Intraluminal Papillary Urinary Bladder Paraganglioma: case report and review of literatures

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ABSTRACT

Urinary bladder paraganglioma is a very rare tumor, accounting for <0.05% of all bladder tumors. They are usually submucosal tumors most commonly located near the urinary bladder trigone. A deeply located urinary bladder paraganglioma may not be seen on cystoscopy, and pelvic sidewall paraganglioma may mimic urinary bladder paraganglioma. Here we present an extremely rare case of urinary bladder paraganglioma presenting as 39-year-old woman incidentally found to have an intraluminal papillary bladder tumor and it turned out to be a paraganglioma. Most of the paragangliomas are usually functional tumors, which frequently cause symptoms related to catecholamine release. In addition to systemic symptoms like other paragangliomas, bladder paraganglioma may cause specific symptoms during urination and sexual intercourse that warrant attention for diagnosis and management. Preoperative, correct diagnosis of such case with palliative medication using catecholamine antagonist may be crucial to prevent adverse complication.

CASE REPORT

A 39-year-old female patient complained of weight gain from 54kg to 57kg for months. Physical examination and subsequent gynecology sonogram showed a huge pelvic cystic mass with septate structure, compatible with ovarian cystic tumor. Physical examination revealed a pelvic mass up to the level of the umbilicus. The laboratory data were unremarkable. Preoperative computerized tomography (CT) of the abdomen showed a 13 x 8.5-cm pelvic cystic mass containing septae structure and soft tissue mural part. A 1.9 x 2-cm nodular enhancing mass (Fig. 1) projecting to the urinary bladder lumen was incidentally found. The tumor was seen arising from the dome and the posterior wall of the urinary bladder, with moderate enhancement on the post-contrast CT study (Fig. 2). Excretory phase of the CT study (Fig. 3) revealed intraluminal location of the tumor. The patient underwent abdominal total hysterectomy (ATH), salpingo-oophorectomy (BSO) and partial cystectomy. The excised left ovarian cystic tumor and urinary bladder tumor were proved to be mucinous ovarian cystic...
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**Figure 1.** Coronal reformation image of post contrast enhanced CT scan shows an ovarian cystic tumor with septal structure and mural part in pelvic region with extrinsic compression of the urinary bladder. Papillary intraluminal bladder mass (arrow) about 1.9 cm x 2 cm in dome of the urinary bladder is incidentally found, with adjacent bladder wall thickening.

**Figure 2.** Axial view of contrast enhanced CT shows an intraluminal mass (arrow) in posterior wall of urinary bladder with well enhancement.

**Figure 3.** Excretary phase clearly demonstrate the papillary nature of the bladder mass.

**Figure 4.** 40 x magnification. The tumor cells are short spindle arranged in a nesting pattern. Necrosis is absent. Mitoses are scanty.
Urinary bladder paraganglioma is an extremely rare tumor, which accounts for less than 0.06% of all urinary bladder tumors and less than 1% of all pheochromocytomas [5]. Paragangliomas are tumors that arise from autonomic nervous system. The autonomic nervous system has a roughly symmetric distribution with extension from the skull base down to the pelvic floor, so paragangliomas are usually found in posterior mediastinum and retroperitoneum [1]. The autonomic nervous system can be divided into two groups: sympathetic and parasympathetic system. Some ganglia are located in internal organs, so paragangliomas can occur beyond the retroperitoneum and close to other internal organs. Uncommon location of paragangliomas has been reported in gallbladder, urinary bladder, prostate, spermatic cord, uterus, and duodenum [1]. Small nest of paraganglionic tissues may migrate to the urinary bladder wall during development [2]. Sympathetic plexus is scattered among all the layers of the urinary bladder wall and most of the bladder paragangliomas are intramural in location [6]. Thus, they usually present as submucosal tumors. Cystoscopy may reveal broad-based, hypervascular submucosal tumors. The intraluminal papillary or cauliflower growth pattern is rarely seen [7]. Bladder paraganglioma projecting into urinary bladder lumen, like our case, make the diagnosis more difficult.

Paragangliomas are functional tumors in more than half of the cases, and patients commonly present with symptoms related to excessive secretion of catecholamine, such as palpitations, headache, sweating, and hypertension [8]. Micturition syncope is the clinical hallmark of catecholamine-secreting bladder paraganglioma. It is found in 65% of patients. Unlike physiological micturition syncope, it is associated symptoms related to catecholamines release [9]. The triad of hypertension, hematuria, and symptoms on micturition or sexual activity is considered almost diagnostic of the condition, and is reported variably in 50 to 100 percent of such patients [10]. Prolonged catecholamine exposure may cause receptor down regulation [3]. Some patients may have only painless hematuria [5]. In case of bladder paragangliomas larger than 2 cm, 65% of patients may develop hematuria [11]. Asymptomatic patient, like our case, has also been reported. It is estimated that 27% of the urinary bladder paragangliomas do not feature any hormonal activity [11].

Extraadrenal paragangliomas have nearly identical imaging features, including a homogeneous or heterogeneous hyperenhancing soft-tissue mass at CT, multiple areas of signal void interspersed with hyperintense foci (salt-and-pepper appearance) within tumor at MRI, and an intense tumor blush with enlarged feeding arteries at angiography [11]. Smaller tumors are more likely to be homogeneous in attenuation and sharply marginated as compared with larger ones. Punctate calcification or focal areas of high attenuation caused by acute hemorrhage may be seen in some cases [12].

Like all paragangliomas, urinary bladder paragangliomas are usually hypervascular tumors; on the other hand, common transitional cell carcinoma of the bladder is a hypovascular lesion [13]. When paraganglioma is suspected, biopsy should be avoided, because it can result in significant bleeding [14]. Besides, it may provoke a hypertensive crisis [15]. Laboratory exam is preferred for confirming the diagnosis if bladder paraganglioma is suspected preoperatively. Endocrine tests, including vanillylmandelic acid (VMA) in 24 hour urine sample, serum epinephrine, and urine catecholamine level in case of urinary bladder paraganglioma, should be performed. In one study of 858 subjects, the most sensitive test (99%) was the plasma free metanephrine level, whereas the urinary VMA level yielded the most specific results 95% [16]. Measurement of urine catecholamine may make the diagnosis particularly for bladder paraganglioma [15, 16].

Treatment modalities include transurethral resection and partial or total cystectomy combined with pelvic lymph node dissection, especially in the presence of proven metastasis [14]. Although laparoscopic removal has been reported [17], partial cystectomy is a more preferred method as the tumor needs to be completely removed to avoid possible recurrence [17]. Preoperative treatment with catecholamine or calcium channel blockers was suggested to prevent catecholamine release crisis during operation [18].

Our case is unique in that it presents intraluminal papillary growth mimicking the growth pattern of ordinary uroepithelial bladder tumor. When characteristic symptoms are absent and the morphology of the tumor mimics ordinary urinary bladder tumor, preoperative diagnosis is difficult. Retrospective review of her clinical history shows neither episode of hypertension nor hematuria. Unlike our case, most of the paragangliomas are functional tumors. Preoperative, prompt diagnosis of such case with palliative medication using catecholamine antagonist may be crucial to prevent adverse complication.
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REFERENCES