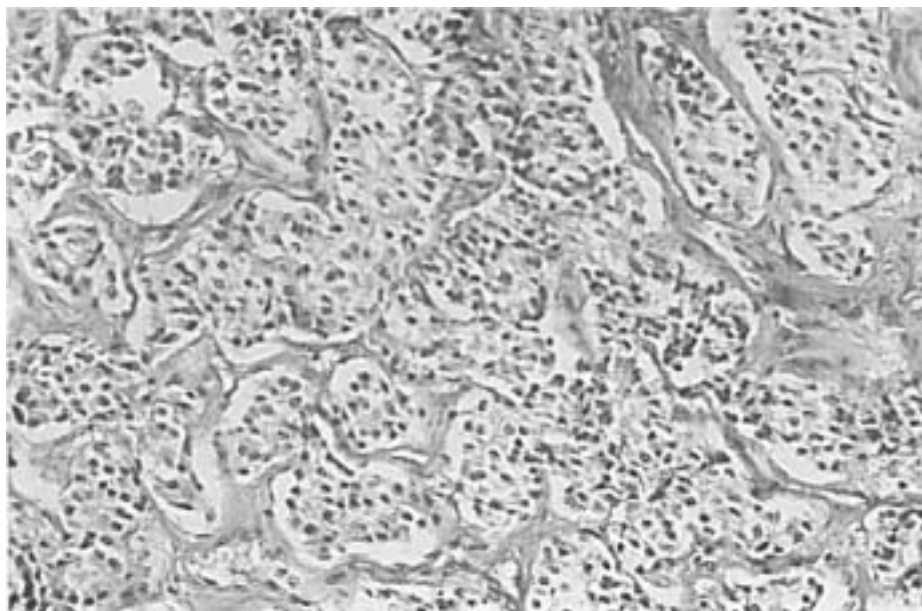


## PHOTOCLINIC



**W**e discuss here the cases of two patients who presented with the chief complaint of painless gross hematuria without hypertension. Ultrasonography of each patient revealed a limited polypoid mass of the posterior wall of the bladder, and cystoscopy showed that each was covered by apparently normal mucosa. Treatment consisted of transurethral resection, and the patients have been followed for 2 years without recurrence.

In case 1, a 24-year-old man presented with gross hematuria since several months before presentation. Sonographic evaluation showed a well-defined mass of 2 cm in diameter in the posterior wall of the bladder.

On cystoscopy, the mass was seen to be covered by mucosa of normal appearance. The patient was hospitalized and necessary tests were performed. Routine blood chemistries were within normal limits and urinary cytology and cultures were negative. Transurethral resection of the ovoid mass, which was located lateral to the left ureteric orifice, was performed.

In case 2, a 35-year-old woman presented with gross hematuria. No history of hypertension was found and she had no other symptoms related to micturition. Sonography showed a 3-cm polypoid mass, which was confirmed by cystoscopy to be on the posterior wall of the urinary bladder. The mass

was resected transurethrally.

The patients' postoperative courses were uneventful, and each was followed for 24 months with cystoscopy, pelvic computed tomography (CT) and measurement of urinary catecholamines. No apparent recurrence of the disease was seen in either case. Examination of excised specimen sections revealed a pink-gray to red-brown, firm, resilient surface.

Histologic slides of sections stained with hematoxylin and eosin showed groups of cells (Zellballen clusters) that were delineated by well-vascularized, fine fibrous networks (Figure). These septae did not penetrate between individual tumor cells. As seen in the Figure, the cell borders cannot be distinguished clearly (present syncytial appearance). Each cell contained round-to-oval shaped open, inconspicuous nuclei and granular eosinophilic cytoplasm. Mild degrees of pleomorphism and mitosis were seen.

**Your Diagnosis?**

See page 129 for diagnosis

**Photoclinic Diagnosis: Primary Paraganglioma of the Urinary Bladder**

Paragangliomas of the urinary bladder are rare neoplasms accounting for less than 0.5% of all bladder tumors and 10% of all extraadrenal paragangliomas. The majority of these neoplasms secrete catecholamines, and more than half of the patients present with hypertension. Common systemic symptoms are episodes of headache, palpitation and sweating, while specific urinary symptoms are dysuria and hematuria. Malignancy is documented in 5% to 15% of cases, but there are no reliable pathologic criteria useful to distinguish benign and malignant tumors. Although immunohistochemical and DNA flow-cytometry studies seem to provide useful prognostic markers, definitive evidence of malignancy is offered only by the presence of metastasis.<sup>1</sup> In 1953, Zimmerman et al reported the first case of pheochromocytoma arising in the urinary bladder.<sup>6</sup> They believed that such tumors arose from paraganglionic tissues present within the bladder wall, most commonly at the dome and the trigon.<sup>2</sup>

The tumors may occur over a wide age range, from childhood to old age,<sup>3</sup> and are found roughly equally in both sexes. Clinically, patients often present with symptoms related to catecholamine excess (headache, tachycardia, dizziness, fainting and hypertension) and, in many, the episodes are associated with micturition. Episodes of paroxysmal hypertension with painless hematuria are practically pathognomonic in these patients. In most cases, diagnosis can be confirmed by measurement of catecholamines and their metabolites in the serum and urine.<sup>4</sup> Tumor size ranges from a few millimeters to 10 cm, and they are generally located in the submucosa, producing an intramural mass covered by intact epithelium, which can infiltrate the adjacent lamina propria and bladder muscle. More rarely, they present as a fungating or multinodular mass, or as an ulcerated lesion.<sup>5</sup>

The patients described above only presented with gross, painless hematuria. Other diagnostic workup such as search for urinary catecholamines or their metabolites, CT or <sup>131</sup>I-iodine-metaiodobenzyl guanidine (<sup>131</sup>I-MIBG) scanning, which are, at present, the most useful tools for diagnosing extraadrenal paragangliomas was not done.<sup>6</sup>

In most cases, the clinical course is benign, but metastasis can develop years after the initial diagnosis and treatment. Paragangliomas are

broadly divided as arising from three anatomic groups: branchiomeric (head and neck), intravagal and aorticosympathetic paraganglia. The tumors arising in the head and neck region are almost always nonfunctional, and tend to be indolent and localized, with a very low incidence of metastasis. Similarly, vagal and mediastinal (aortic body) paragangliomas have a 10% to 15% risk of distant metastasis. The retroperitoneal paraganglioma are divided into adrenal (80–90%) and extraadrenal (10–20%) tumors. The patients with extraadrenal retroperitoneal paragangliomas present with back pain or a palpable mass, and approximately 10% have distant metastasis at the time of diagnosis. Approximately 25% to 60% of the tumors are functional, with symptoms and signs of norepinephrine overproduction compared with the adrenal paraganglioma (pheochromocytoma), with which there may be overproduction of epinephrine and norepinephrine. The metastatic potential of retroperitoneal paraganglioma is higher, ranging from 20% to 42%.<sup>7</sup> On CT scans or magnetic resonance imaging (MRI), paragangliomas show marked contrast enhancement and multiple vascular flow voids. In a comparison of preoperative and postoperative CT and MRI, the <sup>131</sup>I-MIBG scan was found to be the most specific (100%) modality.<sup>5</sup>

Therapy consists of adequate surgical excision. Transurethral resection can be successful in some, but when the intramural location or the size of the tumor does not allow a complete transurethral removal, partial cystectomy is considered more appropriate in most cases.

**Acknowledgment**

*We would like to express our sincere appreciation to Dr. P.V. Kumar for providing photographs and reviewing the slides.*

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## Photoclinic

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### Erratum

1. Please replace the following figure for the figure 3, page 60 of volume 5.



2. The following paragraph should be replaced for the first paragraph in page 1, volume 5.

**Background-Carotid body tumor is not a common disease but it should be considered in the differential diagnosis of neck masses at the mandibular angle. Without a correct diagnosis, the surgeon unexpectedly encounters an unfamiliar and highly hemorrhagic tumor in 23.7% of cases, leading to a fruitless termination of the operation.**