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Incidental Non-Functional Bladder Paraganglioma Discovered after Appendicitis: A Case Report

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Abstract | **Case Report**

Bladder paraganglioma is a rare neuroendocrine tumor arising from extra-adrenal chromaffin tissue and accounting for less than 0.1% of all bladder tumors, with non-functional forms often posing diagnostic challenges due to the absence of specific symptoms. We report the case of a 53-year-old male patient with no significant medical history who presented with febrile right iliac fossa pain and vomiting, without urinary or adrenergic manifestations. Abdominal ultrasound revealed acute appendicitis associated with an incidental bladder mass. Following appendectomy, the patient was referred for further evaluation. Clinical examination was unremarkable, and transurethral resection of the bladder tumor was performed without intraoperative hemodynamic instability. Histopathological and immunohistochemical analysis confirmed the diagnosis of bladder paraganglioma. Postoperative imaging showed no additional lesions, and biochemical evaluation for catecholamine secretion was within normal limits. Partial cystectomy was recommended but declined by the patient.

Keywords: Bladder paraganglioma; Neuroendocrine tumor; Non-functional tumor; Transurethral resection; Incidental finding; Urinary bladder tumor.

1. INTRODUCTION

Paragangliomas are rare tumors developed from extra-adrenal neuroendocrine tissue, originates from the neuroectodermal cells of the sympathetic nervous system, with one-third to one-half being located in the thoracoabdominal region [1,2].

Bladder paraganglioma are very rare, representing 0,006 % of all bladder tumors and 0,7% of extra adrenal paraganglioma [1].

They occur more in female patients than man ones and typically in the age from 20 to 50 years [1].

These tumors are most of the time benign and can be categorized in functional or non-functional types. 61,3% were reported to be functional and manifest in catecholamine release induced by miction and may include palpitation, headache, paroxysmal hypertension, sweating and even syncope [3].

There is no specific guideline for surgical treatment of bladder paraganglioma. About 14% of these tumors are malign and consequently radio and chemoresistants. Standard treatment for non-muscle invasive tumors are transurethral resection [TUR] and partial or radical cystectomy for muscle-invasive ones [4,5].

2. CASE REPORT

A 53 years old male patient with no comorbidities, chronic smoked weaned 30 years ago consulted for febrile right iliac fossa pain associated with vomiting with no urinary signs, notably no hematuria in the past.

Questioning revealed no objective urinary or adrenergic signs, and the patient reported no micturition.

Ultrasound findings revealed appendicitis and a tissue like bladder process.

He had an appendectomy and was referred to us for further treatment.

Clinically, the patient was in good general condition, with no clinical features, Fc: 85bpc, BP: 13/6

with a prostate of 30g, supple and painless on rectal examination.

Transurethral resection of the bladder was performed with no intra- nor postoperative events [no hypertension, tachycardia or heavy bleeding].

Anatomopathological study with immunohistochemistry of the resection revealed a vesical paraganglioma.

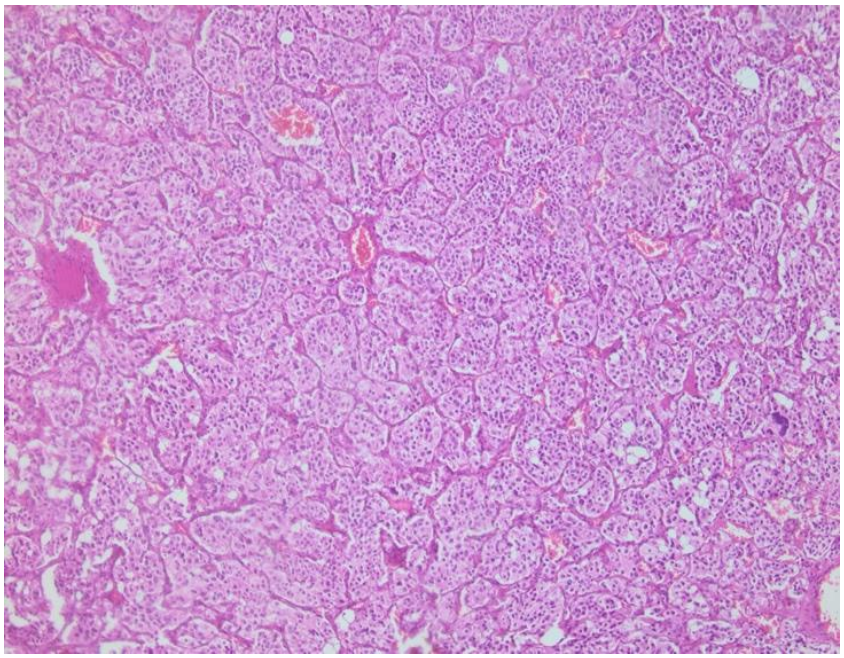

Figure 1: Tumoral proliferation arranged in nests [Zellballen], resting on a scant fibrovascular stroma [photo taken with standard staining, x100].

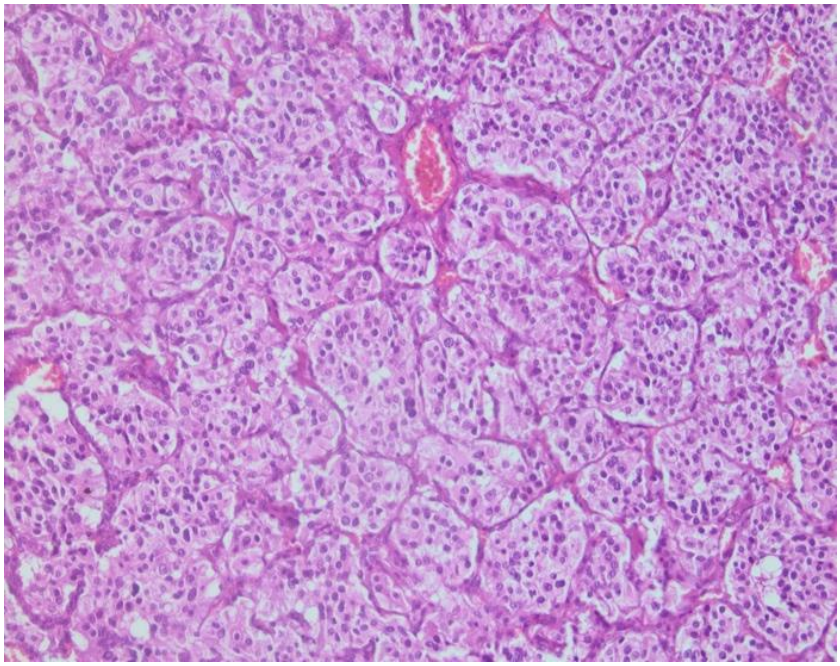

Figure 2: The tumor cells are polygonal, with abundant, eosinophilic, and cleared cytoplasm [image taken with standard staining, x200].

A thoracic-abdominal-pelvic CT scan was performed postoperatively to look for other localizations, and a urinary methoxylate derivative assay was performed.

The methoxylate derivatives came back normal, and the decision of a partial cystectomy was refused by the patient due to the absence of clinical symptoms.

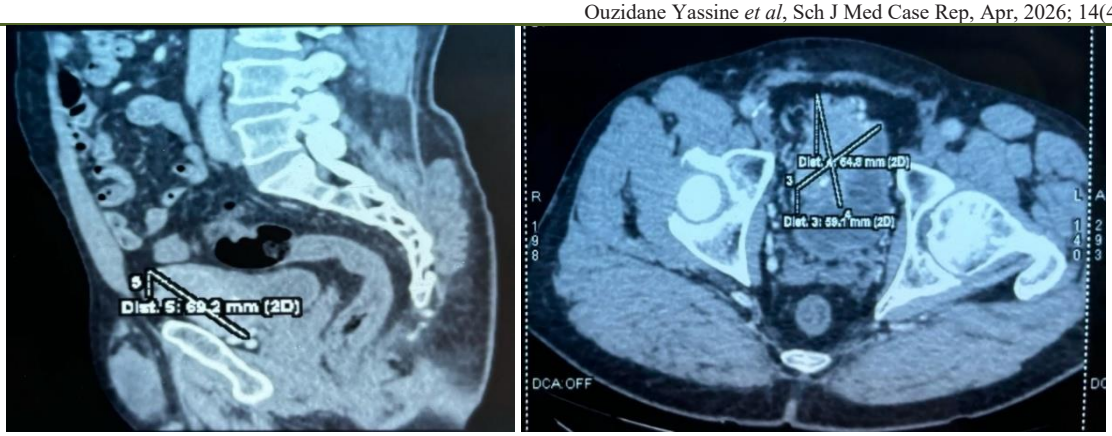

Figure 3: Scanner showing a tumor on the anterior wall

3. DISCUSSION

Paraganglioma originates from the chromaffin tissues of the autonomic nervous system, which is why it frequently develops along the sympathetic chain or in the organ of Zuckerkandl. It can be found in various locations, including the retroperitoneum, bladder, para-aortic region, and pelvic cavity [1].

Described the first time by Zimmerman in 1953, bladder paraganglioma is a very rare kind of neoplasm and can be often mistaken for more common bladder cancers, and it is critical to consider this diagnosis in patients presenting with hematuria and signs of catecholamine excess [3,5].

BPG may be functional or no depending on either catecholamine is over-secreted. Most of them are functional and the classic triad of clinical symptoms is silent hematuria, paroxysmal hypertension and micturition given the catecholamines secretion.

However, our patient didn't show any of those symptoms, that's why non-functional BPG can be easily misdiagnosed as bladder cancer before any surgical approach with only 28,9% of bladder paraganglioma diagnosed before surgery [1].

If the diagnosis is made preoperatively, a careful preparation is critical due to the tumor's ability to secrete catecholamines which can lead to several cardiovascular complications during surgical manipulation. Proper preparation focuses on controlling catecholamine-induced symptoms, especially hypertension, and minimizing the risk of intraoperative hypertensive crises. This involves careful alpha-blockade, potential beta-blockade for heart rate control, volume expansion, and close monitoring of blood pressure.

Both CT and MRI are effective tools for locating the primary tumor and identifying metastases. However, imaging with 131Iodine-metaiodobenzylguanidine [MIBG] has demonstrated superior sensitivity and specificity in detecting pheochromocytomas. Functional imaging, which targets the pathways involved in catecholamine synthesis, storage, and release, is particularly helpful in patients with paraganglioma, especially after surgery or when looking for metastases [6].

When not diagnosed preoperatively, the appearance of a yellow, submucosal tumor should suggest non urothelial kind of tumors [6].

Surgical resection is the most effective treatment for bladder paraganglioma, with options like transurethral resection, partial cystectomy, or total cystectomy. Partial cystectomy is often preferred due to the risks of hypertensive crisis and incomplete removal with transurethral resection. Transurethral resection may be suitable for small, non-functional tumors. Surgical intervention typically ensures long-term effectiveness, but close monitoring is recommended to detect potential recurrence. Treatment decisions should be individualized based on the patient's condition [1].

4. CONCLUSION

Bladder paraganglioma is a rare and potentially challenging diagnosis, particularly in non-functional forms that lack characteristic adrenergic symptoms. It should be considered in the differential diagnosis of bladder tumors, even in asymptomatic patients.

Accurate preoperative diagnosis is essential to prevent intraoperative complications and to guide appropriate management. Surgical resection remains the treatment of choice, and individualized management with long-term follow-up is crucial given the risk of recurrence and malignant potential.

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