Case Report

Pelvic retroperitoneal paraganglioma mimicking an ovarian mass

E. Kuscu[1], *M.D.*; **M. Oktem**[1], *M.D.*; **D. Eroglu**[1], *M.D.*; **A. Haberal**[1], *M.D.*; **B. Bilezikci**[2], *M.D.*; **B. Demirhan**[2], *M.D.*

[1]*Department of Obstetrics and Gynecology, Ankara*

[2]*Department of Pathology, Baskent University Faculty of Medicine, Ankara (Turkey)*

Summary

A 75-year-old hypertensive woman was referred with ultrasound findings of a 40 x 35 mm semi-solid right adnexal mass and right hydroureteronephrosis. She complained of headache and right-sided back pain. Computed tomography demonstrated a cystic adnexal mass that did not appear to originate from the right ovary and grade 2 hydroureteronephrosis. Magnetic resonance imaging indicated that the mass originated from the right ovary. Tumor markers were in the normal range. Exploratory laparotomy was performed to determine the origin of the lesion, and revealed a retroperitoneal mass obstructing the right ureter. The mass was completely removed and and the histopathologic diagnosis was paraganglioma.

Key words: Paraganglioma; Pelvic retroperitoneal mass; Extra-adrenal; Pheochromocytoma; Catecholamine.

Introduction

Extra-adrenal pheochromocytomas, also known as paragangliomas, are rare tumors that arise from chromaffin cells in the autonomic nervous system. These neoplasms usually develop along the paraaortic ganglia chain. Pheochromocytomas occur in approximately 0.1% of the hypertensive population, and 10% are extra-adrenal [1]. The typical clinical manifestations of these tumors are paroxysmal hypertension, headache, excessive perspiration and palpitations. Excess production of catecholamine causes these symptoms. However, some patients exhibit no obvious signs, and these lesions may be discovered during unrelated surgery, at autopsy, or when computed tomography (CT) of the abdomen is performed for evaluation of multiple endocrine neoplasia, abdominal pain or abdominal masses [2].

We describe a patient who was preoperatively misdiagnosed as having an adnexal mass. The lesion turned out to be a retroperitoneal paraganglioma that was obstructing the right ureter.

Case Report

A 75-year-old female patient (gravida 6, para 4, aborto 2) was referred with suspicion of a right ovarian tumor based on pelvic ultrasound examination. Her blood pressure was 150/100 mmHg, and she complained of headaches and right-sided back pain. Abdominal-pelvic ultrasound examination revealed a 45 x 30 mm semi-solid right adnexal mass that was obstructing the ureter and grade 2 hydroureteronephrosis. After an unsuccessful attempt at retrograde catheterization of the right ureter, right percutaneous nephrostomy was performed. Antegrade pyelography showed narrowing of the distal portion of the right ureter, and dilatation of the proximal ureter and the collecting system of the right kidney. These abnormalities were attributed to the right adnexal mass. Abdominal-pelvic CT demonstrated severe hydroureteronephrosis and showed a 40 x 35 mm cystic adnexal mass obstructing the right ureter. The scan indicated that the mass was independent of the right ovary. Abdominal-pelvic magnetic resonance imaging was also performed in effort to determine the origin of the adnexal lesion. In contrast to CT, these images indicated that the mass originated from the right ovary. Upper and lower gastrointestinal endoscopy and bilateral mammography revealed no malignancy. A cervicovaginal smear was assessed as chronic cervicitis. Serum analysis for tumor markers showed nothing significant (CA-125 = 8.5 u/ml; CA 19-9 = 2.32 u/ml; CA 15-3 = 8 u/ml; carcinoembryonic antigen = 2 ng/ml).

We decided to perform an explorative laparotomy to definitively determine the origin of the mass. At surgery, the patient's ovaries, fallopian tubes and uterus all appeared normal. A blue-violet mass that arose from the pelvic retroperitoneum was found wrapped around and obstructing the right ureter. The lesion was excised and frozen-section analysis identified it as benign tissue. After this was established, total abdominal hysterectomy and bilateral salpingo-oophorectomy were performed. Urinary drainage was improved after the surgery. The definitive histopathologic diagnosis was paraganglioma (Figures 1 and 2).

Discussion

The presence of a complex mass in a postmenopausal woman raises suspicion of a neoplastic process. This situation usually warrants surgical exploration because it can be difficult to differentiate a retroperitoneal process from an intraperitoneal entity on ultrasound. Gynecologic disease rarely presents in the form of a retroperitoneal mass. When such an abnormality is encountered on surgical exploration, other etiologies such as neurologic, vascular and lymphatic processes should be considered.

Revised manuscript accepted for publication September 20, 2004

Fig. 1

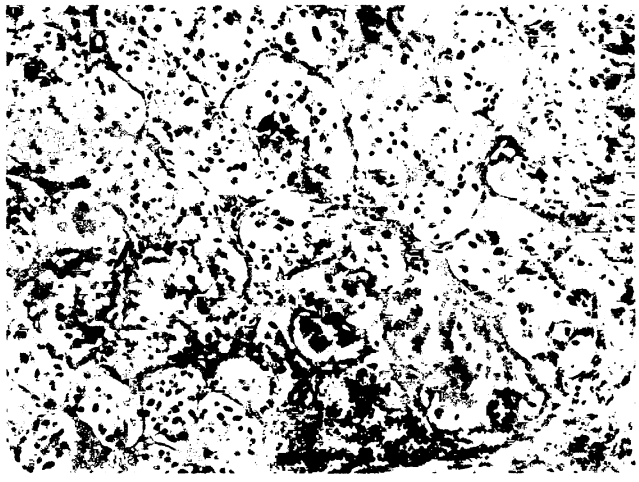

Fig.

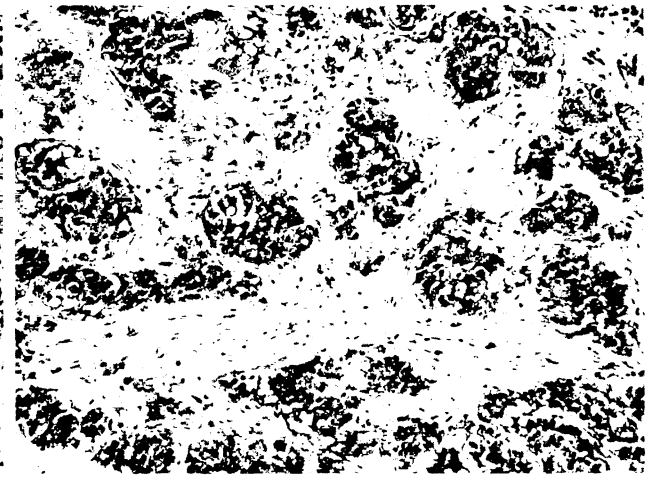

Figure 1. — A section of the paraganglioma reveals rich vascularity and organoid structures (H&E x 20).
Figure 2. — Immunohistochemical analysis revealed positivity for neuron-specific enolase (NSE) in organoid structures (immunoperoxidase x 20).

Like ovarian tumors, pheochromocytomas are morphologically classified as cystic, solid or mixed. Due to this similar range of morphological types, it is difficult to distinguish paragangliomas from ovarian neoplasms on ultrasound or CT. Lane *et al.* reported that the CT findings for paraganglioma are non-specific, but that elevated catecholamine levels and paraaortic lesion location are suggestive of the diagnosis [3]. In our case, it was not possible to differentiate the mass from an intraperitoneal process on pelvic ultrasound or on magnetic resonance imaging.

Careful diagnosis and treatment of pheochromocytomas is necessary in order to avoid the catecholamine crises that are sometimes associated with these tumors. These patients may be at great risk during any surgical procedure [4]. Drugs or physical manipulation may induce life-threatening reactions during surgery [5]. The risk of a hypertensive crisis from surgical excision can be reduced by α- and β-adrenergic blockade. Our patient did not experience a hypertensive crisis during her surgery.

In conclusion, any gynecologist preparing to surgically remove a suspicious or undiagnosed pelvic mass should consider the possibility of a retroperitoneal tumor such as a paraganglioma. Unfortunately, when these tumors are diagnosed during surgery, the risks of morbidity and mortality are higher.

References

[1] Landsberg L., Young J.B.: "Pheochromocytoma". In: Fucci A.S., Braunwald E., Isselbacher K.J., Wilson J.D., Martin J.B., Kasper D.L., Hauser S.L., Longo D.L. (eds.). Harrison's Principles of Internal Medicine. 14th ed. New York: McGraw-Hill, 1998, 2057.
[2] Krane N.K.: "Clinically unsuspected pheochromocytomas. Experience at Henry Ford Hospital and a review of the literature". *Arch. Intern. Med.,* 1986, *146,* 54.
[3] Lane R.H., Stephens D.H., Reiman H.M.: "Primary retroperitoneal neoplasms: CT findings in 90 cases with clinical and pathological correlation". *Am. J. Radiol.,* 1989, *152,* 83.
[4] Hull C.J.: "Pheochromocytoma. Diagnosis, preoperative preparation and anaesthetic management". *Br. J. Anaesth.*, 1986, *58*, 1453.
[5] Pullerits J., Ein S., Balfe J.W.: "Anaesthesia for pheaochromocytoma". *Can J. Anaesth.*, 1988, *35,* 526.

Address reprint requests to:
M. OKTEM, M.D.
Onur Sokak 38/9
06570 Maltepe Ankara (Turkey)