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CASE REPORT

Intra Renal Paraganglioma: Exceptional Localization

Yddoussalah O, Saouli A, Karmouni T, Elkhader K, Koutani A, Ibn Attya Andaloussi A

Department of Urology B, Ibn Sina University hospital, Faculty of Medicine and Pharmacy, Mohamed V University, Rabat, Morocco.

Abstract

Introduction: Intra renal paraganglioma are exceptional tumors. A thorough literature review found only twelve additional cases of intrarenal paraganglioma reported up to date.

Case presentation: The authors report the case of a paraganglioma intra discovered on renal nephrectomy piece performed on a right kidney destroyed by pelvic calculi.

Conclusion : The treatment of these tumors requires a more complete surgical resection possible since the prognosis depends. There exists against no consensus on the usefulness of complementary therapies, which nevertheless may be symptomatic title in extra. Through this observation we insist on the clinical, paraclinical and therapeutic characteristics of this anatomo-pathological entity.

Keywords: Intrarénal, Nephrectomy, Paraganglioma.

Introduction

Paragangliomas are extra-adrenal pheochromoctomas found along sympathetic and parasympathetic chain, The adrenal seat is usual (90%), the extrasurrenal location is rare, representing 10% of the paragangliomas with an incidence rate of 2-8 cases per million persons / year [1]. They have a variable topography and functional retroperitoneal form accounts for 2% of cases [2]. Renal paragangliomas are not only rarely encountered, but also difficult to distinguish clinically and pathologically from renal cell carcinoma. We report a case, diagnosed by the histological study of the surgical specimen.

Case Presentation

The patient is aged 62 years. Clinical history of this woman began with abdominal pain in the right side with two episodes of renal colic right. Abdomen was flexible.

The CT scan, performed before and after injection of contrast material, showed a right kidney destroyed with a parenchyma lamine upstream from a calculi staghorn renal pelvis and lower caliceal (Figure 1), a suspicious top pole right renal lesion was identified which Takes the contrast (Figure 2).

Additional biological assays had eliminated unusual hormonal secretion.

Given the negativity of the balance and the destroyed aspect of the right kidney, surgery was performed by left lobotomy. A total right nephrectomy is then performed. The operative sequences were simple.

A macroscopic examination, there is a nephrectomy piece weighing 70g.avec has cutting there is a dilation of

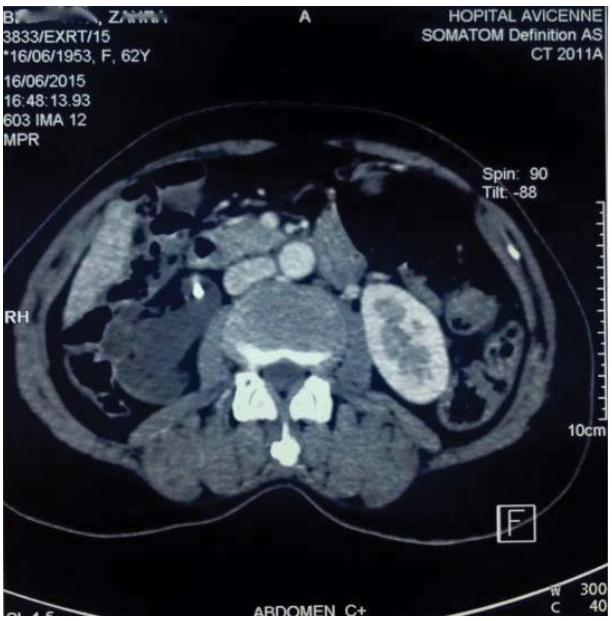

Figure 1: CT scan showing a right kidney seat of a huge DPC, upstream of a lithiasis image.

Pyellocalicious cavities and the presence of several stones coraliformes. We notice the presence of a nodule under whitish capsular measuring 1x0, 5cm, located At the upper polar level.

Correspondence to: Othmane Yddoussalah, residence Wifaq building 2 apartments 7 -Hay Riad- rabat. Tél: 0021268517870 Email: yddoussalah[DOT]urob[AT]gmail[DOT]com

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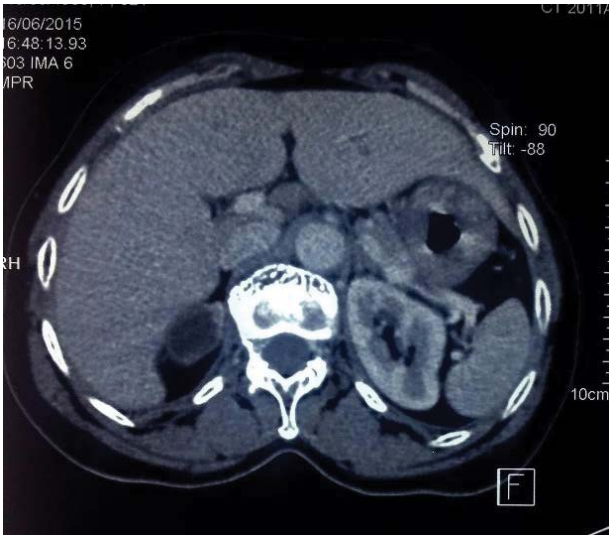

Figure 2: CT scan showing a nodule higher polar tissue with contrast uptake.

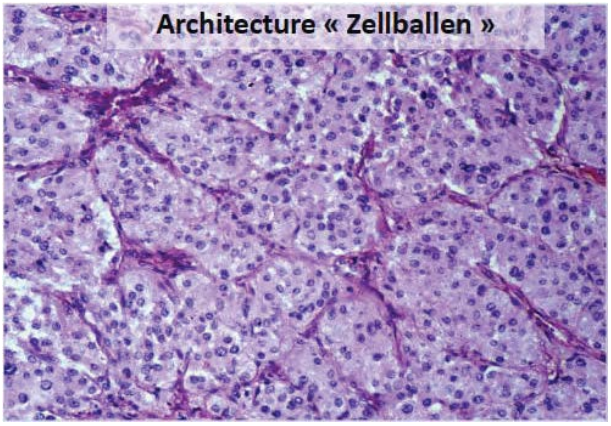

Figure 3: Histological aspect of an endocrine tumor, realizing an aspect of zellballen.

The histological examination, the cuts analyzed in the kidney shows an aspect of non-specific chronic pyelonephritis. The cuts made at the upper polar nodule, show a tumor proliferation made of cord, and nests spans supported by fusiform addition sprawling cell performing one aspect of zellballen within a dense stroma hyalinized and richly vascularized. the tumor cells have pleomorphic nuclei and eosinophilic cytoplasm. Mitosis is rare (Figure 3).

This proliferation contains foci of haemorrhagic and fibrous remodeling. There has been no evidence of tumor vascular embolism.

This morphological analysis concluded a Paraganglioma at the upper polar nodule.

Clinical and paraclinic evolution is favorable with a follow-up of 24 months, the patient is still alive.

Discussion

Heterotopic adrenal tissue or adrenal rests have been reported in various locations, such as testis and related structures, female genital tract and thorax. In kidneys it is encountered more frequently than is generally appreciated. Usually it is located in sub capsular region. Only adrenal cortical tissue or both cortical and medullar tissue may be present [3]. Like in our case, many of the reported cases of renal paragangliomas were not associated with hypertension or other symptoms of catecholamine hypersecretion and were labeled as nonsecretory. However, determination of urine catecholamines was not performed in most cases. None of the reported cases was diagnosed as or suspected to be paraganglioma or pheochromocytoma preoperatively. The presence of hypertension, especially when encountered in a younger patient or associated with paroxysmal headaches and palpitations, may raise the suspicion of paraganglioma or pheochromocytoma; however, in the presence of a mute kidney a Renovascular Hypertension can explain hypertension arterial.

The gross and microscopic appearance of paragangliomas is practically the same regardless of the location

And indistinguishable from that of adrenal pheochromocytoma. Well-defined nests of cuboidal cells ("Zellballen") are separated by highly vascularized fibrous septa. The individual cells have a moderately abundant granular cytoplasm and mitoses are exceptional. Ultra structurally, the tissue cells contain large number of cytoplasmic neurosecretory granules whose appearance is similar to those seen in normal paraganglia [4]. A second cell type is the sustentacular cell, which wraps around the chief cells and lacks dense-core granules.

Application of the Pheochromocytoma of the Adrenal Gland Scaled Score PASS scoring system that was proposed to separate benign from malignant pheochromocytomas showed a score of <4, which also suggested a benign tumor [5]. Nonetheless, histology is an imperfect tool to predict the behavior of paragangliomas, as there are no absolute criteria of malignancy apart from the presence of invasion andmetastasis, and no paraganglioma or pheochromocytoma can be considered unequivocally benign [6].

Our observation perfectly summarizes the main characteristics of paragangliomas. Their evolution is slow, pauci-symptomatic. Their discovery is often fortuitous or belated. In our case, the elements of the clinic and the results of imaging were in favor of a kidney destroyed by a lithiasic pathology. But the results of immunohistochemical and histological examination were characteristic of an intrarenal paraganglioma. Paragangliomas can take many aspects in imaging [7].

Regardless of the anatomic location of the paragangliomas the treatment of choice is complete surgical excision of the

primary as well as recurrent or metastatic disease [8,9]. In the present patient tumor was completely excised & at two-year follow-up no signs of recurrence have appeared. If the diagnosis is performed intraoperatively a preparation of the patient is obligatory. Recently a case of robotic partial nephrectomy for an intra renal paraganglioma was reported by an American team [10].

A thorough literature review found only twelve additional cases of intrarenal paraganglioma reported up to date,

Occurring in 6 men and 7 women with a mean age of 42.6 years. In the older literature, some of these cases were

Reported as “renal pheochromocytoma” in accordance with the nomenclature used at the time. The exact mechanism

By which paragangliomas occur in the kidney is unknown; one theory suggests an origin in ectopic adrenal tissue or

Adrenal rests located in the kidney [11]. Ectopic adrenal tissue is caused by multiple adrenal primordia or from fragmentation of the adrenal primordial during embryogenesis; the more commonly found ectopic adrenal rests that migrate with the developing gonads consist only of cortical tissue, while the ectopic adrenal rests situated close to the original position of the adrenal gland may also contain medulla. Another possible explanation suggested for the occurrence of intrarénal paragangliomas is renal-adrenal fusion, in which the heterotopic adrenal gland is entirely contained within the renal capsule. However, such cases show only adrenal cortical tissue and lack adrenal medulla [12]. In most of the reported cases of renal paraganglioma, the ipsilateral adrenal gland was identified within the nephrectomy specimen and was morphologically unremarkable.

Renal paragangliomas have been reported to arise in the upper pole or hilum or, as in the case presented herein, from the lower pole of the kidney. The location of the tumors within the kidney appears to be an important determinant of the patients’ clinical presentation. Tumors located in the renal pelvis/hilum have been reported to cause hypertension renal artery stenosis [8]. The reported sizes of the tumors varied from 2.5 to 18 cm; larger tumors were cystic. The size of these tumors is similar to the size of renal cell carcinomas. The relatively large size of the tumors may be ascribed to their lack of symptoms, since even small paragangliomas (and pheochromocytomas) can be detected early if they cause symptoms like palpitations, sweating, and headaches due to hormone hypersecretion.

Conclusion

The present case appears to be the thirteenth case of intra-renal paraganglioma in English literature. Particular

Feature of the present intra-renal paraganglioma is the appearance of destroyed kidney, which is in contrast to other

Reported cases of intra-renal paragangliomas and led to difficulty in accurate preoperative diagnosis. Possibility of this rare condition should be kept in mind.

Consent of patients

Written informed consent was obtained from the patient’s next of kin for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing Interests

The authors declare that they have no competing interests.

Authors’ Contributions

O. Yddoussalah wrote the manuscript. All authors read and approved the final manuscript.

Acknowledgement

NA

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