CASE REPORT

Pelvic Pheochromocytoma: A Rare Lethal Tumor Initially Presenting as Tuboovarian Mass to Gynaecologist

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Received: 16 March 2013 / Accepted: 2 September 2013 / Published online: 27 September 2013
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Introduction

Extra-adrenal pheochromocytomas are rare, but potentially lethal, tumors which may initially present to gynecologists masquerading as a tuboovarian mass leading to a potentially dangerous exploration in an unprepared patient. We present a case of a large extra-adrenal pheochromocytoma which was diagnosed intraoperatively because of sudden hypertension.

Case Report

A 45-year-old female with no medical co-morbidities underwent exploration for a tuboovarian mass at a private hospital where she presented with vague pelvic pain. During manipulation, her blood pressure rose to 240/116 mmHg; the procedure was abandoned after taking a biopsy and the patient was referred to our center. The biopsy showed features of a pheochromocytoma. Upon presentation to our institute, she was hemodynamically stable with a pulse rate of 90/min and BP of 160/92 mmHg, and her surgical wound had healed. Upon retrospective questioning, she came out with a history of episodic flushing, sweating, and palpitations for the past 2 months. On examination, there was a palpable $12 \times 10 \text{ cm}^2$ pelvic mass. Contrast enhanced computed tomography (CECT) showed a hypervascular pelvic mass separate from the uterus and adnexa in close relation to the left common iliac vessels, deriving blood supply from them as well as the aorta (Fig 1a, b). A Ga68 DOTATATE PET scan showed marked uptake into the tumor (Fig 1c). Her routine hemogram, biochemical profile, and 24-h urine and plasma metanephrines were within normal limits. She was started on the alpha-blocker Prazosin and later on the Beta-blocker Metoprolol for 2 weeks to control her hypertension. Upon exploration, a $15 \times 12 \text{ cm}^2$ retroperitoneal mass was seen in the pelvis overlying the left common iliac vessels, close to the uterus and adnexa, and was safely removed (Fig 2a, b). The histopathology report was consistent with pheochromocytoma (Fig 2c). The postoperative period was uneventful.

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Fig. 1 **a** Sagittal section of CECT abdomen showing a $12 \times 10 \text{ cm}^2$ pelvic mass in close relation with the uterus. **b** CT Angiography shows the relationship of the mass with the left common iliac vessels and its feeding vessels. **c** Ga68 DOTATATE PET scan showing intense uptake by the tumor

Discussion

Pheochromocytomas are rare endocrine tumors that can present insidiously and remain undiagnosed until death or onset of clear manifestations of catecholamine excess, most commonly as spells of headaches, sweating, palpitations, and hypertension. These tumors can no longer be regarded as a uniform disease entity, but rather as a highly heterogeneous group of chromaffin cell neoplasms with different ages of onset, secretory profiles, locations, and potential for malignancy according to the underlying genetic mutations [1]. Paragangliomas in the retroperitoneum are usually located in close proximity to the aorta and inferior vena cava [2]. Patients with pheochromocytoma may develop complicated and potentially lethal cardiovascular and other complications, especially in the setting of diagnostic or interventional procedures, e.g., upon induction of anesthesia or during surgery due to the potent effect of paroxysmal release of catecholamines [3].

Pelvic paraganglioma may present primarily to the gynecologist and may be treated initially as preeclampsia or other pelvic tumors [4–6]. The patient might present with vague pelvic symptoms and may not be forthcoming with the classical symptoms [5], and the unsuspecting gynecologist might miss these important clues leading to a potentially dangerous unplanned exploration. Suboptimal Imaging may reveal the mass to be related to the uterus and adnexa, and careful thin-section imaging with 3D reconstruction might reveal the exact anatomy of the mass. Plasma and 24-h urine metanephrines might be normal, further adding to the confusion, and sudden intraoperative hypertension upon tumor manipulation may be the first hint toward this pathology, as happened in our patient's case.

A detailed history, physical examination, and a complete laboratory and cardiac evaluation are essential in preparation of a patient with pheochromocytoma for surgery. Echocardiography can be helpful to delineate the degree of cardiac compromise, especially in patients with long-standing hypertension. Normalization of blood volume minimizes the possibility of protracted hypotension or shock resulting from sudden diffuse vasodilation at the time of tumor removal; if catecholamine-induced hyperglycemia is found preoperatively, close monitoring should be initiated [3]. Surgery is the only curative treatment; while preoperative management has changed little over the last 20 years, surgical procedures have evolved so that laparoscopic resection is the standard of care if feasible

Fig. 2 **a** Intraoperative photograph showing the pelvic mass in close relation to the uterus. **b** Cut section of the tumor showing a well-encapsulated mass with areas of hemorrhage and necrosis. **c** Photomicrograph showing Zellballen pattern (arrow) (H&E $\times 100$)

even for parauterine location [1, 8]. Radical surgical resection is the treatment of choice for giant extra-adrenal pheochromocytomas [2]. Although preoperative selective angioembolization does not prevent intraoperative hypertensive crises, by facilitating the pharmacological management of the perioperative hemodynamic instability, we may avoid the onset of major adverse cardiovascular complications [7]; however, we have not found it to be necessary as adequate preoperative adrenergic blockade, careful intraoperative tumor handling, and early ligation of the feeding vessels as guided by the imaging prevented fluctuations of blood pressure in the intraoperative period.

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