Laparoscopic Adrenal Surgery for Giant Ganglioneuroma

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Abstract: We present herein a case of a 24-year-old woman with incidentally diagnosed right adrenal ganglioneuroma with 14cm in size that was removed completely by the laparoscopic approach. The patient was asymptomatic and the tumor was first diagnosed on abdominal ultrasonography. A subsequent computed tomography (CT) of the abdomen confirmed a 12x 11x9cm complex expansive mass of right adrenal, with well-defined outlines. Magnetic resonance imaging (MRI) showed a solid lesion measuring 11 x 9 x 11cm arising from the right adrenal. Laparoscopic complete excision of the mass was accomplished through a transabdominal lateral approach. The surgical specimen weighed 665 g and 14 x 10 x 7 cm in size. There was no complication in postoperative period, and the patient was released from the hospital two days after the operation. The patient resumed her normal activities in one week. Histology was consistent with an adrenal ganglioneuroma. A control CT was made one year after the surgery with no evidence of lesion suggestive of relapse**.** Adrenal ganglioneuromas are rare lesions with a benign behavior in which surgery is the only possible form of treatment. In centers of advanced laparoscopy this method of access can be used, even for larger lesions.

Keywords: Urology, Oncological Surgery, general laparoscopy.

INTRODUCTION

Ganglioneuromas (GN) are the most highly differentiated and invariable benign tumors of the ganglia cells which also include ganglioneuroblastomas with intermediary differentiation and highly malignant neuroblastoma. These are rare neoplasias and may appear in any peripheral ganglia. 56 -70% of cases are located in the posterior mediastinum and retroperitoneum associated with the paravertebral sympathetic plexus, and 20-30% involve the adrenal gland [1] representing 0-6% of the incidentalomas. GN generally occurs between the first and second decade and at the end of the third decade of life. Due to their slow growth these tumors are commonly asymptomatic and are discovered incidentally. When they are located in the adrenal, they may cause epigastralgia or might be diagnosed during a physical examination. These neoplasias occasionally may secrete active hormones such as androgens, catecholamines and VIP (vasoactive intestinal peptide) generating respective symptoms of virilization, hypertension and diarrhea [3-6]. The present golden standard treatment for small benign adrenal tumors (< 6 cm) is laparoscopic adrenalectomy (LA) [7-10]. However, larger lesion s (> 6 cm) present a high risk of malignancy [11-12], increasing the possible risk of peritoneal cancer dissemination during this access [13-15]. Various recent studies have demonstrated that LA may be extended to any adrenal tumor where there is no radiological evidence of adrenal infiltration or venous invasion [16-19]. This paper reports a case of a voluminous adrenal ganglioneuroma diagnosed incidentally which was completely removed *via* laparoscopy.

CASE REPORT

A 24-year-old woman was referred to the Urology Department of our hospital with an incidental diagnosis of a 14 cm mass in the right adrenal during an ultrasound examination to investigate dysmenorrhoea. The patient did not have any other symptoms and had a healthy appearance. Her blood pressure was of 110 x 80 mmHg and there is no abdominal palpable mass. An x -ray of the thorax was found to be normal as were the routine blood tests. A directed hormone evaluation was made which demonstrated that all values were normal in accordance with Table **1**. A computerized tomography (CT) of the abdomen (Figure **1**) showed a complex expansive formation with well-defined outlines located at the right adrenal measuring 12 x 11 x 9 cm with enhancement by contrast. Soon after the computerized tomography, the patient was submitted to an magnetic resonance imaging (MRI) which visualized a solid lesion measuring 11 x 9 x 11 cm located in the right adrenal (Figure **2**) with homogeneous hyposignal with relation to hepatic parenchyma in a T1 weighed sequence and heterogeneous hypersignal in T2. There was a growing and delayed impregnation of paramagnetic contrast by the tumor in dynamic study.

The findings in the image made the diagnosis of a benign lesion, particularly a possible ganglioneuroma

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Table 1: General Endocrinology Data

Exam	Result	Reference Value
Glucose	81,80 mg/dl	65-99 mg/dl
Creatine	0,87 mg/dl	<1,4 mg/dl
Dehydroepiandrosterone sulfate	115,00 ug/dl	35-430 ug/dl
Aldosterone	13,50 ng/dl	4-31 ng/dl
Renin-angiotensin activity	1,0 ng/ml/hora	0,3-1,6 ng/ml/hora
Cortisol	0,15 ug/dl	1-25 ug/dl
Dopamine	16,29 pg/ml	< 30 pg/ml
Adrenaline	84,90 pg/ ml	< 140 pg/ ml
Noradrenaline	803,10 pg/ ml	< 1400 pg/ ml
Free Cortisol	26,70 mcg/24 hrs	10- 90 mcg/24 hrs
Urinary Metanefrine	22,20 mcg/24 hrs	> 400 mcg/24 hrs
Urinary Normetanefrine	89,90 mcg/24 hrs	> 800 mcg/24 hrs

which oriented our efforts towards a laparoscopic resection despite the considerable size of the tumor.

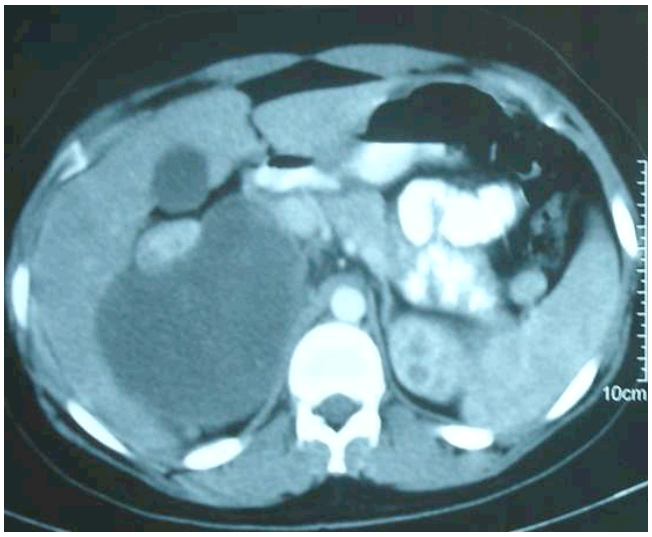

Figure 1: Computed tomography showing a mass with well-defined outlines located at the right adrenal with enhancement by contrast.

Surgical Technique and Evolution

We opted for left side decubitus. The Hansen technique was used to introduce a 10-mm trocar followed by a peritoneal insufflation and the introduction by puncture from another portal o f 10-mm and two of 5-mm. Throughout the surgical procedure we placed the portal of the camera below the original position to visualize the dissection of the tumor on the other side. The tumor was easily visible in the abdominal cavity and no metastatic lesions were identified. After the access of the retroperitoneum and mobilization of the right colon, the inferior vena cava and the adrenal vein were identified with the latter clipped by hem-o-lock and sectioned. The other vessels of the tumor were clipped using a metallic or polymer clip according to the caliber. No ultrasonic scissor was used. To remove the surgical specimen was used a 15-mm extraction bag. The specimen was removed integrally by Pfannestiel incision. The duration of the surgery was 300 minutes with a blood loss of 150ml. The surgical specimen weighed 665g and 14x10x7cm in size. The patient was released from the

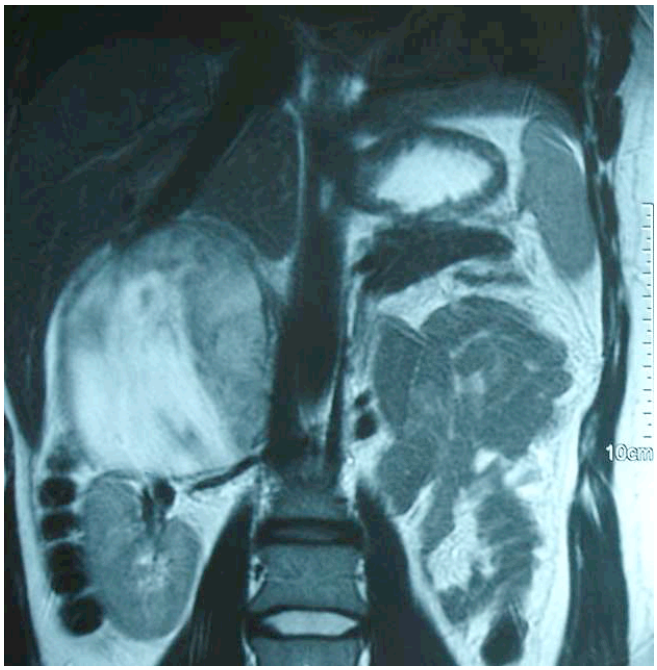

Figure 2: Magnetic resonance imaging. After intravenous injection of gadolinium, the mass showed a progressive, heterogeneous, and delayed enhancement.

hospital two days after the operation and resumed her normal activities in one week. A control TC was made one year after surgery with no lesion suggestive of recurrence.

Histopathological Characteristics

The tumor was a solid globular and encapsulated tumor of a dark yellow color with a firm elastic consistency with 13.7 x 9.5x 7.2 cm in size (Figure **3**). The surface was bright and wound with the lesion weakly adhered to the capsule of the adrenal gland. Microscopically (Figure **4**), the tumor was composed of arranged in fascicules. Mature spread or arranged in agglomerations were also noted. These characteristics ma de the diagnosis of ganglioneuroma possible and no other additional therapy for this benign tumor was required.

DISCUSSION

Ganglioneuroma tumors are rare which makes an accurate estimate of their occurrence difficult. They most frequently occur in the posterior mediastinum with

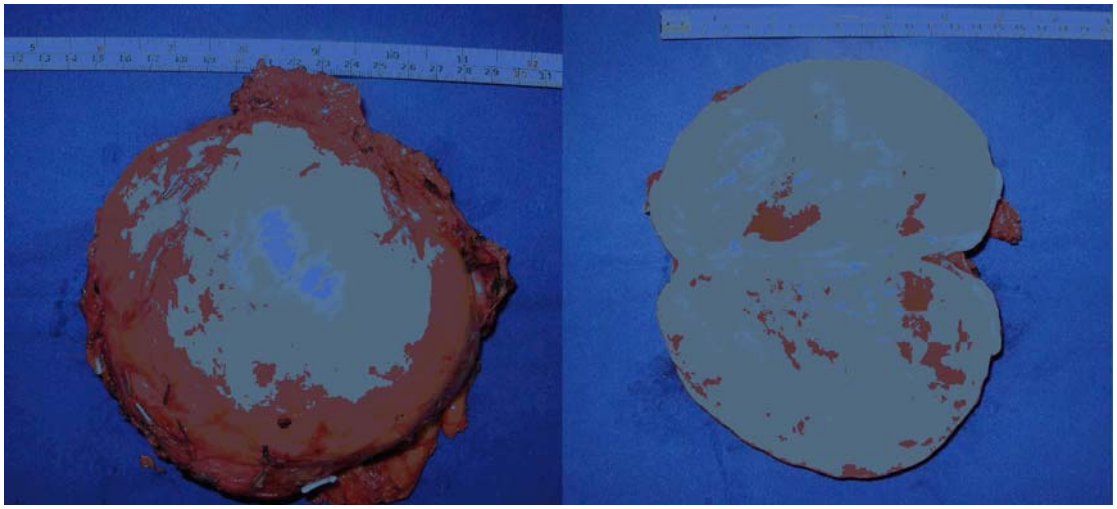

Figure 3: The tumor was solid and encapsulated with a firm elastic consistency measuring 13.7 x 9.5 x 7.2 cm.

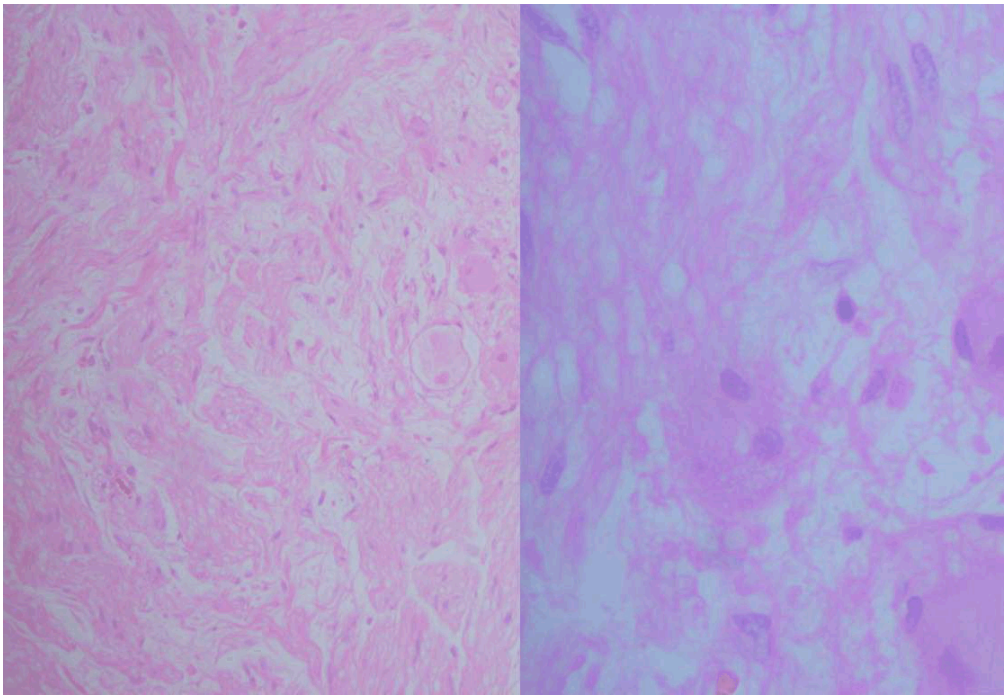

Figure 4: The tumor was composed of fusiform cells arranged in fascicules. Mature ganglionary cells spread or arranged in agglomerations were also noted (H&E 20x).

the incidence in pediatric population reaching 1 in 100,000 children per year [1]. With greater facility in accessing images in examination, incidental diagnosis of adrenal lesions has become more frequent. Approximately 1 to 10% of all CTs show findings that were suspected to adrenal tumors, with ganglioneuroma occupying 0 % - 6% of these lesions [2].

In both TC and RM, retroperitoneal or adrenal ganglioneuroma appears as an ovoid lesion, well defined and lobulated. This tumor has a tendency to partially or completely involve main vessels without, however, causing significant compression to the lumen. In a TC without contrast, the lesion may take on a homogeneous aspect with attenuation inferior to the muscle. Calcification may also occur in 20% of the cases which tend to be discrete and point -like. These aspect helps in the differentiation of neuroblastomas which have coarse amorphous calcifications [20]. After the infusion of contrast, varied standards of distinction were described. Ichikawa *et al* described the standard of heterogeneous and delayed distinction which occurs due to the abundance of myxoid matrix being more common. Its presence leads to the accumulation of material in contrast in extra cellular space [21]. In the evaluation of MRI, the ganglioneuromas appear as homogeneous lesions with a low intensity signal in TI and hypersignal in T2 depending on proportions between myxoid stroma and component cells as well as the quantity of collagen fibres present in the tumor. The higher proportion of stroma myxoid, the more intense is the T2 signal. The ganglioneuromas are distinguished prematurely after the administration of paramagnetic contrast with a progressive increase in signals in the more delayed phases [20]. Laparoscopic adrenalectomy was introduced in 1991 [22] and has become the gold standard method in the removal of the adrenal in benign diseases due to the significant advantages such as shorter hospitals stays, earlier return to work, better cosmetic results, lower risk of incisional hernias and even lower total cost than the traditional open technique [23-26]. In cases of suspected malignant adrenal neoplasias, the open technique continues to be the first choice because of the possible risk of peritoneal dissemination of neoplastic cells and the recurrence of the disease at the portals of laparoscopic access [13-15]. In the absence of a local invasion or metastases, there is no other clinical, biochemical or radiological exam that can exclude the malignant adrenal tumor in the preoperative phase [19, 27-28]. We know that the larger the adrenal tumor, the greater the risk of malignancy, although the majority of voluminous adrenal masses are benign, both histological and in their behavior [29]. Furthermore, the small size of the tumor alone does not guarantee the absence of the malignancy because 13.5% of adrenal carcinomas are smaller than 5 cm in diameter on their diagnosis [30]. In experienced hands, the capsular rupture (main cause for the peritoneal dissemination of neoplastic cells) during the laparoscopic adrenalectomy tends to be less frequent [17-19]. An open or conversion surgery is, however, recommended when malignancy is suspected (fixed tumor, invasion of the pancreas, spleen or superior pole of the kidney, thrombosis of the vessels, lymphadenopathy, or distant metastasis) in the preoperative phase or during the laparoscopic surgery. Various authors, in advanced laparoscopic centers, have published good results with laparoscopic ressection of large adrenal tumors with benign aspect. The only apparent disadvantage is a longer period of time in surgery and greater blood loss when compared to the open technique [17-19, 31-34]. To our knowledge this is the largest adrenal tumor removed by laparoscopy.

CONCLUSION

Adrenal ganglioneuromas are rare lesions with a benign behavior in which surgery is the only possible form of treatment. In centers of advanced laparoscopy this method of access can be used, even for larger lesions.

Disclosure Statement No competing financial interests exist.

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