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The surrenal ganglioneuroma: a rare anatomo-clinical entity (About a case)

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ABSTRACT

The ganglioneurom is a rare benign tumor of the adult and the adrenal localization is only 20% of all the localizations. We report the case of an adrenal ganglioneurosis in a 51-year-old patient. She had moderate pain in her right flank. Clinical examination was normal. Ultrasound showed a rounded hypoechoic formation between the upper pole of the right kidney and the liver of 37mm diameter evoking a right adrenal process. Computed tomography (CT) showed the presence of a well-defined 50/30 mm oval-shaped adrenal mass with a few thin endolesional calcifications. The biological balance of which a hormonal evaluation was normal. The patient had undergone a complete resection of the sural tumor. On histological examination, the diagnosis is that of a ganlioneurome. The surgery was simple.The diagnosis of ganlioneurome is difficult and is definitely retained only after the immunohistochemical study of the surgical specimen. Complete monobloc resection is the only treatment.

Keywords : Ganglioneurome, adrenal tumor, diagnosis, resection, histopathology.

INTRODUCTION

Ganglioneuromes are rare benign tumors located in the adrenal gland (20%), along the sympathetic chain and particularly in the posterior mediastinum (40%) and retroperitoneum (30%) (1). They belong to the group of neurogenic tumors, developing at the expense of sympathetic ganglionic chains, which also includes ganglioneuroblastomas and neuroblastomas (2).

We report a case of a 51-year-old woman who had moderate right flank pain isolated, with a subnormal hormonal balance. The diagnosis was made on the abdominal CT scan, the histological study of the surgical specimen confirmed the ganglioneurome.

MATERIALS AND METHOD

Patient of 51 years, was operated 20 years ago for vesicular lithiasis under laparotomy, which had presented isolated and moderate pains of the right flank without other associated signs. The clinical examination had found a normotensive patient in good general condition. The abdomen was supple. There was no lumbar contact or palpable masses. The ganglionic areas were free.

The blood count was normal, the function

Renal and blood crass were without abnormalities.

The hormone balance showed a slight increase in plasma cortisol at 22.65 µg / dl at 8 am, the plasma aldosterone assay was normal at 129 pg / ml, urinary normetanephrine at 0.16 mg / 24h, metanephrine at 0 , 12 mg / 24h, vanilmandelate (VMA) at 7.60 mg / 24h.

Ultrasound showed a rounded hypoechoic formation between the upper pole of the right kidney and the liver of 37mm diameter evoking a right adrenal process. Computed tomography (CT) showed the presence of a well-defined 50/30 mm oval-shaped adrenal mass with a few thin endolesional calcifications. The patient was operated on and a complete surgical excision of the right adrenal mass was performed.

The histological study of the surgical specimen showed a nodular formation of 23g measuring 4x4x2,5cm of firm consistency of white - greyish appearance corresponding to a fusocellular proliferation without atypia nor of mitosis, it associates some mature ganglion cells grouped in nests And presence of foci of calcification.

An immuno-histochemical study showed that tumor proliferation diffuse PS 100 and does not express desmin and AML. The morphological and phenotypic aspect of a ganglioneuome was retained.

The postoperative follow-up was simple. The exit was decided on the 4th day. The follow-up was 12 months, no local or remote recurrence was observed.

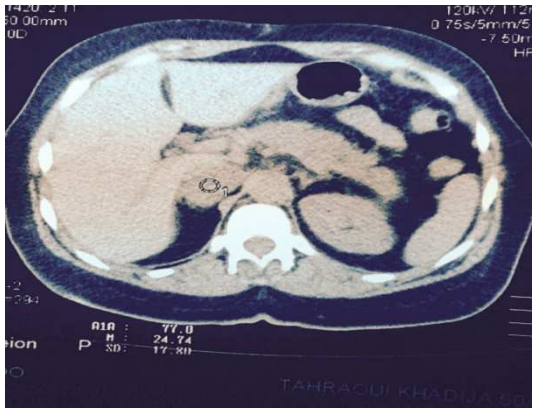

Figure 1

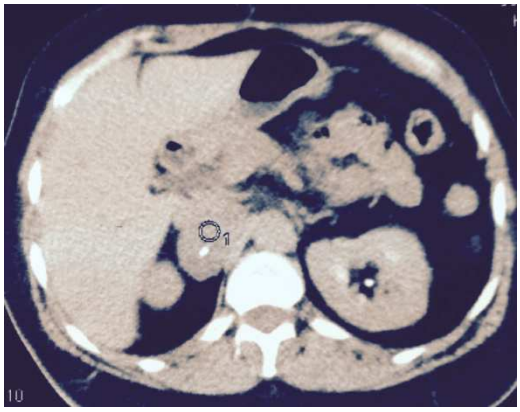

Figure 2

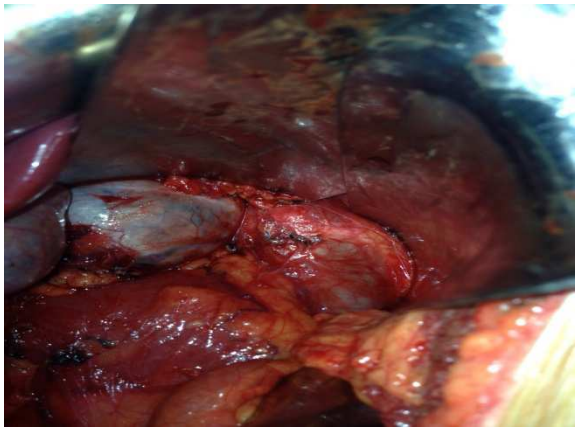

Figure 3

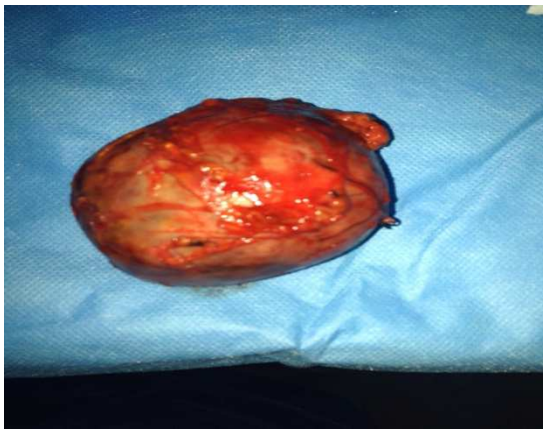

Figure 4

RESULT AND DISCUSSION

Ganglioneurom is a benign nervous tumor of the child and young adult. The first case was described by LORETZ in 1870.

This is a rare benign tumor of neuroectodermal origin, that grows like neuroblastoma and ganglioneuroblastoma, from the sympathetic nervous system that goes from neck to pelvis. It is composed of mature ganglion cells and stroma containing nerve cells associated with a contingent schwann, unlike neuroblastoma and ganglioneuroblastoma which are composed of more immature ganglion cells whose evolutionary potential is more important (3). Retroperitoneal localization is the most frequent after the mediastinal. The neck, the anterior mediastinum, the ganglioneuroma represents only 0,7à1,6% of primary retroperitoneal tumors which, in turn, represent less than 1% of all tumors. It affects children and adolescents preferentially before the age of 20 years. Women are more affected than men; The sex ratio is 0.72-0.77 (4).

This tumor evolves at low noise, it is discovery most often fortuitous on the occasion of a radiological examination for assessment of another condition or a palpable mass. Sometimes it is manifested by non-specific pain such as in our patient, or urinary compression signs, neurological, vascular or gastrointestinal (3-4).

Ganglioneuroma is usually considered a non-secreting tumor; But there is an exceptional possibility of secretion of catecholamines (4-5).

The radiological diagnosis of this tumor is difficult. However, the imaging makes it possible to specify the seat of the tumor as well as the relations with the neighboring organs, in particular the vessels. Ultrasound is not very specific and often reveals a heterogeneous tissue mass with well-defined contours of the adrenal lodge. The tumor can come close to the vessels without invading them. Computed tomography can reveal calcifications in 50% of variable and fine (5-6) cases. Before injection, the tumor is homogeneous, hypodense, with regular contours and well limited. After injection, the contrast setting remains low to moderate, the mass becoming heterogeneous or remaining homogeneous. The magnetic resonance imaging (MRI) showed a homogeneous tumor with relatively low signal intensity on T1 and heterogeneous with predominant signal on T2 (4-5).

Despite this multitude of explorations, the ganglioneuroma still poses a differential diagnosis with other retro peritoneal tumors, namely ganglioneuroblastoma and neuroblastoma (1-3-5), but these tumors are suspected in front of Scanner signs of invasion or locoregional At a distance and in front of the infiltrating character in peroperative. Nevertheless, the diagnosis of certainty will only be carried out after a histological study of a biopsy puncture of the mass or on the surgical part (3-4-10).

The treatment remains surgical (4-6-9). It consists of tumor excision; An intervention which is all the more difficult because the tumor is large and has intimate relations with neighboring structures, in particular the large vessels (VCI and aorta). The approach is generally a transperitoneal laparotomy, mainly for the large masses. The laparoscopic pathway is favored for the small, well-defined retroperitoneal masses without an intimate relationship with the large vessels. A complete analysis of the excision piece remains necessary because of the possibility of neuroblastoma contingents but also of pheochromocytoma within the ganglioneurome. 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. Local recurrence is exceptional, however the possibility of a malignant transformation into a ganglioneuroblastoma is possible (5-7).

CONCLUSION

The adrenal ganglioneurom is a rare and benign non-secretory tumor. Nevertheless it deserves to be known. Its clinical picture is often uncouth or asymptomatic. Imaging, especially CT and MRI confirm the retroperitoneal seat of the tumor, its relationships and predict its reseability. Its extremely favorable prognosis after surgery justifies complete resection.

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