



Pheochromocytoma of the prostate: An unusual location

Faten Hadj Kacem^a, Khouloud Boujelben^{a,*}, Wiem Feki^b, Khansa Chaabouni^c, Nadia Charfi^a, Mohamed Abid^a

^a Endocrinology Department, Academic Hospital Hedi Chaker, Sfax, Tunisia

^b Radiology Department, Academic Hospital Hedi Chaker, Sfax, Tunisia

^c Biochemistry Department, Academic Hospital Habib Bourguiba, Sfax, Tunisia

ARTICLE INFO

Keywords:

Pheochromocytoma
Ectopic
Paraganglioma
Prostate

ABSTRACT

Ectopic pheochromocytomas, also called paragangliomas, are defined as catecholamine-secreting tumors, which develop outside the adrenal medulla.

Pheochromocytomas of the urinary tract represent less than 1% of all paragangliomas and are most commonly located in the bladder. Nevertheless, prostatic pheochromocytoma is an extremely rare clinical entity and only a few cases have been reported in the medical literature.

Herein, we report a case of ectopic pheochromocytoma arising from the prostate, revealing with hypertensive crisis occurring immediately after ejaculation.

1. Introduction

Pheochromocytoma is a rare endocrine tumor that arises commonly from adrenal medulla. The annual prevalence ranges from 0,4/1 million to 9,5/1 million.¹ The rule of 'tens' is usually considered to describe pheochromocytomas' distribution and histology, with 10% malignant, 10% bilateral and 10% ectopic in origin.²

Ectopic pheochromocytoma is defined as pheochromocytoma occurring in extra-adrenal chromaffin tissues.

Due to the wide range of clinical presentations, diagnosis of ectopic pheochromocytoma can be challenging.

In fact, ectopic pheochromocytomas in the abdomen are commonly located at the aortic bifurcation or the inferior mesenteric artery where there are chromaffin tissues.²

Nevertheless, pheochromocytomas involving the prostate are extremely rare with no more than 15 cases reported in the medical literature.

2. Case presentation

A 27-year-old man presented to our department with palpitations, headaches and sweating that occurred immediately after ejaculation for more than two years.

He additionally complained of intermittent dysuria for several years. His past history included also paroxysmal hypertension treated for the

previous eight years.

Physical examination showed a heart rate of 124 beats/min and blood pressure of 160/80 mmHg. The patient was found to have a rubbery enlarged prostate on digital rectal examination but no nodules were palpable.

The serum level of prostate-specific antigen (PSA) was 0,31 ng/ml (normal range: <4 ng/ml).

Twenty four-hour urinary catecholamines measured after a single ejaculation subsequent to masturbation were significantly elevated at 37, 69 μmol/24 hours (normal range: <5,5 μmol/24 hours).

Examination of endocrine tumor biomarkers revealed that calcitonin and chromogranin A were within normal ranges.

Pelvic ultrasound showed a well-defined hypoechoic mass arising from the right lobe of the prostate measuring 12 × 14 mm (Fig. 1).

Contrast enhanced computed tomography (CT) scan of abdomen and pelvis didn't detect any adrenal lesion but revealed a 16 × 14 mm heterogeneously-enhanced mass located in the right lobe of the prostate (Fig. 2). There were no abdominal or pelvic pathological lymph nodes.

The body scintigraphy with iodine-123 metaiodobenzylguanidine (MIBG) demonstrated an increased uptake at the same lesion and ruled out the presence of multifocal lesion or metastases (Fig. 3).

Considering the patient's clinical manifestations including Menard triad (palpitations, headaches and profuse sweating), hypertension at an early age, as well as an increased catecholamine level and imaging findings, the diagnosis of ectopic prostatic pheochromocytoma was

* Corresponding author. Endocrinology Department, Academic Hospital Hedi Chaker, Route El Ain Km 0.5, 3000, Sfax, Tunisia.

E-mail address: khouloud25boujelben@gmail.com (K. Boujelben).

<https://doi.org/10.1016/j.eucr.2022.102108>

Received 28 March 2022; Received in revised form 29 April 2022; Accepted 4 May 2022

Available online 7 May 2022

2214-4420/© 2022 Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



Fig. 1. Ultrasound image revealing a hypoechoic mass located in the right lobe of the prostate.

made. Thus, a radical prostatectomy was considered but the patient declined any surgical options.

3. Discussion

Ectopic pheochromocytomas, also called paragangliomas, are slow growing neoplasms derived from the paraganglion tissue. They are uncommon in adults, accounting for 10% of all chromaffin cell-derived neuroendocrine tumors.²

As extra adrenal pheochromocytomas arise from the chromaffin cells, the majority of these tumors are abdominal. Of these last, about 90% develop in the para-aortic area, as well as at the Zuckerkandl organ.²

Pheochromocytomas of the urinary tract represent less than 1% of all paragangliomas and are most commonly located in the bladder.³ However, prostatic pheochromocytoma is rare with only 15 cases reported in the medical literature.

Clinical presentations of paragangliomas are various, depending on the size and location of the tumor, as well as its functional status.

In the case of prostatic pheochromocytoma, the most common symptoms include painful urination, hematuria, hemospermia and difficult defecation.² If the tumor is functional, other clinical



Fig. 2. Contrast enhanced CT scan showing a heterogeneously enhanced mass located in the right lobe of the prostate.

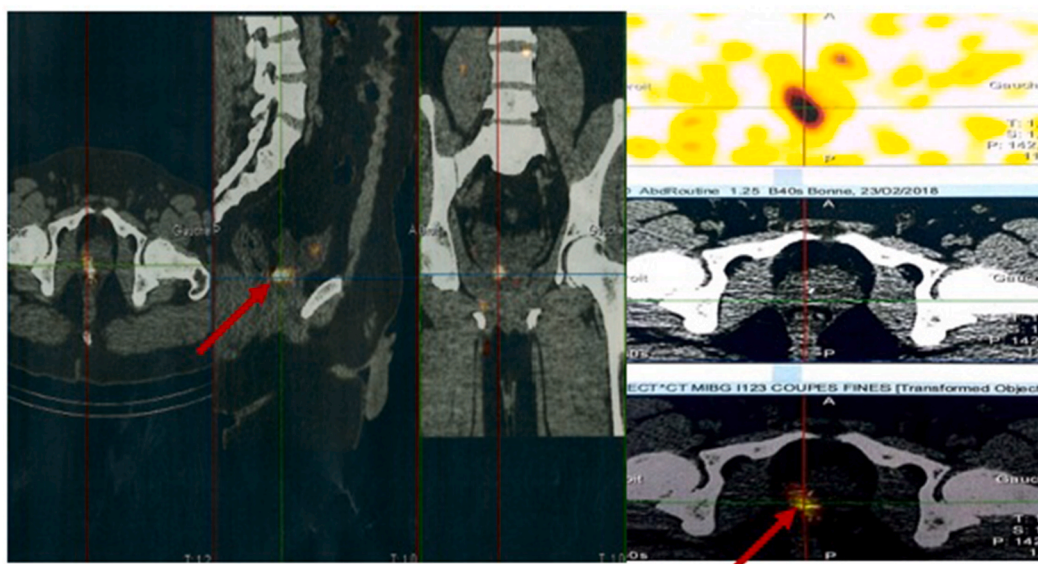


Fig. 3. MIBG scintiscan showing an increased uptake at the right lobe of the prostate.

manifestations resulting from catecholamine overproduction may include palpitations, sweating, headaches, anxiety and hypertensive crisis triggered by defecation, micturition or rectal examination³

In the current case, the chief complaint was hypertensive crisis occurring immediately after ejaculation.

Confirmation of pheochromocytoma is accomplished through measurement of plasma-free catecholamines or its derivatives in serum or urine of 24 hours.

Catecholamines 4-fold above the upper reference limit confirm pheochromocytoma with high sensitivity.⁴ Our patient had 6-fold above the upper reference urinary methoxylated derivatives.

For imaging diagnosis, CT, magnetic resonance imaging (MRI) and metaiodobenzylguanidine (MIBG) scintigraphy have sensitivity of 90%, 93%, and 91%, respectively⁴

Thus, scintigraphy-using iodine 123 or 131 labeled MIBG has a specificity of nearly 100% as regards to characterizing a lesion detected by CT or MRI. It is also performed to reveal multifocal lesions or secondary sites of a malignant tumor.²

The presence of metastases at non-chromaffin sites is the only indication of malignancy. Usual locations of metastases are lung, bone and lymph nodes. Metastases to lymph nodes in prostatic pheochromocytoma have been reported in 5 cases. Metastases to other sites have not been described.

Because of its rarity, the most common differential diagnosis for prostatic pheochromocytoma is adenocarcinoma. Negative results in immunohistochemical evaluation for prostate-specific antigen (PSA) and/or prostate acid phosphatase as well as elevated catecholamines levels are evident differential diagnostic criteria.⁵

After making the definite diagnosis and as in adrenal pheochromocytomas, the main treatment in the case of isolated and solitary prostatic pheochromocytoma is the complete surgical removal⁴

As pheochromocytoma's manipulation may result in excessive catecholamines release leading to hypertensive crises, its surgical excision is associated with a significant rate of morbidity and mortality. Thus, preoperative preparation with optimal adrenergic blockage and volume expansion is crucial in the management of pheochromocytoma⁵

Different operative approaches for the treatment of ectopic prostate pheochromocytoma may be considered such as transurethral resection of the prostate, radical prostatectomy or adding pelvic lymphadenectomy. In addition, chemotherapy, as well as palliative radiotherapy,

may be recommended for patients with metastatic disease⁴

Because there were no evidence of metastatic tumor in our patient, radical prostatectomy was recommended, but refused by the patient.

4. Conclusion

The present case illustrates a rare condition of prostatic pheochromocytoma. Clinicians should be aware that the prostate is uncommon but possible location of ectopic pheochromocytoma. Because of the nonspecific clinical signs and the insidious course in many cases, the diagnosis of the tumor remains challenging.

The first-line treatment for ectopic pheochromocytoma is surgical excision of the tumor. Thus far, the prognosis of prostatic pheochromocytoma is unrevealed because of the lack of valid parameters and the small number of cases in the medical literature.

Early diagnosis and close monitoring are therefore essential for recognizing paraganglioma recurrence.

Consent

Consent of the patient for publishing was obtained.

Declaration of competing interest

No conflict of interest to be noted.

References

1. Fishbein L. Pheochromocytoma and paraganglioma: genetics, diagnosis, and treatment. *Hematol Oncol Clin North Am.* févr. 2016;30(1):135–150.
2. Zhou J, Wu W-F, Zhang W, Xin J, Lei W-H. Successful transurethral resection of the prostate in ectopic prostate pheochromocytoma: a case report. *Medicine (Baltimore).* AVR. 2020;99(17), e19852.
3. Chan VSH, Chan DLH, Hui SY, Lam WWM. Occult extra-adrenal pheochromocytoma in the urinary bladder. *BMJ Case Rep.* 8 Mar 2019;12(3), e229267.
4. Worrest TC, Gilbert EW, Sheppard BC. Pheochromocytoma: 20 years of improving surgical care. *Am J Surg.* Mar 2019;217(5):967–969.
5. Liu H, Li B, Yu X, Huang Y. Perioperative management during laparoscopic resection of large pheochromocytomas: a single-institution retrospective study. *J Surg Oncol Suppl.* Sept 2018;118(4):709–715.