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Retroperitoneal Ganglioneuroma in Young Adults, Report of A Case

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Abstract **Case Report**

Retroperitoneal ganglioneuroma is a benign tumor of the sympathetic nervous system that is part of the neuro-cristopathies and whose pathogenesis remains even less well known. It is a rare tumor, it constitutes 0.07 to 0.2% of tumors. In Morocco, 3 cases of ganglioneuroma have been published. The peak frequency is between 20 and 30 years old with a slight female predominance. It is an often asymptomatic tumor falling within the framework of fortuitous diseases and whose diagnosis is based on radiological investigations, in particular ultrasound, CT and MRI. The histological diagnosis is based on the anatomo-pathological study after surgical excision.

Keywords: Ganglioneurone, benign tumor, abdominal MRI, surgical treatment.

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INTRODUCTION

Ganglioneuroma is a benign nerve tumor in children and young adults. It is a rare tumor, located in the adrenal gland (20%), along the sympathetic chain and particularly in the posterior mediastinum (40%) and retroperitoneum (30%) [1]. It belongs to the group of neurogenic tumors, developing at the expense of the sympathetic ganglion chains, a group which also includes ganglioneuroblastomas and neuroblastomas [2]. Indeed, these pose a problem of clinical and histological diagnosis, as well as a therapeutic problem because of the relationships with neighboring organs, and in particular with the large vessels (inferior vena cava, aorta).

RESULTS

23-year-old patient followed for hemorrhoids. A patient presented for heaviness of the right hypochondrium with a palpable mass. Physical examination was normal in our patient, except for a palpable abdominal mass.

Abdominal MRI: individualization of a lesion process lateralized to the right, well limited, with regular contours, in clear T1 hypo signal, heterogeneous hyper signal T2 hyper signal diffusion decreasing with the value of b, slightly enhanced and heterogeneously after injection of glonium Figure 1.

The surgical intervention was carried out through a median incision which discovered a 20*15 cm retroperitoneal cystic mass adhering to the left renal vein, the aorta and the adrenal gland and right kidney without carcinomatosis, ascites or hepatic metastasis.

The post-operative aftermath was simple. The logical anatomo-pathological study of the surgical specimen (Figure 2) finds a tumor of firm and slightly yellowish and adipose consistency weighing 720 grams (Figure 2).

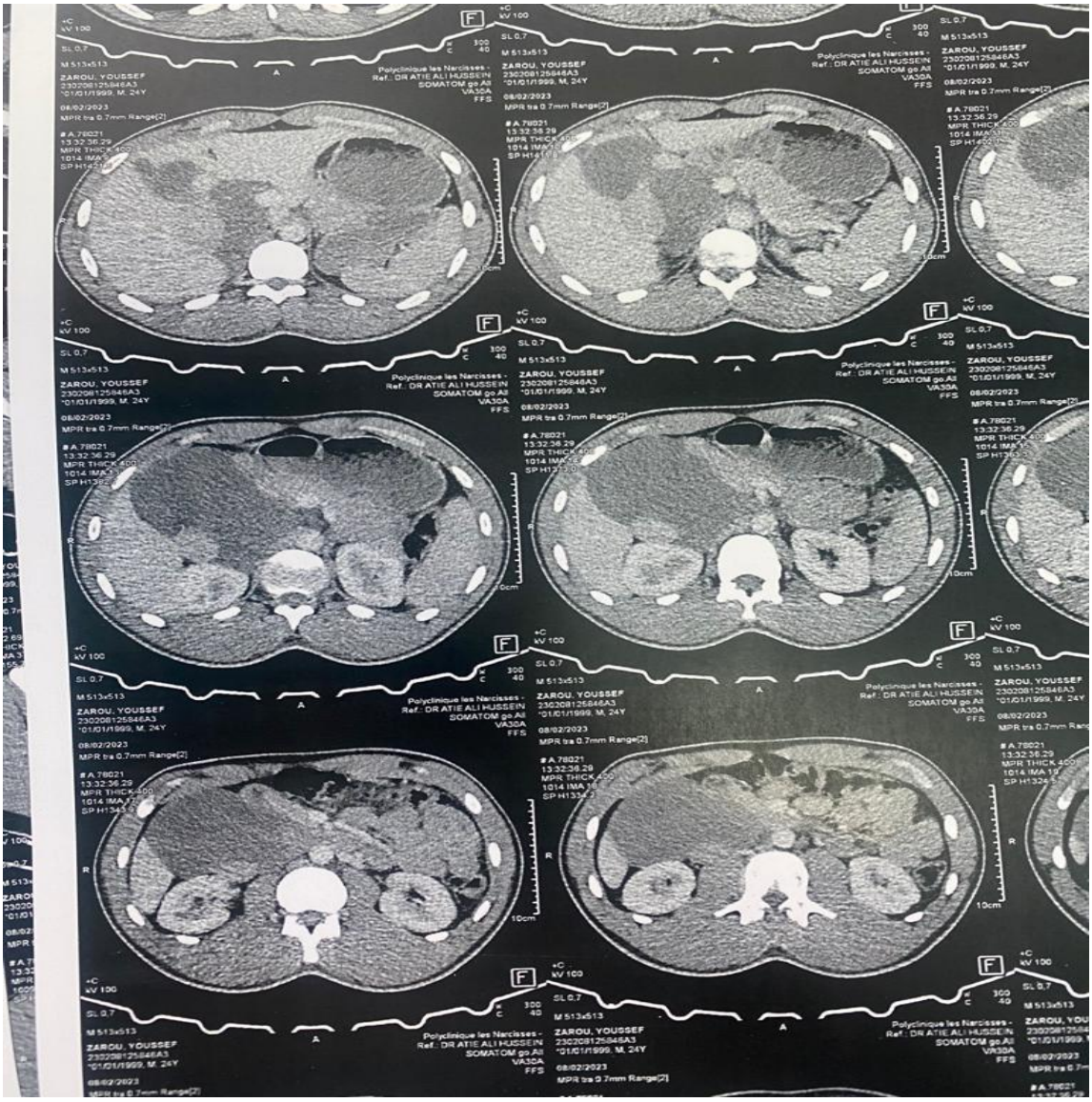

Figure 1: Lesional process lateralized to the right

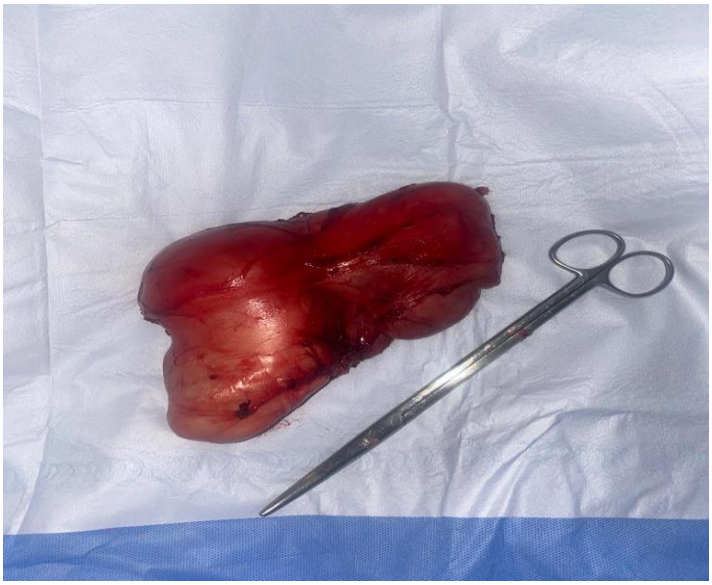

Figure 2: Mass of 20*15 cm with solidocyst appearance

DISCUSSION

Ganglioneuroma is a rare benign tumor of neuroectodermal origin. This tumor develops, like neuroblastoma and ganglioneuroblastoma, from the sympathetic nervous system. It is composed of mature ganglion cells and a stroma containing nerve cells and a Schwan contingent, whereas neuroblastoma and ganglioneuroblastoma are composed of more immature ganglion cells with a greater evolutionary potential [3]. In children, ganglioneuroma can represent the mature form of neuroblastoma. Conversely, the transformation of a ganglioneuroma into a neuroblastoma in adulthood is exceptional [4].

Ganglioneuroma develops along the sympathetic chain that runs from the neck to the pelvis. The retroperitoneal location is the most common after the mediastinal. The neck, the anterior mediastinum, the gastrointestinal tract are also possible but rarer locations [5]. It occurs at all ages, however it is mainly found in children and young adults [6]. The female sex is more often affected with a sex ratio of 0.75 [7].

These tumors progress at low noise and are most often discovered incidentally during a radiological examination to assess another condition or even during a palpable mass as is the case of one of our patients [8]. Sometimes, these tumors manifest themselves by non-specific pain, by signs of urinary, neurological, vascular or digestive compression which can even lead to a picture of acute intestinal obstruction. In fact, Cronin *et al*., report a case of intestinal obstruction by a ganglioneuroma occurring 18 years after the initial diagnosis [9].

The radiological diagnosis of these tumors is difficult. However, imaging makes it possible to specify the location of the tumor as well as the relationships with neighboring organs, particularly the vessels [10].

Ultrasound is not very specific and often reveals a heterogeneous tissue mass with well-defined contours in the adrenal compartment. The tumor can come close to the vessels without invading them [11].

CT scanning is nonspecific. Calcifications are encountered in approximately half of the cases. These vary in appearance, but are typically fine. Before injection, the tumor is homogeneous, hypodense, with regular and well-defined contours. After injection, contrast enhancement remains low to moderate, the mass becoming heterogeneous or remaining homogeneous [11].

Magnetic resonance finds a very limited mass, capable of reaching the adrenal compartment, not invading neighboring organs. Calcifications are difficult to analyze using this technique. In T1, the tumor presents a homogeneous hyposignal. In T2, the tumor can take two aspects:

A hyper signal or an intermediate signal, this essentially depends on the richness of stroma in the lesion. Contrast enhancement after Gadolinium injection is not specific, ranging from no contrast enhancement to weak inhomogeneous enhancement or even sometimes very strong enhancement [12].

The hormonal secretion assessment is in the majority of cases normal. Ganglioneuroma is generally a non-secreting tumor, but some authors report rare cases of ganglioneuroma with secretion of catecholamines or vasoactive intestinal polypeptide (VIP), responsible for diarrhea and high blood pressure [12, 13].

However, despite this multitude of explorations, ganglioneuroma still poses a differential diagnosis with other retroperitoneal tumors, namely ganglioneuroblastoma and neuroblastoma, but these tumors are suspected by scannographic signs of locoregional or distant invasion and by the character infiltrating intraoperatively [13].

However, the definitive diagnosis will only be made after a histological study of the surgical specimen. Indeed, the preoperative biopsy, although it allows the diagnosis to be made, a complete analysis of the excision site remains necessary due to the possibility of contingent neuroblastoma but also pheochromocytoma within the ganglioneuroma [2].

The treatment remains surgical. It consists of tumor excision; intervention all the more difficult as the tumor is large and has intimate relationships with neighboring structures, in particular the large vessels (IVC and aorta) [13].

Treatment should be performed early not only to confirm the nature of the mass, but also to prevent its increase in volume and compression of adjacent structures. The approach is generally a transperitoneal laparotomy, mainly for large masses [7].

The laparoscopic approach remains possible and even preferred for small, well-defined retroperitoneal masses with no intimate relationship with the large vessels. The evolution of these tumors is slow, but the increase in volume is the rule in the absence of treatment. Their prognosis is good in case of complete excision. The complications are mainly mechanical [7]. Local recurrence is exceptional, however the possibility of malignant transformation into ganglioneuroblastoma is possible, hence the interest in prolonged monitoring [1].

CONCLUSION

Despite its rarity and benignity, retroperitoneal ganglioneuroma deserves to be known. Diagnosis is often late. Imaging, particularly CT and MRI, confirms the retroperitoneal location of the tumor, its relationships and predicts its resectability. Its extremely favorable

prognosis after surgery justifies complete excision. Local recurrences, although rare, require periodic monitoring.

REFERENCES

1. Adraoui, J., El Jai, S. R., Chehab, F., Khaiz, D., & Bouzidi, A. (2008). Retroperitoneal ganglioneuroma, *Moroccan Journal of Urology*, 10, pp. 34-36.
2. Pelletier, F., Manzoni, P., Heyd, B., Algros, M. P., Ringenbach, F., Delroeux, D., ... & Humbert, P. (2006). Ganglioneuroma revealed by complicated nephritic colic. *La Revue De Médecine Interne*, *27*(5), 409-413.
3. Cocieru, A., & Saldinger, P. F. (2011). Images in surgery: retroperitoneal ganglioneuroma. *The American Journal of Surgery*, *201*(1), e3-e4.
4. Cyriac, S., Srinivas, L., Kathiresan, N., Sundersingh, S., Mahajan, V., & Sagar, T. G. (2009). Ganglioneuroma of the retroperitoneum presenting as a pediatric renal mass. *Indian Journal of Medical and Paediatric Oncology*, *30*(03), 105-107.
5. Singh, K. J., Suri, A., Vijjan, V., Singh, P., & Srivastava, A. (2006). Retroperitoneal ganglioneuroma presenting as right renal mass. *Urology*, *67*(5), 1085-e7.
6. Hajri, M., Ben Moualli, S., Ben Amna, M., Bacha, K., Chebil, M., & Ayed, M. (2001). Retroperitoneal ganglioneuroma, About a case. *Annals of Urology*, 35, pp. 145-147.
7. Sarf, I., el Mejjad, A., LBadre, L., Mani, A., Aboutaieb, R., & Meziane, F. (2003). Giant retroperitoneal ganglioneuroma, *Progress in Urology*, 13, pp. 502-505.
8. Papavramidis, T. S., Michalopoulos, N., Georgia, K., Kesisoglou, I., Valentini, T., Georgia, R., & Papavramidis, S. T. (2009). Retroperitoneal ganglioneuroma in an adult patient: a case report and literature review of the last decade. *South Med J*, *102*(10), 1065-1067.
9. Cronin, E. M. P., Coffey, J. C., Herlihy, D., Romics, L., Aftab, F., Keohane, C., & Redmond, H. P. (2005). Massive retroperitoneal ganglioneuroma presenting with small bowel obstruction 18 years following initial diagnosis. *Irish journal of medical science*, *174*, 63-66.
10. Nelms, J. K., Diner, E. K., Lack, E. E., Patel, S. V., Ghasemian, S. R., & Verghese, M. (2004). Retroperitoneal ganglioneuroma encasing the celiac and superior mesenteric arteries. *The Scientific World Journal*, *4*, 974-977.
11. Dubois, C., Jankowski, A., Gay-Jeune, C., Chabre, O., Pasquier, D., & Ferretti, G. (2005). Imaging of adrenal ganglioneuroma: a case report. *Journal de Radiologie*, *86*(6 Pt 1), 659-662.
12. Cai, J., Zeng, Y., Zheng, H., Qin, Y., Kaiyong, T., & Zhao, J. (2010). Retroperitoneal ganglioneuroma in children: CT and MRI features with histologic correlation. *European journal of radiology*, *75*(3), 315-320.
13. Gargouri, M. M., Sfaxi, M., El Atat, R., Mouelli, S. B., Hassine, L. B., Chebil, M., & Ayed, M. (2007). Retroperitoneal ganglioneuroma. A case report. *La Tunisie medicale*, *85*(1), 78-80.

Quoted by (4)

- Attafi, S., Zehani, A., Chelly, I., Smichi, I., Azouz, H., Nouira, Y., ... & Kchir, N. (2016). Unusual adrenal incidentaloma: the ganglioneuroma. *African Journal Of Urology*, *22*(4), 240-242.
- Kora, C., Oulad Amar, A., El Arabi, S., Rockson, O., Nasri, S., & Skiker, I. (2021). Retroperitoneal ganglioneuroma presenting as an obstructive pyelonephritis: a case report. *Journal of Surgical Case Reports*, *2021*(4), rjab104.
- *Bouzid, A., Belhadj, A., Saidani, A., Bokal, Z., Khefacha, F., & Chebbi, F. (2021). Unusual retrorectal ganglioneuroma: a case report of laparoscopic assisted approach. Pan African Medical Journal*, *38(1)*.
- Baïzri, H., Zoubeir, Y., Elhadri, S., Elanzaoui, J., Chahbi, Z., Kaddouri, S., ... & Zyani, M. (2014). Hepatic cytolysis revealing adrenal ganglioneuroma. *The Pan African Medical Journal*, *17*, 224-224.