Malignant paraganglioma of the urinary bladder is a rare tumor. Clinical diagnosis of this tumor relies on high index of suspicion and high levels of blood and urine catecholamines. The malignancy of the paraganglioma is established upon the presence of local aggressive behavior or distant metastasis. Imaging studies play an important role in detecting tumor foci, in determining the extent of the tumor invasion as well as in pre-surgical evaluation. We have presented the CT and MR imaging features of a case of malignant paraganglioma of the urinary bladder. The tumor demonstrated high signal intensity on both T1-weighted images and T2-weighted images and being enhanced by MRI contrast medium. MR imaging is superior to CT in delineating the extent of the tumor invasion into the bladder wall because of its better tissue contrast and multiplanar imaging capability.

Key words: CT, MR, urinary bladder tumor, malignant urinary bladder paraganglioma

Paraganglioma of the urinary bladder is rare, consisting of 10% of extra-adrenal paragangliomas, and only accounts for less than 0.06% of all bladder tumors [1-3]. Paraganglioma arises from the cells of neural crest embryonically and belongs to the amine-precursor-uptake decarboxylation system [4]. The origin of paraganglioma of the bladder is uncertain. Cheng and coworkers have suggested that the tumor originates from paraganglionic cells that migrate into bladder muscle [5]. Malignancy and recurrence rates of this tumor are approximately 5% and 18%, respectively [1]. The diagnosis of paraganglioma of the bladder relies on a high index of suspicion based on the clinical histories and is confirmed by the quantitation of the serum level of catecholamine as well as their metabolites in 24-hour urine. Imaging studies of the paraganglioma of urinary bladder provide pre-operative evaluation and surgical planning. Here, we present the CT and MR imaging features of a malignant paraganglioma of the urinary bladder.

CASE REPORT

A 31-year-old woman presented with intermittent palpitation and hypertension for approximately eight months. She experienced the first episode of headache and dizziness during micturition about eight years ago. Physical examination showed an elevation of blood pressure (up to 210/110 mmHg) and slightly tachycardia (80 ~ 90/min), which exacerbated especially after micturition and defecation. The blood chemistry work-up demonstrated elevated levels of plasma catecholamine, including norepinephrine (7012 pg/ml) and dopamine (531...
The 24-hour urine vanillyl mandleelic acid (VMA) was 19.1 mg/day (normal: 1.0-7.0 mg/day). The other physical examinations and laboratory data were unremarkable. Under impression of pheochromocytoma, the non-contrast CT scanning of the abdomen and the pelvis was performed to survey the lesion foci and demonstrated the tumor mass protruding from the base of the urinary bladder (fig. 1). MR imaging depicted a bulky mass, measuring 5x3.3x6 cm in size, at the base of urinary bladder with slightly and heterogeneous high signal intensity on both T1-weighted images and T2-weighted images (fig. 2). After intravenous gadolinium contrast administration, the lesion was homogeneously enhanced (fig. 3). The mass caused indentation to the urinary bladder and encroached the upper portion of the urethra. Enlarged lymph nodes at left obturator and left external iliac regions were also seen (fig. 4). The patient underwent a partial cystectomy with reconstruction of bladder neck and pelvic lymph node dissection. The operative findings include: (1) a 5x5 cm solid mass at the base and anterior wall of urinary bladder with the presence of engorged veins surrounding the tumor, and (2) enlarged lymph nodes at left obturator and the left iliac groups, which showed malignant cells infiltration on the frozen section. Histopathologic

Figure 1. Non-contrast CT image of malignant paraganglioma of urinary bladder. The malignant paraganglioma (arrow) is located at the base of urinary bladder and shows iso-dense attenuation to the uterine cervix or bladder wall.

Figure 2. MR image of malignant paraganglioma. T2-weighted image (4000/96) of the tumor on sagittal view depicts a cauliflower-like mass with inhomogeneous higher signal intensity (in comparison with muscular layer) on the base of urinary bladder. The tumor has penetrated through the muscular layer of urinary bladder to the perivesical fat (arrow).

Figure 3. Gadolinium-enhanced MR image of malignant paraganglioma of urinary bladder. Axial demonstrates the tumor (arrow) is well enhanced and disrupted the muscular layer of urinary bladder.

Figure 4. Gadolinium-enhanced MR image of pelvis. Enlarged lymph nodes with well enhancement (arrow) at left internal iliac nodes are shown. The histopathologic examination confirmed malignant paraganglioma cells in the nodes.
studies of the tumor and resected lymph nodes demonstrated a typical paraganglioma with lymph node metastasis. The tumor cells infiltrated detrusor muscle and peri-vesical soft tissue. Vascular invasion was noted. Metastases were identified in four out of ten sampled left obturator lymph nodes. After operation, the regularly follow-up MR imaging of the pelvis, the serum catecholamine and the urine VMA level were within normal ranges for 19 months.

**DISCUSSION**

As in other parts of the body such as adrenal medulla, neck region, and retroperitoneum, paraganglioma of the urinary bladder may secrete catecholamines, resulting in hypertension, palpitation, headache, sweating and anxiety [6]. However, approximately 60% of the cases with paraganglioma of bladder present symptoms of hematuria [1]. The most common location of the tumor is trigone of the urinary bladder, followed by the dome and the anterior wall [1]. Malignant paraganglioma of the urinary bladder is very rare and is diagnosed on the basis of its aggressive behavior, such as regional lymph node or distant metastasis. Cheng et al. recently reviewed the morphologic features of 16 cases (13, benign; 3, malignant) of paraganglioma of the urinary bladder and suggested that histological criteria, such as DNA ploidy, p53 alteration, or MIB-1 monoclonal antibody labeling index, may not be reliable in separating benign from malignant paraganglioma. Instead, they suggested tumors of the advanced stage (T3) at presentation were virtually associated with higher risk of distant metastasis [5]. As a result, precise imaging of the tumor in respect to the location and extent of invasion in the bladder muscle as well as the lymph node involvement not only provides the information for pre-operative planning but also helps in predicting the prognosis. MR imaging provides better tissue contrast and multiplanar imaging capability. On MR imaging, the malignant paraganglioma showed iso- to hyperintense signal on T1-weighted images and hyperintense signal on T2-weighted images. The tumor was enhanced after gadolinium administration. The MR features of paraganglioma of the urinary bladder are, however, not specific. Other differential diagnoses include leiomyoma, neurofibroma, hemangioma, and inflammatory pseudotumor [7]. In our case, MR imaging clearly demonstrated tumor invasion through the deep detrusor muscle and perivesical fat tissue; while, on CT, the invasion of tumor cannot be evaluated confidently due to inherent limitation of transverse section and poor contrast of muscle layer and perivesical fascia.

In conclusion, we have presented the CT and MR images of malignant paraganglioma of the urinary bladder. MR imaging is superior to CT in depicting the extent of tumor invasion, which is important for pre-operative planning and may be used as an important prognostic indicator.

**REFERENCE**

膀胱惡性副神經節瘤：病例報告

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發生在膀胱的惡性副神經節瘤為一罕見的腫瘤，臨床上診斷這種腫瘤需要依靠血液及尿液中，偽茶酚胺的濃度上昇指數來判斷。對於副神經節瘤的惡性變化之判定，在於腫瘤是否造成周圍組織的侵犯以及遠端轉移。因此影像學的檢查在偵測腫瘤位置，腫瘤侵犯程度以及術前之評估上扮演了重要的角色。在此我們報告一例發生在膀胱的惡性副神經節瘤在電腦斷層及磁振造影之影像。這個腫瘤在磁振造影的 T1 及 T2 影像上呈現高訊號，並在施打對比劑後有顯影的情形。相較於電腦斷層攝影，磁振造影影像可藉由顯示腫瘤與周圍組織的對比，以及多軸切面影像呈現，將腫瘤侵犯膀胱的狀況更清楚表現出來。

關鍵詞：電腦斷層，磁振造影，膀胱腫瘤，惡性膀胱副神經節瘤