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Oncology

Atypical calcified paraganglioma of the urinary bladder: A new case report

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Introduction

Paraganglioma is a rare neuroendocrine tumor defined by the World Health Organization (WHO) as catecholamine-producing tumor originating outside the adrenal glands.

The paraganglioma of the bladder is very rare and represents less than 6% of the whole range of paragangliomas and less than 1% of the bladder tumors.

This case aims to remind the clinical, histological and therapeutic features of this rare tumor.

Case presentation

We report a case of a 45-year-old woman, with no significant past medical history, suffering since one year of irritative lower urinary tract symptoms associated with recurrent episodic pelvic pain reducing life quality. There was no history of micturition attacks including headaches, palpitations, syncope and profuse sweating. Physical examination was normal and blood pressure was normal at 130/80 mmHg. Cytobacteriological examination of the urine was negative.

Ultrasonography of urinary tract had shown a mass depending of the left bladder wall.

The CT scan, with intravenous administration of contrast medium, revealed a well limited nodular calcified lesion, measuring 4 cm with increased density in its soft parts, showing intense, heterogeneous enhancement, in the left lateral wall of the bladder (Fig. 1).

Cystoscopy was therefore performed showing a calcified 4-centimeters mass depending on the left bladder wall.

The patient underwent complete and deep transurethral resection of the bladder tumor. The mass was sessile and its tip was completely calcified.

The histological examination of the tumor confirmed the diagnosis of vesical paraganglioma showing tumorous small cells with a positive immunostaining for synaptophysin and a strong positivity for chromogranin (Fig. 2).

24 hours urinary catecholamine levels (norepinephrine, vanillylmandelic acid (VMA) and metanephrines) were normal.

During the follow-up, two weeks after the endoscopic resection an abdominal and pelvic Computerized Tomography scan was performed and showed a thickening of the left lateral wall extended to the dome of the bladder measuring 8 mm with integrity of the perivesical fat.

We have therefore considered doing partial cystectomy as a curative treatment but preoperative abdominal and pelvic computer tomography, performed after six months, revealed no signs of loco regional or distant recurrence. So we decided to continue the monitoring.

Meta-iodobenzylguanidine (MIBG) scan was performed and no other associated or metastatic localizations of the paraganglioma were shown (Fig. 3).

Two control cystoscopies made after respectively, seven and ten months did not show any recurrence.

Discussion

Pheochromocytomas are neoplasms developed from the chromaffin tissues of the sympathetic nervous system. Commonly known arising from the largest aggregate of such tissues in the adrenal medulla, about 10% of pheochromocytomas can occur in the ectopic or extra adrenal sites and they are called in this case paragangliomas.[3]

The Pheochromocytoma of the bladder was first described by Zimmermann in 1953[1], and more than 170 cases have been spotted since then. The paraganglioma of the bladder is very rare and represents less than 6% of the whole range of paragangliomas and less than 1% of the bladder tumors.

The average age of the patients are relatively low, nearby 40 years, with a range going from 10 to 78 years, with high frequency during the adolescence and the thirties.

The most common associated symptoms with bladder paragangliomas are hypertension (54.7%)[5], headache (48.1%), hematuria (47.2%), and syncope/palpitations (43.4%). Micturition attacks, including headache, palpitations, syncope and visual disturbances, were reported in 52.8% of patients.

24 hours urinary and plasma metanephrine and catecholamine levels are measured for biochemical confirmation. Elevated urinary

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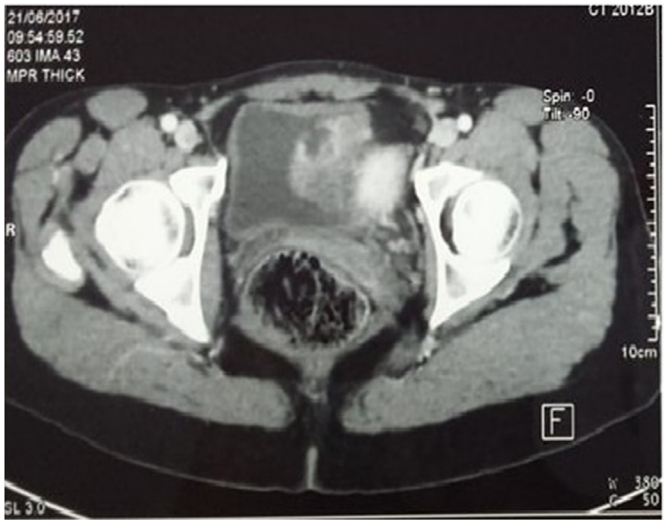

Fig. 1. CT scan showing a nodular calcified lesion, measuring 4 cm in the left lateral wall of the bladder.

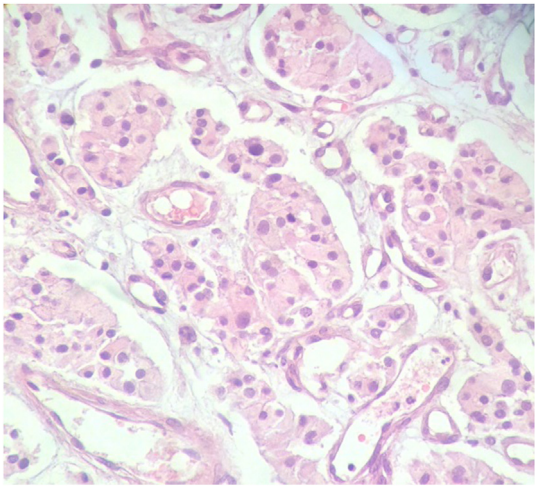

Fig. 2. HE x40: The tumor cells are round, polyhedral with eosinophilic cytoplasm.

metanephrine levels are both sensitive and specific for active paragangliomas, 96% and 98% respectively.

Imaging methods used in the investigation of paragangliomas include Ultrasonography and Computerized Tomography (CT), Magnetic Resonance Imaging (MRI), and Scintigraphy. Those examinations give also the opportunity to look for concomitant locations, as well as potential ganglionic or visceral metastasis.

For the detection of adrenal pheochromocytomas > 1.0 cm in diameter, CT and MRI have a sensitivity of nearly 95% and 100%, respectively, and MRI has greater specificity than does CT. On MRI, pheochromocytoma typically manifests as an expansive lesion with low signal intensity on T1-weighted sequences and high signal intensity on T2-weighted sequences, with intense impregnation after contrast administration. However, in rare cases, pheochromocytoma can present low signal intensity on T2-weighted sequences.

Histological examination of the tumor tissue will confirm diagnosis with diffuse, strong positivity for neuron-specific enolase (NSE), synaptophysin, and/or chromogranin.

The treatment of choice for paraganglioma is surgical resection, due to the multilayer involvement of the bladder wall.[2] The most commonly reported treatment for patients with paraganglioma of the urinarybladder was partial cystectomy.

Other treatment modalities include trans-urethral resection of bladder tumor (TURBT) and radical cystectomy.

External beam radiation therapy (ERBT) at > 40 gray in patients with paragangliomas has shown clinically significant symptomatic relief for at least one year or until death. For patients in which surgery or local radiation therapy is not viable, palliative system options include 131-iodine-labeled meta-iodobenzylguanidine (131I-MIBG) therapy and cytotoxic chemotherapy.[4]

In our case, the paraganglioma of the bladder was mimicking a typical urothelial bladder tumor with lower urinary tract symptoms but without hematuria. However, it had a calcified appearance at cystoscopy. The typical triad did not stand out. Biological cathecholamines measurement was normal, but performed after complete resection of the tumor.

During a ten-month follow-up, controls, made by two computerized tomography abdominal and pelvic scans and two cystoscopies, have not shown any kind of loco regional or distant recurrence.

This proves that complete and deep transurethral resection of bladder paragangliomas if well performed can be considered as an interesting less invasive first-line treatment with good outcomes. However a life-term clinical, biological and endoscopic monitoring

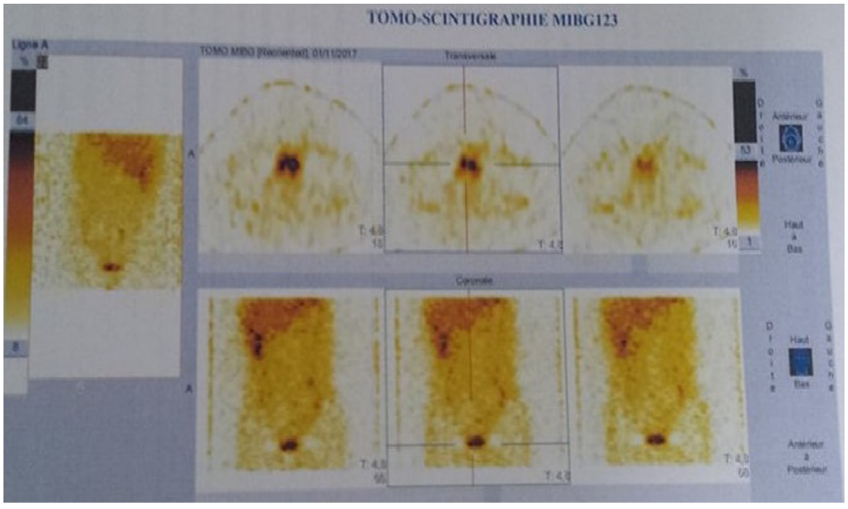

Fig. 3. Meta-iodobenzylguanidine (MIBG) scan.

should be done to decide whether or not re-resection should be performed.

Conclusion

Pheochromocytoma of the bladder is a rare tumor. Its diagnosis is usually suspected in the presence of paroxysmal hyperblood pressure paired with a bladder tumor. However, atypical paucisymptomatic forms exist. The partial or radical cystectomy remains the treatment of choice but other treatment modalities such transurethral resection can be considered as effective first-line treatments.

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