Patients With Urinary Bladder Paragangliomas: A Compiled Case Series From a Literature Review for Clinical Management

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Pheochromocytomas are neuroendocrine tumors that arise from chromaffin tissues, typically of the adrenal medulla.[1] Approximately 10% are found in extra-adrenal sites along the sympathetic chain, and as such, they are termed as paragangliomas. The urinary bladder is an unusual primary location, accounting for <0.06% of all bladder tumors and <1% of all paraganglionas.[2] Typical presentation is related to symptoms of catecholamine excess, such as hypertension, post micturition syndrome, and painless hematuria.[1] Here, we report a case of paraganglioma of the urinary bladder found incidentally by computed tomography (CT) in a patient recently diagnosed with a pancreatic mass and a subsequent literature review of paragangliomas.

A 66-year-old African American woman presented to the emergency department with 1-day history of sudden generalized abdominal pain and jaundice. Her past medical history was significant for hypertension, diabetes, and dyslipidemia. Patient denied any history of chest pain, shortness of breath, palpitations, or urinary symptoms except for some vaginal itching. Physical examination was remarkable for hypertension with a blood pressure of 140/80, tenderness in the left upper quadrant of the abdomen, and generalized jaundice with bilateral scleral icterus. CT of the abdomen and pelvis demonstrated a 3 × 3 cm uncinate mass with secondary biliary dilatation and possible superior mesenteric vein involvement, as well as a 1-cm mural high density bladder mass associated with the right bladder wall (Fig. 1).

Cystoscopy with transurethral resection of bladder tumor was performed under general anesthesia. The mass, located at the 10-o'clock position within the right vesical wall, had a surrounding hypervascularity. It was seen as being almost perfectly spherical in shape and well circumscribed (Fig. 2). The intraoperative course was uneventful, even though the preoperative blood pressure was 163/88 and the immediate postoperative blood pressure was 148/81.

Histologically, the bladder mass was a well-circumscribed tumor in the deep lamina propria extending to the deep resection margin. On higher magnification, the tumor cells were arranged in a solid and nested pattern with abundant eosinophilic cytoplasm and a speckled chromatin pattern. Immunohistochemical stains demonstrated tumor cell positivity for synaptophysin and chromogranin and negativity for cytokeratins and uroplakin. Sustentacular cells surrounding the tumor cell nests were positive for S100 protein. The histology and immunophenotype were consistent with a paraganglioma.

Twenty-four-hour urinalysis of catecholamines was performed postoperatively and revealed normal levels of normetanephrine (391 μg; normal, 8-473 μg) and metanephrine (202 μg; normal, 573-1933 μg). As well, urine cytology was negative for malignant epithelial cells.

Several days after the bladder tumor resection, a pancreaticoduodenectomy was performed. Histopathology revealed a well-differentiated, invasive, ductal adenocarcinoma of the pancreas extending into duodenal mucosa with ampullary involvement and focal extension of peripancreatic soft tissue. Lymph vascular and perineural invasions were present with negative margins. Final pathologic staging was pT3N1M0.

DISCUSSION

Paragangliomas are rare primary tumors of the urinary bladder. The first documented case was reported by Zimmerman et al[3] in 1953, and since then, >250 cases have been described worldwide. We performed our literature review with a PubMed search for published case reports of paragangliomas, using the terms "Paragangliomas of the bladder", "Pheochromocytomas of the bladder," and "Extra adrenal Pheochromocytomas." A total of 143 cases were found in this manner. Of the 143

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Figure 1. Computed tomography imaging.

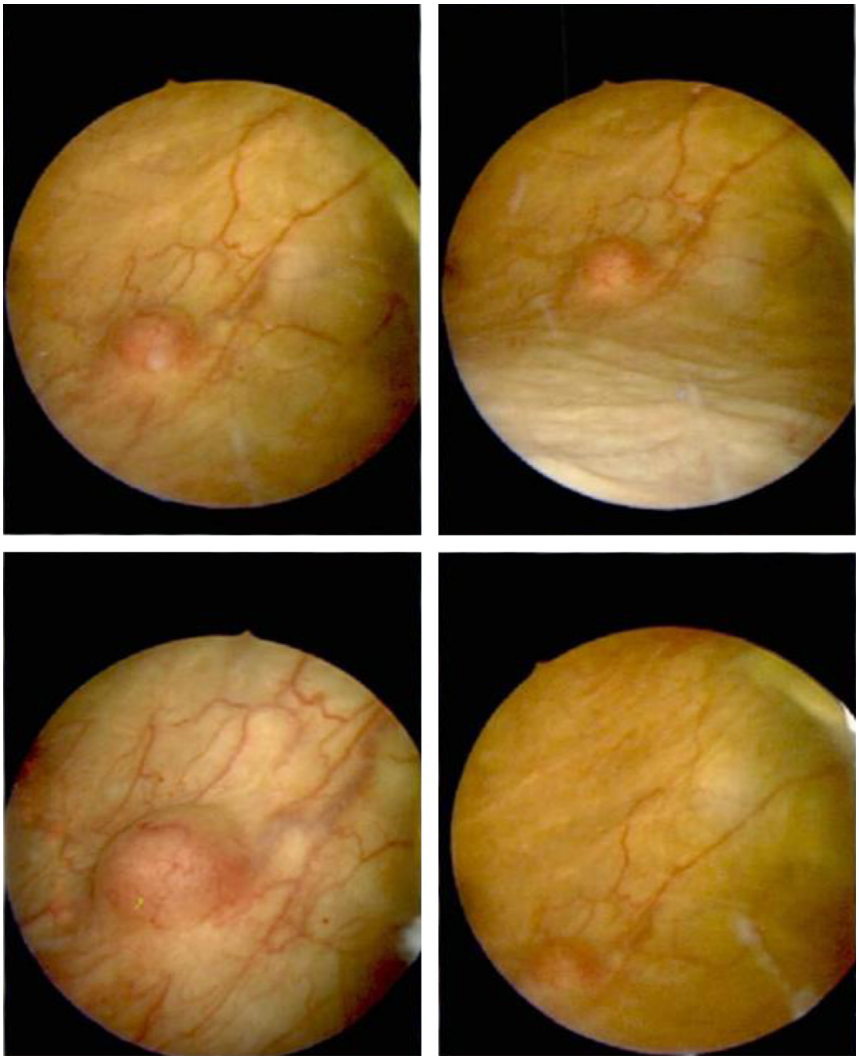

Figure 2. Cystoscopy view of bladder mass on right vesical wall: spherical in shape and well circumscribed.

cases, 128 were reported as case series and 15 were individual case reports. With our case included, the total number of cases in this series was 144 (Tables 1 and 2).

Bladder paragangliomas have been classically reported to present with a triad of symptoms including gross hematuria, hypertension, and a collection of symptoms known as postmicturition syndrome. Postmicturition syndrome includes symptoms of catecholamine release such as sweating, palpitations, headaches, hypertension, syncope, or flushing provoked by over distension of the bladder or micturition. In our case series, we found that gross hematuria was reported in 51% of the cases and was