi rezeksiyondur. Her ne kadar malign paragangliomaların metastaz ve rekürrens riski olduğu bilinse de; benign paragangliomaların metastaz, malign dönüşüm ve rekürrens potansiyelleri henüz bilinmemektedir. Bu nedenle, malign paragangliomalar gibi benign paragangliomalar da sık aralıklarla uzun dönem takip edilmelidir. Burada, 75 yaşında postmenopozal pelvik kitle olarak başvuran ve veziko-uterin boşluk retroperitonunda apseleşmiş benign paraganglioma tespit edilen bir hasta sunulmuştur.

Anahtar Kelimeler: Ekstraadrenal paraganglioma; retroperitoneal boşluk; pelvis tümörleri; apse

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Paragangliomas (PG) are rare tumors orginating from neural crest cells in the autonomous nervous system.[1] These tumors originate from carotid bifurcation, arcus aorta, zuckerkandl organ and retroperitoneum which are the most common localizations of chemoreceptor bodies.[2] Although PGs may develop at any localization where extra-adrenal chromaffin cell-formed paraganglial tissue exists, the most common localization for these tumors are paraaortic region.[3] Chromaffin tissue related tumors consist 10-20% of PGs and 10% of PGs are located in the retroperitoneal area[4]. Here in we report, a patient with non-functional benign pelvic retroperitoneal paraganglioma mimicking adnexal mass.

CASE REPORT

A 75 year-old postmenopausal, G5 P4 woman was referred with abdominal swelling, pelvic mass, fever and diabetes mellitus. Her past medical history was significant for hypertension for 15 years. She used to take amlodipine besylate 5 mg once daily since then. Her physical examination revealed an approximately 10 cm mass extending up to umbilicus. Its borders and relation with uterus could not be clearly identified. She had generalized abdominal tenderness. With the bimanual gynecological examination the huge mass fulfilling the left adnexal region was palpated. In her laboratory workup complete blood count and routine biochemistry analyses were normal (hemoglobin: 11.1 g/dL, white blood cell: 6.100/uL). Her acute phase reactants were recorded as erythrocyte sedimentation rate (ESR): 60 mm/h, C-reactive protein (CRP): 15.5 mg/dL. The tumor markers were within normal ranges (CA 125: 26.8 U/mL, CA 15-3: 27.3 U/mL). Transvaginal sonography (TVS) showed an 11 x 9 cm, well encapsulated, cystic mass with internal echogenicity, in the left adnexal region. The uterus and right ovary were atrophic, and there were no significant abdominal free fluid. Left ovary could not be clearly examined with TVS. The pelvic computerized tomography (CT) showed a 13.5 x 11.5 x 10.5 cm mass with regular margins which seemed to be arising from uterus and extending over the bladder, containing air and contrast media (Figure 1). In her follow up in hospital, she had been febrile twice with body temperature at least 38^0C. No infectious sources were determined in her physical examinations during the febrile periods. The urine and blood cultures were negative for bacteria. After her written informed consent was taken, she was operated via median infraumbilical laparotomy for postmenopausal adnexal mass. In gross examination, a retroperitoneal mass between the uterus and bladder, which was approximately 10 cm in diameter and abscessed with a thick membrane, was observed. It was adherent to the left pelvic floor. Consequently, the mass was excised completely and the frozen sections were reported to be benign. The patient was discharged at the postoperative 3rd

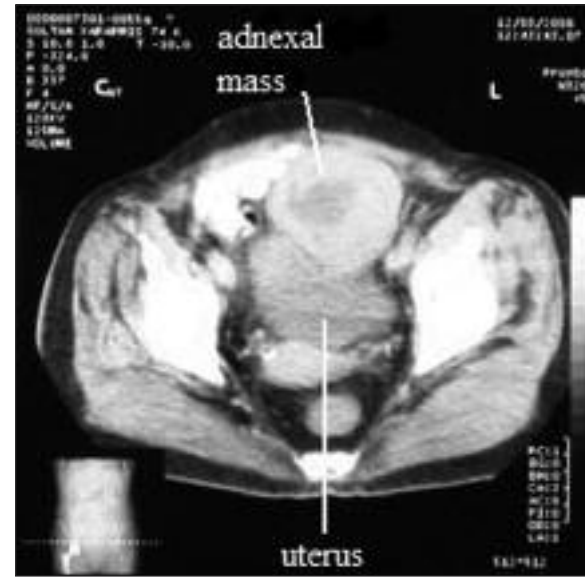

FIGURE 1: The CT image showing the mass with regular margins which seemed to be arising from uterus and extending over the bladder, containing air and contrast media.

day without any complication. The pathological evaluation revealed a benign PG. The hematoxylene-eosine staining revealed tumor cells with homogenous, eosinophilic and polygonal cytoplasms and nuclei with eccentric location (Figure 2A). In the immunohistochemical analysis, the sustentacular cells surrounding the tumor cells were stained positive with S-100 and the tumor cells' cytoplasm were stained weakly positive with chromogranin A and synaptophysin (Figure 2B).

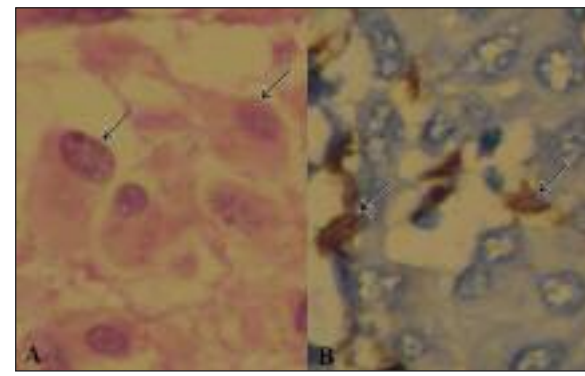

FIGURE 2: Pathological findings in paraganglioma of the pelvic retroperitoneum. A Hematoxylene-eosine staining; tumor cells with homogenous, eosinophilic and polygonal cytoplasms and nuclei with eccentric location (arrows), x40. B Immunohistochemical staining; The sustentacular cells surrounding the tumor cells stained weakly positive with chromogranin A (arrows), x20.

DISCUSSION

Paragangliomas originate from carotid bifurcation, arcus aorta, zuckerkandl organ and retroperitoneum where the chemoreceptor bodies commonly localize. Retroperitoneal PGs typically localize between abdominal aorta and inferior vena cava.[5] However, in this case the tumor localization was very uncommon: retroperitoneal area of vesicouterine space. PGs are classified as functional and non-functional according to catecholamine secretion. Functional PGs may present with headache, palpitation, sweating, hypertension and weight loss whereas a non-functional PG presents with mass and symptoms of compression depending on size and localization as in this case.[6,7] In the present case, as we did not suspect PG preoperatively due to its rare localization in the adnexal region, we did not measure catecholamine levels. However, the patient did not have symptoms like headache, palpitation, sweating or weight loss.

PGs in the pelvic retroperitoneal localization were rarely reported previously.[8,9] In this localization they generally cause compression symptoms like hydroureteronephrosis and back pain.[8]

The prognosis of extra-adrenal PGs is poor when compared to the adrenal PGs. The rates of malignancy for PGs arising from retroperitoneum range between 29% and 40% and these tumors generally display a slow progression.[7] The metastasis rates of malignant PGs range between 28% and 50% and retroperitoneal PGs are much more metastatic than the others.[10] Metastases can be found 7-33 years after the first diagnosis.[2,11] These tumors may spread hematogenously and lymphatic drainage. However, the risks of recurrence, malignant transformation and metastasis are not well known yet for benign PGs. Thus, benign PGs should be followed-up in long term as malignant PGs. Pathologically; malignant PGs can not be clearly differentiated from benign PGs. There are some criteria such as increased mitotic index and necrosis, weak staining with chromogranin A and S-100. There are also some surgical clues differentiating malignant PGs such as incomplete resection and local invasion.

Primary treatment for PGs is surgical excision of the mass. Especially in functional PG operations, the possibility of hypertensive attacks during manipulation of the mass should be kept in mind. The treatment of malignant PGs may also include radiotherapy, chemotherapy and MIBG or combination of these modalities.

In conclusion, although it's very rare pelvic retroperitoneal PGs should be kept in mind in the differential diagnosis of adnexal masses. The symptoms and findings of the patient may include important clues particularly regarding functional PGs. Late metastases and recurrence might complicate malignant PGs. Although malignant transformation and recurrence potentials for benign PGs are not well documented yet, these tumors should also be followed up frequently for long term.

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