Combined partial cystectomy and cesarean delivery in a pregnant female with bladder pheochromocytoma

Ryan S. Hsi, MD,[1] Christopher J. Welty, MD,[1] Mathew D. Sorensen, MD,[1] Aashiyana F. Koreishi, MD,[2] Hunter Wessells, MD[1]

[1]Department of Urology, University of Washington School of Medicine, Seattle, Washington, USA
[2]Department of Pathology, University of Washington School of Medicine, Seattle, Washington, USA

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We report the first known case of concurrent partial cystectomy and cesarean delivery in a pregnant female with bladder pheochromocytoma. A 28-year-old G4P2 female presented at 28 weeks gestation with labile blood pressures requiring three antihypertensive medications. Urinary catecholamines were elevated, and a subsequent MRI showed a 2.6 cm x 3.2 cm bladder wall mass. She underwent combined cesarian section and partial cystectomy at 37 weeks. Fluid resuscitation and vasopressors were required in the immediate postoperative period. While bladder pheochromocytoma with pregnancy is a rare occurrence, concurrent delivery and removal of the bladder tumor can be performed safely.

Key Words: pheochromocytoma, paraganglioma, pregnancy, gravidity, hypertension, urinary bladder neoplasms

Introduction

Pheochromocytoma is a catecholamine-secreting tumor originating from the chromaffin cells of the adrenal medulla and is the underlying etiology of hypertension in 0.1-0.6% of patients with diagnosed hypertension.[1] At least 15% of all pheochromocytoma are extra-adrenal, of which up to 40% are malignant.[1,2] Bladder pheochromocytoma represent less than 1% of all pheochromocytoma.[3] Additional signs and symptoms due to its location in the bladder include painless hematuria, which occurs in half of patients; and paroxysmal hypertension or syncope when the bladder fills or contracts to empty.[3]

We report the first case of concurrent partial cystectomy and cesarean delivery in a pregnant female with bladder pheochromocytoma.

Case report

A 28-year-old gravida 4, para 2 female at 28 weeks gestation presented with preterm contractions. She reported a 7 year history of symptoms triggered by urination, including palpitations, diaphoresis, substernal chest pain, dyspnea, and elevated blood pressure. She denied gross hematuria. She had labile blood pressure from systolic 110-190 and diastolic 70-120.

On admission, she was given intravenous steroids for preterm contractions and was placed on nifedipine, atenolol, and phenoxybenzamine. Work up for pre-eclampsia was negative, with normal urine protein.

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Address correspondence to Dr. Ryan S. Hsi, Department of Urology, University of Washington School of Medicine, 1959 NE Pacific Street, Seattle, WA 98195 USA

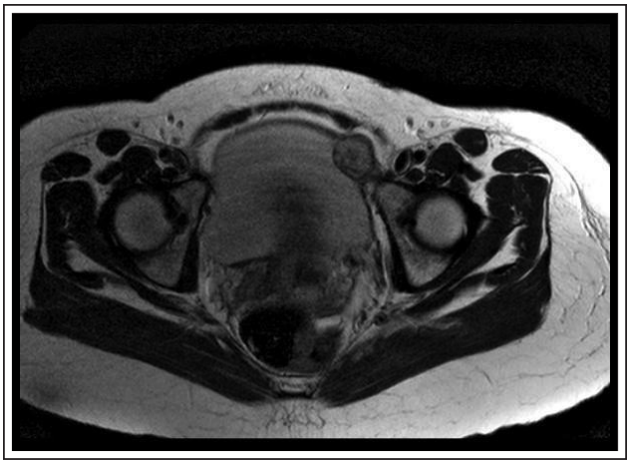

Figure 1. T2-weighted axial MRI image showing a 2.6 cm x 3.2 cm mass adjacent to the left anterior bladder.

She had a urine metanephrine level of 2971 mcg/24 hr (normal 142-510 mcg/24 hr) and normetanephrine level of 2881 mcg/24 hr (normal 103-390 mcg/24 hr). An MRI was performed which demonstrated a 2.6 cm x 3.2 cm mass adjacent to her left anterior bladder, Figure 1. A MIBG scan was not pursued due to concern for fetal toxicity from the radiolabeled tracer, as well as poor bladder visualization from urinary excretion and increased uterine positivity. The patient underwent a scheduled Caesarean section and partial cystectomy at 37 weeks gestation under general anesthesia with an epidural block. Through a Pfannenstiel skin incision, a classical uterine incision was performed. No bladder flap (dissecting the urinary bladder from the lower segment of the uterus as part of a standard part of Caesarean section) was created. The 2.3 kg infant was found to be in a frank breech presentation and was delivered with Apgar scores of 4, 7, and 8. After The uterus was closed, the space of Retzius was developed with blunt dissection. Through a cystotomy medial to the palpable mass, the mass was excised with the accompanying bladder mucosa with a 0.5 cm margin. The bladder wall was closed in two layers.

She required four pressors, including oxytocin, phenylephrine, vasopressin, and norepinephrine after removal of the bladder mass. She was weaned to norepinphrine and vasopressin as the surgical procedure concluded and was transferred to the intensive care unit. She was weaned off pressors the next day, and had an otherwise uneventful recovery with normalization of urinary catecholamines. She was discharged home on postoperative day 6 on a single beta-blocker. Her Jackson-Pratt drain and Foley catheter were removed on post-operative day 3 and 14, respectively. At her two year follow up, she had normal blood pressure and no evidence of recurrence. She will have yearly plasma catecholamine testing and MR of the abdomen and pelvis for a total of 5 years, after which intervals may be increased if there is no evidence of disease.

Gross examination of the surgical specimen revealed a 3.8 cm well-circumscribed, orange-brown mass. Microscopic examination showed a solid mass arising within the bladder wall muscularis propria. The tumor cells were arranged in well-defined nests surrounded by delicate vasculature (the characteristic 'zellballen' architecture), Figure 2. The cells had round to oval nuclei

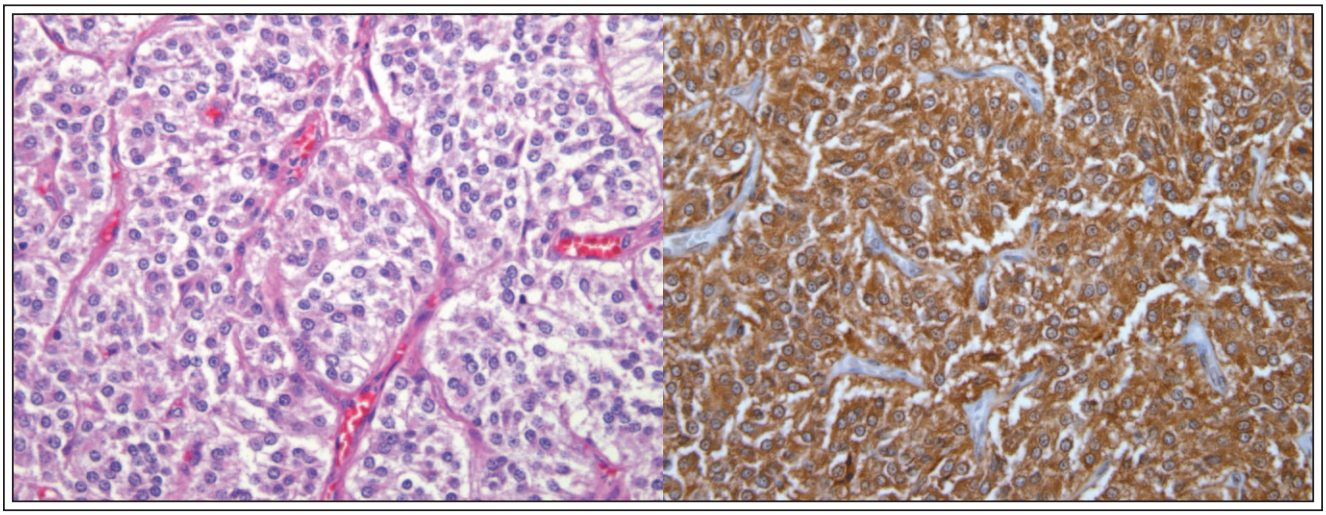

Figure 2 and 3. Microscopic examination showed the mass arising within the bladder wall muscularis propria with the characteristic 'zellballen' architecture (2-Hematoxylin and eosin 200x). The tumor cells were positive for chromogranin (3-400x).

and abundant granular cytoplasm. Mitotic figures and necrosis were not identified. An immunohistochemical stain revealed that the neoplastic cells were diffusely and strongly positive for chromogranin, Figure 3.

Discussion

Pheochromocytoma diagnosed during pregnancy has been previously reported.[4] While the diagnosis of pheochromocytoma is made antepartum in approximately 80% of cases,[4] it is often delayed in the context of pregnancy because signs and symptoms can mimic eclampsia and pre-eclampsia. Pheochromocytoma involving the bladder during pregnancy is a rare occurrence. Previous reports of bladder pheochromocytoma during pregnancy[5-9] involved cases where the diagnosis of pheochromocytoma was either made unexpectedly post-partum,[6,7] or when the surgical excision was delayed until after delivery.[5,7,9] Most pretreatment anti-hypertensive regimens begin with an alpha blocker (e.g. phenoxybenzamine via non-competitive inhibition), with the subsequent addition of calcium channel blockers and/or beta blockers.[1] Despite alpha and beta blockade, patients undergoing pheochromocytoma resection may still have intraoperative blood pressure lability, as demonstrated in our patient. With aggressive blood pressure control in the perioperative period, combined caesarean delivery and partial cystectomy may be performed safely for both mother and child. □

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