Paraganglioma of urinary bladder. Report of a very rare case with one year follow-up

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KEY WORDS

minor pelvis tumor ◗ bladder tumor ◗ paraganglioma ◗ pheochromocytoma

ABSTRACT

Diagnosis of minor pelvis tumors is often very difficult and unclear. Paraganglioma is a very rare tumor; in addition perivesical localization is occasionally reported. This article covers a case of a 36-year old man with a perivesical tumor where his first symptom was acute abdominal pain. The patient underwent a laparotomy in the surgical department because of a major peritoneal bleeding. After further urological treatment, a perivesical tumor was resected. Histopathological research indicated a bladder's paraganglioma. Our report presents this unusual case including way of treatment, following laboratory and radiological diagnostics during one-year observation.

INTRODUCTION

Paraganglioma is a neoplasm that proliferates from the sympathetic nervous system. It belongs to the pheochromocytoma group. The name of these tumors comes from chromophil granules that turn brown under the influence of chrome salt (Fig. 1). Pheochromocytoma of the suprarenal gland is most common (75-85%) [1]. Other localizations are possible almost everywhere because of the wide-spread elements of the sympathetic nervous system. Most of them occur in a retroperitoneal area near suprarenal gland, in a renal hilus and in aortal zone(85%) [1, 2]. Perivesical area and bladder are involved in 1% [1, 2, 3]. Rarely, these tumors proliferate in chest (12%) and neck (2%) [4]. The etiopathogenesis of pheochromocytoma is not explained, but in 20-25% of cases a strong connection with congenital gene mutations (RET, VHL, NF-1, SDH) was reported [5, 6, 7]. These are responsible for hereditary syndromes such as multiple endocrine neoplasia-MEN 2A and 2B, von Hippel-Lindau syndrome, neurofibromatosis type 1, and familial pheochromocytoma-paraganglioma syndrome [5, 6, 7]. These tumors are benign in most cases, but nearly 10% have a malignant course [2, 5]. A higher percentage of malignant transformation occurs in an extrasuprarenal localization. At present, there are no clear signs that could confirm a malignant type of tumor with certainty. Only one confident criterion of malignancy is the undisputed presence of distant metastases in regions normally free of typical autonomous nerve tissue. Morphologically malignant tumors are usually bigger, infiltrate adjacent structures, and include necrotic and hemorrhagic areas. Mitotic activity is also higher than in benign neoplasms [8]. Pheochromocytoma is very rare but has an influence on secondary hypertension and, what is most important, could provoke an acute life-threatening condition. Paraganglioma's symptoms are connected with the tumor's presence (haematuria – 55-60% [2, 9], urgency, a painful sensation in abdomen) or with endocrine activity (hypertension – 85% [7, 8], tachycardia, flushing, loss of consciousness during micturition [2, 9]). In diagnosis of paraganglioma structural, pictorial examinations (CT, USG, MRI) [4] are applied, which can assess the morphology of a tumor. Scintigraphy of the adrenal system performed with a metaiodobenzylguanidine marker (^{123}ImIBG) plays a very important role [2, 3]. It also allows estimating the functionality of the transport system and prevailing storage of biogenic amine (Vesicular Monoamine Transporter: VMAT). Furthermore, scintigraphy of somatostatin receptors (SRS),

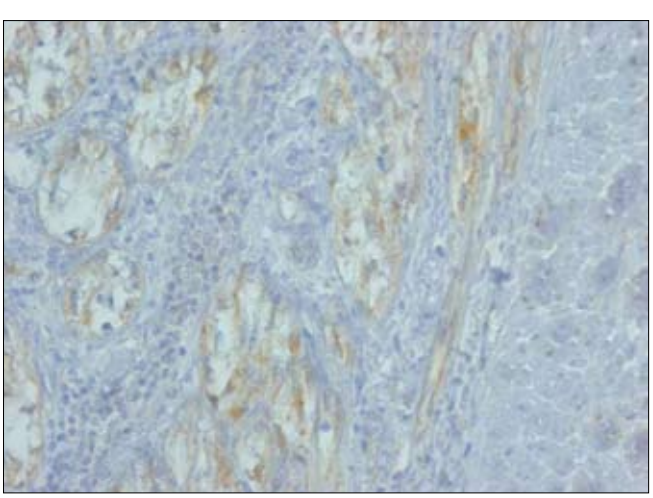

Fig. 1. Positive reaction to chromogranin presence.

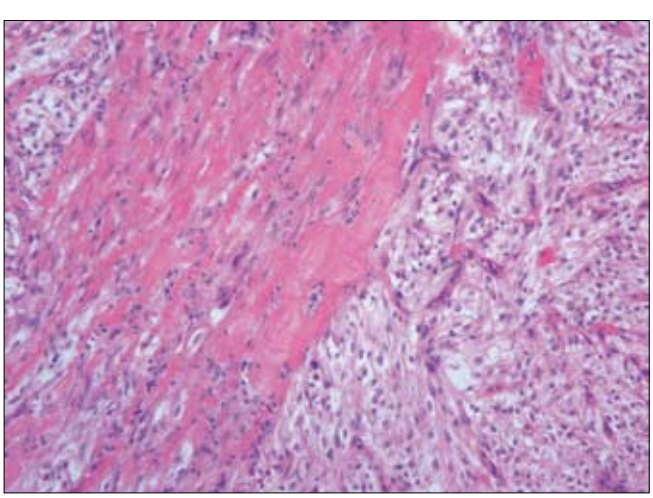

Fig. 2. Infiltration of bladder's muscular lamina.

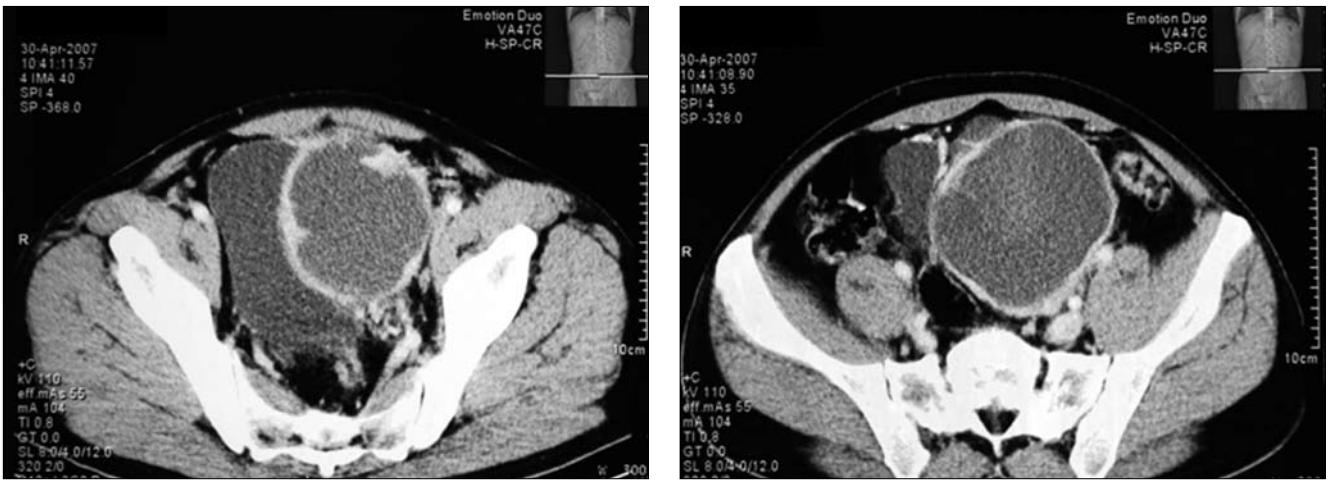

Fig. 3, 4. CT. Paraganglioma of the urinary bladder – preoperative view. Tumor is to the left of the bladder.

which is most often performed with ^{99m}TcHYNICTOC, is very helpful in functional diagnosis of these kinds of tumors [10].

CASE REPORT

A 36-year's old man was accepted at our Clinic after laparotomy, which was performed one month earlier. On the basis of the patient's medical documentation and history, we established that during minor exercise the patient felt acute abdominal pain that was particularly intense in the hypogastrium. He was admitted to a regional hospital. During an abdominal ultrasound, a notable amount of liquid was confirmed, but without visceral trauma. Performed cystography was also normal. The decision to perform a laparotomy was made, which was done by transversal incision. Blood was present in the peritoneum, but the liver, spleen and other peritoneal organs weren't damaged. The only source of bleeding was "intramural disruption" of the bladder wall, which was sutured. No complications were reported in the postoperative period. In course of further diagnostics, abdominal and pelvic computer tomography was performed. A solid cystic tumor, localized extravesically, was detected. The dimensions of the lesion were 10 x 9 x 7 cm (Fig. 3). No particular conclusion about the characteristics of the tumor were stated. The patient was admitted to our department. He did not complain about abdominal pain, dysuria, or hematuria. He did not suffer from hypertension. We performed a bladder ultrasound and diagnosed a cystic structure near the left-posterior wall of the bladder. The cistern failed to empty after micturition. During cystoscopy, the urothelium was normal. There was no tumor or diverticula detected. An invagination on the left-posterior wall was visible, suggesting compression of the bladder wall from the outside. As an operation was expected in the near future, a D-J catheter was inserted into the left kidney. After further analysis, we came to the conclusion that the reason of bleeding was damage to the unusual tumor, which was recognized as a bladder and sutured during the surgical operation. The patient was prepared for resection of the lesion. We performed a laparotomy by median-hypogastric incision. We confirmed a large (about 10 cm in diameter) cystic tumor, localized extravesically, and infiltrating the bladder wall. The tumor was surrounded by large blood vessels. Despite intense bleeding, we resected the tumor radically with a part of the bladder wall. The patient was reoperated after six hours because of bleeding. During the operation and the day after the operation the patient's blood pressure was very high. He had to receive high doses of anti-hypertensive medicines. Further postoperative time was uncomplicated. In histopathological investigation we received the following result: A- paraganglioma- tumor 10 cm in maximum diameter, necrotic in the central part, adjacent to the muscular lamina of the bladder wall, resected with capsule (Fig. 2). B-part of the bladder wall free of neoplasmic infiltration, a neoplasmic focus 1 cm in diameter was found near a neural bundle. Two months after operation, because of histopathological diagnosis, adrenergic system scintigraphy (^{123} ImIBG) was performed and somatostatin receptors (SRS) were investigated. This was done using the Single Photon Emission Computed Tomography (SPECT) technique to achieve fusion scintigraphy and a structural tomography image. In both scintigraphic investigations, no pathological accumulations of the marker were detected in the site where the tumor used to be located or others regions like abdomen, chest, neck and brain. Five months after the operation, during a computed tomography of the abdomen, chest and brain, one very small, hypervascular focus in the second segment of the liver was discovered. The lesion was diagnosed as an angioma. During the next control computed tomography executed ten months after operation, the morphology of this transformation was identical. There were no other pathologies. Clinically the patient was in an excellent condition. To exclude potential genetic syndrome (SDH mutation), blood samples were collected, which are still being processed. The patient is under an attentive further follow up.

DISCUSSION

Paraganglioma of the urinary bladder is very rare, but should not be missed in distinction of unusual lesions in the minor pelvis. Up to now, there are no detailed algorithms providing guidance on the management of the patient following the operation for paraganglioma of the urinary bladder. Structural investigations, most often computed tomography of the enclosed trunk, neck and brain and functional research such as scintigraphy ^{123}ImIBG and SRS ^{99m}TcHYNICTOC similar extension to CT are absolutely required in every case where histopathological confirmation of paraganglioma or pheochromocytoma is made. The above mentioned investigations should determine the stage of progression of the disease. Diagnostics should be focused on searching for metastases and detection of potential local recurrence. Genetic research is obligatory because of the possibility of a hereditary background [9]. Clinical observation of the patient, laboratory investigations estimating catecholamine metabolites in serum and urine, and pictorial control (structural and functional) should be performed regularly. The dilemma is how often these investigations should be executed? The question remains open and requires further exploration.

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