Scholars Journal of Medical Case Reports (SJMCR) **ISSN 2347-6559 (Online)**
Abbreviated Key Title: Sch. J. Med. Case Rep. **ISSN 2347-9507 (Print)**

A United of Scholars Academic and Scientific Society, India

Retroperitoneal Paraganglioma in Children (About a Rare Case)

Driss Hanine*, Meryam Ramzi, Zakaria Aboulam, Rachid Oulahyane, Mounir Kisra
Visceral Pediatric Surgery Department "A" – Children Hospital of Rabat – Morocco

***Corresponding author**
Driss Hanine

Article History
Received: 14.10.2018
Accepted: 23.10.2018
Published:30.10.2018

DOI:
10.36347/sjmcr.2018.v06i10.025

Abstract: Retroperitoneal paragangliomas are rare tumors. They are defined as extra-adrenal chromaffin tumors and represent approximately 1 / 5th of chromaffin tumors. They are often asymptomatic and can reach important dimensions. We report the case of a 7-year-old boy operated on for a giant retroperitoneal tumor whose pathological examination concluded that he had a paraganglioma. The malignant forms, more frequent than the benign forms, present a locoregional invasion and metastasize late. Management of paragangliomas should be multidisciplinary but only surgical treatment is curative. On the other hand, there is no consensus on the usefulness of complementary therapies, which may nonetheless be supportive as a symptom. Paragangliomas are genetic in 25% of cases. A genetic survey must be systematically proposed.
Keywords: retroperitoneal tumor, paraganglioma, pheochromocytome, child.

INTRODUCTION

The paragangliomas, or extra-adrenal pheochromocytomas, are neuroendocrine tumors developed at the expense of the parasympathetic nervous system [1]. They are defined as extra-adrenal chromaffin tumors and represent approximately 1 / 5th of chromaffin tumors. Non-functional retroperitoneal paragangliomas are rare tumors [2,3] and are less frequent than other locations (head, neck) [2].

They are often asymptomatic and can reach important dimensions. Management of paragangliomas should be multidisciplinary but only surgical treatment is curative. Through an observation of a child having had an extra-adrenal tumor with arterial hypertension, we take stock of this pathology as well as the various diagnostic and therapeutic problems.

MATERIALS & METHODS

This is a 7 year old boy from a sibling of 3 non-consanguineous parents and no particular pathological background. The story of his illness goes back to 1 month before admission by the installation of headaches, profuse sweating, abdominal pain with vomiting and constipation, all evolving in a context of apyrexia and conservation of the general condition. The clinical examination showed a mass of the right hypochondrium with arterial hypertension (with tensionel numbers ranging from 120 to 170 mmHg for the systolic pressure and from 90 to 100 mmHg for the diastolic pressure).

A first abdominal ultrasound was performed, showing a mass under right hepatic, within and slightly above the right renal hile, iso echogenously discrete heterogeneous non-calcified or necrotic, ovoid, fairly well limited 42 x 33 x 40 mm , surrounded by a fine hypoechoic, avascular line distorting the sinus of the right kidney which seems harmoniously pushed back out, seeming to laminate the inferior vena cava behind, and coming in to the contact with the pancreatic head (Figure 1).

A complement by abdominal CT was subsequently performed, objectifying a mass, intimate hilar contact anterior-internal right kidney, isodense, homogeneous, not raised after injection of contrast, non-adrenal evoking a pheochromocytoma or paraganglioma (Figure 2). Biological assessments showed iron deficiency anemia at 9.3 and elevated urinary catecholamines (metanephrine and normetanephrine).

The rest of the biological assessment and imaging were normal. Patient is initially on antihypertensive treatment based on calcium channel blockers in the absence of alpha-blockers that were unavailable. After multidisciplinary staff bringing together pediatric surgeons, pediatricians, pediatric oncologists, radiologists and anatomopathologists, we decided to start with a first surgery.

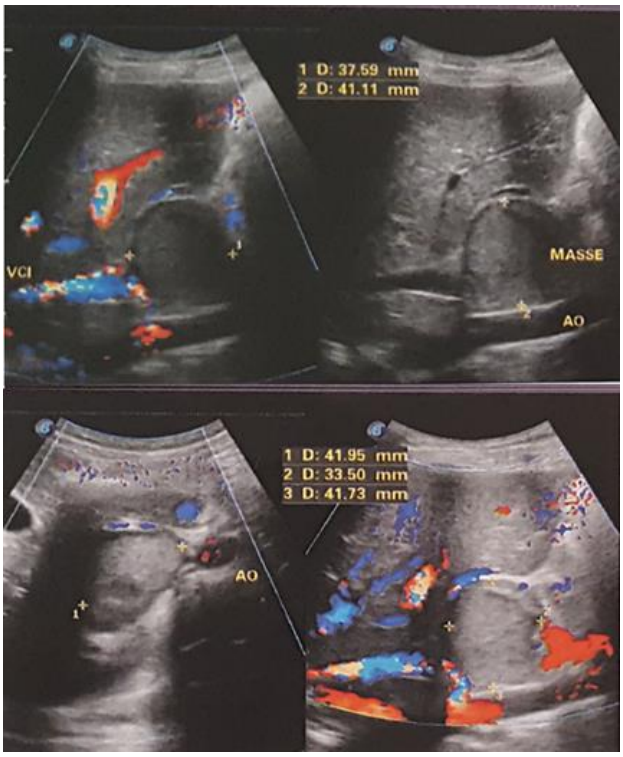

Fig-1: Doppler-coupled ultrasound image showing mass at right renal hile

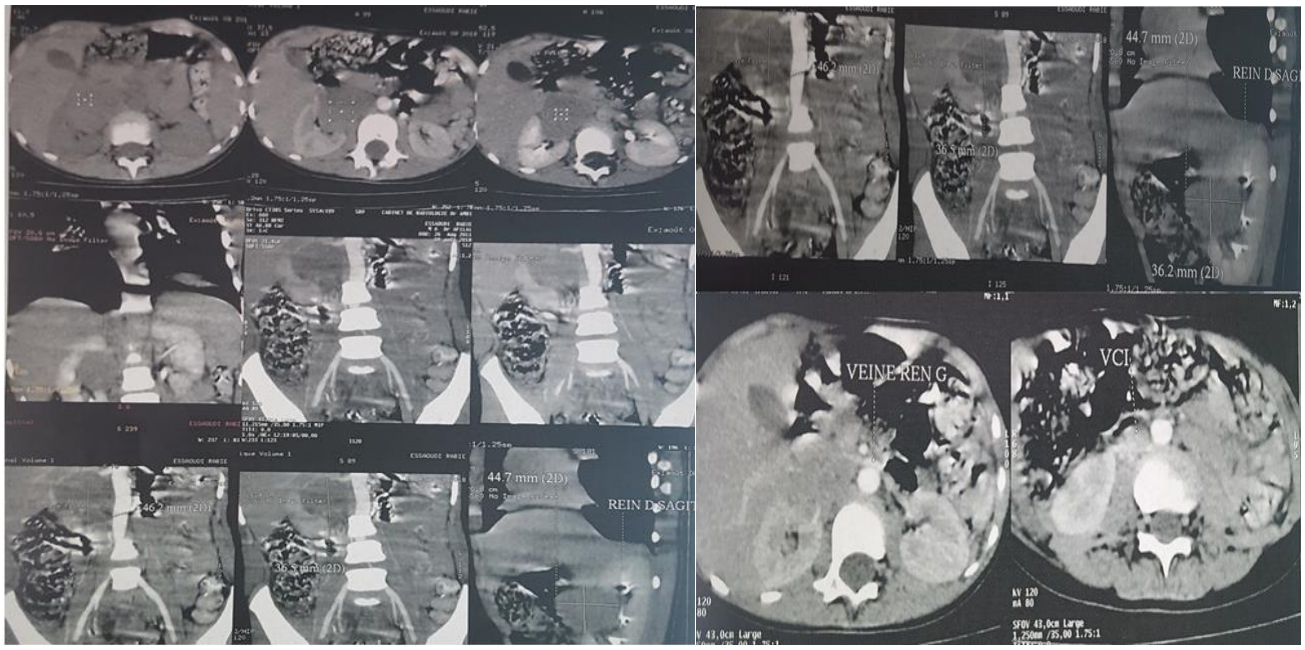

Fig-2: Scanning image objectifying the mass as well as its various reports

RESULTS

After right transverse umbilical incision, the surgical exploration showed a mass in the right renal hile, including the renal pedicle (renal artery in total and half of the renal vein) and half of the inferior vena cava. The adrenal is intact. The mass having multiple vascular connections with the aorta, IVC and renal pedicle, a careful dissection of the mass after section ligation of these vascular connections was performed. Complete resection of the mass and biopsy of part of the adrenal gland.

The anatomopathological study concluded in a retroperitoneal paraganglioma secreting. Immediate postoperative outcomes noted some episodes of hypotension and then normalization of blood pressure in the medium and long term. Catecholamine dosages have normalized.

DISCUSSION

RP Tumors are classified according to their embryonic origin, so paragangliomas are ectodermal tumors of neurogenic origin and are defined as extra-adrenal chromaffin tumors. The extra-adrenal pheochromocytomas or paragangliomas represent about 1 / 5th of the chromaffin tumors and are a rare entity and are secreting in 60% of cases [4]. Embryologically, chromaffin cells extend from the base of the skull to the epididymis. Most involute; and only those of the adrenal gland and retroperitoneal and thoracic ganglionic chains persist. Extra-adrenal pheochromocytomas or paragangliomas account for 18% of chromaffin tumors. They occur at an earlier age, 10 to 30 years, but more common in young adults [4]. They are multifocal in 15 to 24% of cases.

They are much more often malignant than intra-adrenal pheochromocytomas about 40% against 10%. The malignant forms occur earlier than the benign forms and are characterized by the occurrence of a local invasion (lymphatic type) or at a distance (invasion of the lung, bone, liver) in 30% of cases [3,4]. They secrete predominantly norepinephrine, often exclusively. Their origin is genetic, which corresponds well to their multicentricity and metachronous recurrences in other sites. Only 1% of paragangliomas in the head and neck secrete catecholamines while those with thoracic, abdominal, retroperitoneal or pelvic seals secrete more often [5]. Eleven cases of pulmonary paragangliomas have been published. Finally, only two cases of paragangliomas or pheochromocytomas originally intrahepatic have been described. Pheochromocytoma does not only appear to be a tumor of the adrenal gland, but rather a disease of the chromaffin system. The genetic origin is evoked more when the tumors are multiple, while the malignancy is even higher when the tumor is ectopic.

Retro-peritoneal forms are most often isolated [3,4]. They are sometimes associated with other pathologies, notably Carney's triad [6], multiple endocrine neoplasia type 2 [3] and neurofibromatosis type 1. Non-functional retroperitoneal paragangliomas are characterized by their asymptomatic appearance (absence of HTA) and normal levels of blood and urine catecholamines [3] and clinical latency most often [4], sometimes non-specific signs are found [4]. The preoperative positive diagnosis is biological [7].

Abdominal ultrasound notes a well-defined, solid oval mass with many central cystic formations [3,6]. The abdominal CT shows the characteristics of the tumor including its retroperitoneal seat, its size, its single or multiple character with search for locoregional invasion and distance, the most often found aspect is that of a round or oval solid mass, homogeneous, but can be cystic or necrotic in its center or calcified [3,8,9] whereas MRI is the examination of choice for the diagnosis and the morphological assessment of the lesions, for the CT it is equivalent of the MRI for the morphological assessment but anyway its effectiveness is less.

Meta-iodo-benzyl-guanidine scintigraphy has little value preoperatively, but occupies a major place in operative monitoring. The diagnosis of certainty is histological [3,4], with an appearance of an enormous rounded tumor, encapsulated with a firm, elastic, highly vascularized consistency, but it is immunohistochemistry that makes it possible to affirm the diagnosis, on the other hand there is no histological criterion to distinguish between benignity and malignancy of the tumor [8]. A good management of paraganglioma requires a precise morphological assessment, given the vascular complexity of these tumors hence the interest to be based on the angio-scanner. Radical surgery is the basis of treatment with radical resection in 75% of cases [4].

Complementary therapies such as chemotherapy and external radiotherapy may find their place in metastatic forms with a positive response in about 50% of cases, but without significantly influencing the prognosis, only surgical excision allows a significant improvement, with a rate survival without recurrence of 75% at 5 years and 45% at 10 years [4]. The average survival is of the order of 3 years in the metastatic forms and 4 years in the case of incomplete excision [4].

CONCLUSION

The paragangliomas, or extra-adrenal pheochromocytomas, are neuroendocrine tumors developed at the expense of the parasympathetic nervous system. Non-functional retroperitoneal paragangliomas are rare tumors. They are often asymptomatic and can reach important dimensions The management of paragangliomas must be multidisciplinary but only the surgical treatment is curative Complementary therapies, like chemotherapy, external radiotherapy could find their place in the metastatic forms but without influencing the prognosis significant.

REFERENCES

1. Cavero Rebollo O, Urruchi Fernandez P, Cay Diarte A, Gil Sanz MJ, Gonzalez Enguita C, Rioja Sanz LLA. Non functioning retroperitoneal paraganglioma. Actas Urol Esp. 1991; 15(2): 181-184.
2. Crozier F, Lechevallier E, Eghazarian C, Andre M, Sammama D, Wilshire P, Vidal V, Pascal P, Bartoli JM. Paragangliome non sécrétant rétropéritonéal. Journal de radiologie. 1999;80(2):150-2.
3. Giudicelli T, Bruneton JN, Duplay H, Abbes M, Balu Maestro C, Raffaelli C, Rogopoulos A, Bittmann O. Imagerie des paragangliomes rétrpéritonéaux non fonctionnels: à propos d'un cas. J Radiol. 1991; 72(11): 617-619.

4. Farthouat P, Platal JP, Meusnier F, Pourrire M, Thouard H. Paragangliome rétropéritonéal sécrétant: à propos d'un cas. J Chir.1997; 134 (5-6): 248-251.
5. Fernandez Cruz L, Taura P, Saenz A, Benarroch G, Sabater L. Laparoscopic approach to pheochromcytoma: hemodynamic changes and catecholamine secretion. World J Surg. 1996; 20(7): 762- 768.
6. Le Blanc I, Laurent M, Bokobza B, Michot F, Teniere P. Triade de Carney: un nouveau cas associé à un adénome cortico surrénalien. Gastroenterol Clin Biol. 1990; 14(4): 399- 401.
7. Hayes WS, Davidson AJ, Grimley PM, Hartman DS. Extraadrenal retroperitoneal paraganglioma: clinical, pathologic and CT finding AJR Am. J Roentgenol. 1990; 155 (6): 1247- 1250.
8. Pagliano G, Michel PH, La Fay Th, Duverger V. Paragangliomes de l'organe de Zuckerkandl. Chirurgie. 1994-1995; 120(3) : 128-133.
9. Pfister CH, Scotte M, Sibert L, Meunier Y, Kuhn JM, Michot F, Teniere P. Tumeur chromaffine ectopique sous hépatique non sécrétante. Ann Chi. 1995; 49 (10): 963- 967.