Case Report ISSN 2689-1093

Surgical Research

Pheochromocytoma and Extraadrenal Paraganglioma: Case Report and Review of the Laparoscopic Approach

Saúl Eduardo Torres Arano[1]*, Bryan Armando Morales Eslava[2], Manuel Guadalupe Torres González[1], Carolina Flores Castro[1] and Federico López Rosales[1]

[1]Department of General Surgery, Hospital Regional de Alta Especialidad de la Península de Yucatán, Mérida, Yucatán, México.

[2]Department of General Surgery, Hospital Regional de PEMEX Villahermosa, Villahermosa, Tabasco, México.

***Correspondence:**

Saúl Eduardo Torres Arano, address: Doctores Street Number 17, Santa Isabel, Postal Code 24157, Ciudad del Carmen, Campeche, México, Phone number: +52 9999934391.

Received: 02 Jul 2022; **Accepted:** 04 Aug 2022; **Published:** 10 Aug 2022

Citation: Arano SET, Eslava BAM, González MGT, et al. Pheochromocytoma and Extraadrenal Paraganglioma: Case Report and Review of the Laparoscopic Approach. Surg Res. 2022; 4(2): 1-7.

ABSTRACT

The reported percentage of catecholamine-producing tumors is around 80-85% in pheochromocytomas and 15-20% in paragangliomas. The case of a 34-year-old female patient with a clinical, imaging and histopathological diagnosis of pheochromocytoma with paraganglioma with successful laparoscopic surgical resolution with a retroperitoneal approach is described.

The cardinal symptom is uncontrolled hypertension despite medical management with multiple antihypertensives. The initial diagnostic approach is with the measurement of metanephrines in plasma and urine, if this is not conclusive, the clonidine test can be performed. With a positive laboratory result, an imaging study should be performed for localization and finally surgical resolution with a preference for the laparoscopic approach, with less risk of post-surgical complications.

Keywords

Paraganglioma, Pheochromocytoma, Adrenalectomy, Laparoscopic, Surgery.

Introduction

Pheochromocytoma is a tumor that originates in the adrenal gland, and is characterized by being a producer of catecholamines: epinephrine, norepinephrine and dopamine. Paraganglioma is a tumor derived from extra-adrenal chromaffin cells of paravertable, thoracic, abdominal and pelvic sympathetic ganglia or parasympathetic ganglia located along the glossopharyngeal and vagus nerves located in the neck and base of the skull, however these do not they are producers of catecholamines [1]. The reported percentage of catecholamine-producing tumors is around 80-85% in pheochromocytomas and 15-20% in paragangliomas [2]. The incidence of these tumors in patients with hypertension varies between 0.2 and 0.6%.

It is important to suspect this pathology and arrive at the diagnosis, because if the timely treatment is not carried out; morbidity and mortality is high due to its implication at the cardiovascular level, if it evolves it can cause a mass effect spreading to adjacent tissues and organs.

The presentation of paraganglioma and pheochromocytomas is very rare with an incidence of 5 patients per year. The presentation of cases is highly variable and most doctors have little experience for timely diagnosis, which is why a multidisciplinary team is required for the approach. Our objective with the presentation of this clinical case is to describe the diagnostic and surgical approach of the pathology, to describe a colic case with the two entities in the same patient, since during the review of the literature we observed that there are no reported cases with the two entities. Simultaneous with surgical resolution with a laparoscopic approach.

Clinical Case

A 34-year-old female with a history of high blood pressure diagnosed 5 years ago, difficult to control, treated with telmisartan, prazosin, and propranolol.

He began his condition 5 years ago presenting increased BP, accompanied by tachycardia, headache and diaphoresis. He also mentioned that since the diagnosis of high blood pressure, he has presented difficult control of BP. She was sent to Endocrinology and during the evaluation, a tomography of the abdomen was requested, where a 40x34 mm solid lesion with increased uptake was found in the body of the right adrenal gland, suggestive of pheochromocytoma (Figure 1). There is another adjacent retrocaval lesion of 28 x 20 mm with a hypodense center with probable central necrosis to consider extra-adrenal paraganglioma. Urine metanephrines met: 60 mg/dl normeta : 200 mg/dl. Total metanephrines: 260 DHEA: 117, ACTH: 16.7.

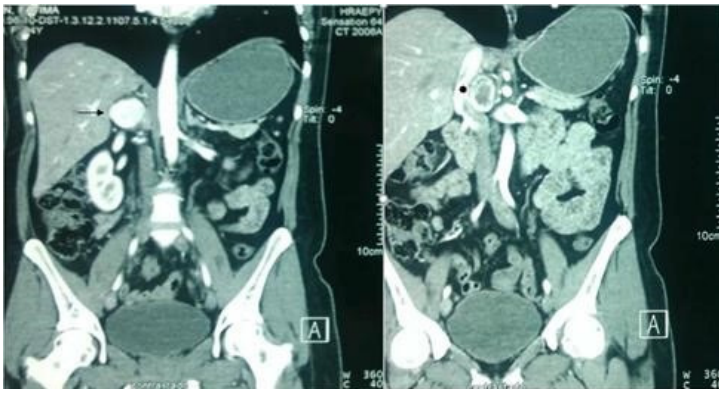


Figure 1: rounded lesion with regular edges and increased uptake at the infrahepatic level (arrow) suggestive of pheochromocytoma. In the image on the right, a heterogeneous retrocaval lesion with irregular edges (asterisk) can be seen adjacent to the first image with possible central necrosis suggestive of a paraganglioma.

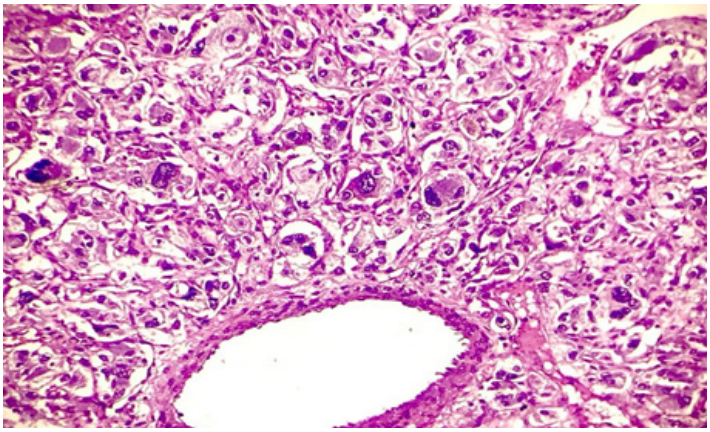


Figure 2: Histological section of pheochromocytoma. Pleomorphism detail, nuclear grade 4.

Despite the absence of an increase in metanephrines and catecholamines in the urine, it was decided to perform a laparoscopic right adrenalectomy due to a high clinical suspicion of pheochromocytoma. During the procedure, she presented multiple episodes of severe hypertension despite being managed with preoperative blockade with propranolol and prazosin. During the postsurgical follow-up, the piece was sent to the pathology laboratory, where the following findings were found: pheochromocytoma of 3.5 x 2.5 cm. And a 2.5x2.2 cm paraganglioma with 2-point Weiss criteria for malignancy. She was sent for follow-up by Surgical Oncology to rule out MEN 2 A and B (Figure 2-5).

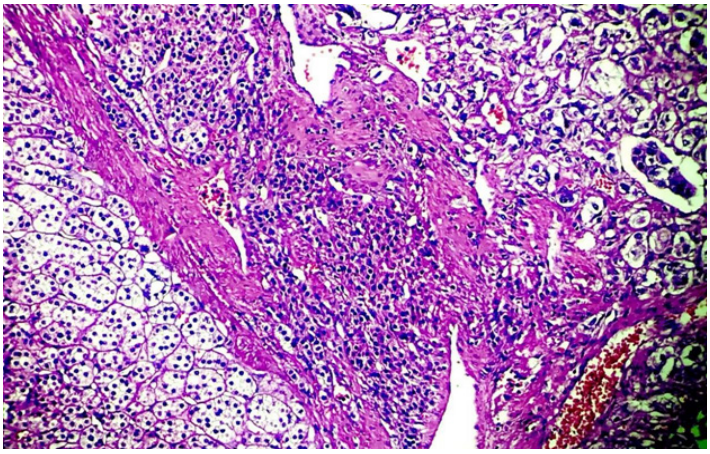


Figure 3: Histological section showing the contrast between the pheochromocytoma (right) and the normal residual adrenal gland (left).

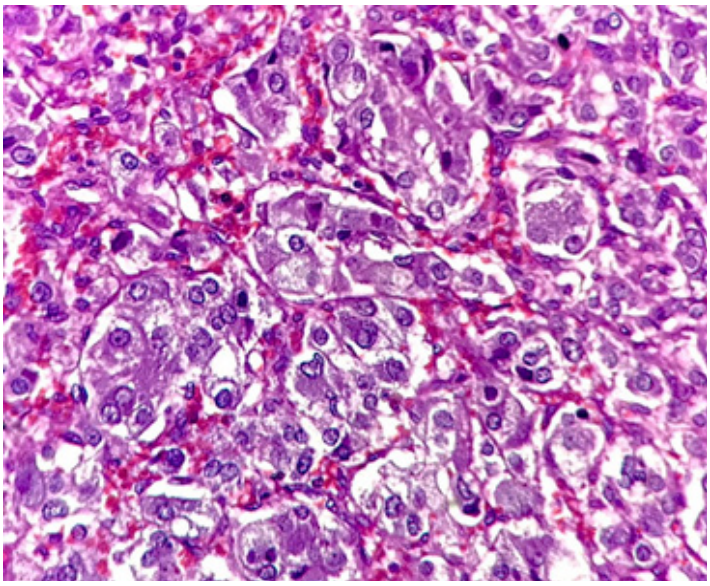


Figure 4: Histological section of the extraadrenal paraganglioma

The surgical technique is described below:

With the patient in left lateral decubitus at 90°, pneumoperitoneum was performed with a Veress needle at 15 mm Hg and trocars were

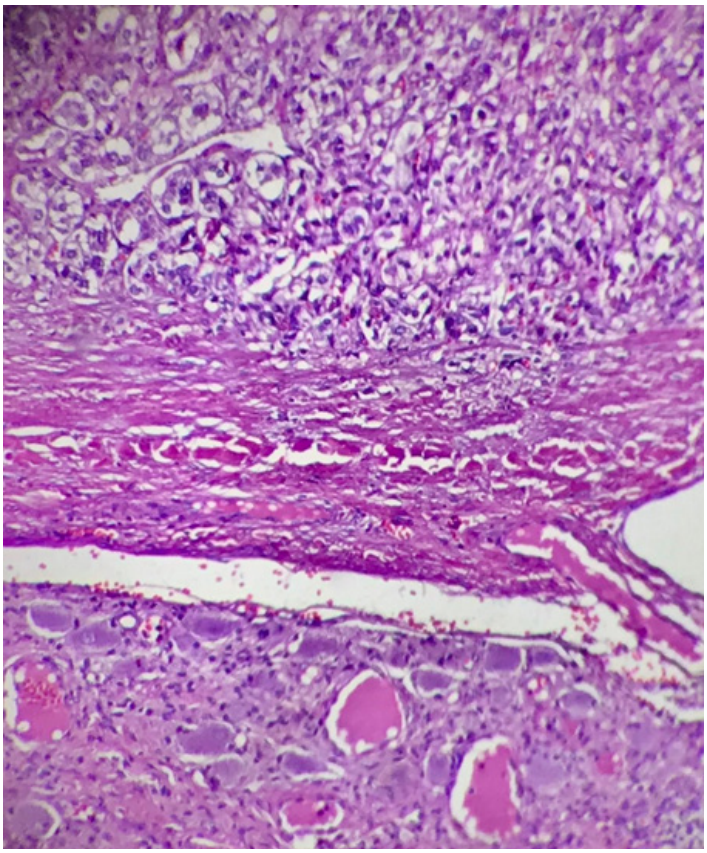
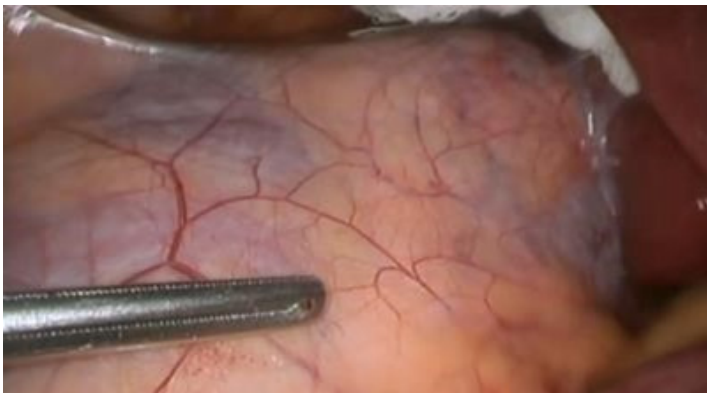


Figure 5: Retrocaval extraadrenal paraganglioma (top) with spinal ganglion (bottom).

placed, one 12 mm in the midaxillary line for the video camera, another 12 mm in the anterior axillary line for the right-hand instrument, one 5-mm posterior axillary line for the left-hand instrument, and one 5-mm midclavicular line for the liver retractor. It can be seen that the patient had fatty liver, which made exposure with the separator somewhat difficult. The kidney and adrenal gland cranial to it are identified and we begin the incision of the peritoneum that covered the gland, just below the hepatic border at the entrance of the gland to the retrohepatic portion (Figure 6 and 7). This is done with blunt and sharp dissection with the help of the harmonic scalpel until reaching the triangular ligament to be able to move and separate the liver medially and superiorly and to be able to expose the retrohepatic portion of the adrenal tumor. Subsequently, we began the dissection between the medial side of the adrenal tumor and the vena cava, finding small vessels from the retroperitoneum, which were ligated with hemolocks and cut with a harmonic scalpel (Figure 8). We proceed with this dissection cephalad to locate the suprarenal vein and be able to ligate it early (Figure 9). We do this dissection in a blunt and sharp manner with the harmonic scalpel and gently pulling the gland laterally, avoiding excessive manipulation (Figure 10). At this moment, the suprarenal vein can be seen, which is ligated with a hemolock and cut with a harmonic scalpel. It is worth mentioning that until now, no hypertensive peak had occurred despite the manipulation that can be observed. Subsequently, we began the dissection of the caudal portion of the gland to separate it from the upper pole of the kidney and moving laterally until exposing the posterior muscles of the abdomen (Figure 11). After that, we returned to the medial mobilization of the gland, which is already separated from the vena cava, which exposed the medial portion of the paraganglioma, with an interface of highly vascularized tissue between both structures. We tried to dissect these vessels by ligating them with hemolocks, but they were difficult to control, so we decided instead to use the gland as a means of traction to expose the paraganglioma, since we had not presented hypertensive peaks, and to resect them en bloc (Figure 12). Then, tractioning the gland laterally allowed us to progressively expose the retrocaval paraganglioma, performing a combination of blunt dissection with the suction cannula and sharp



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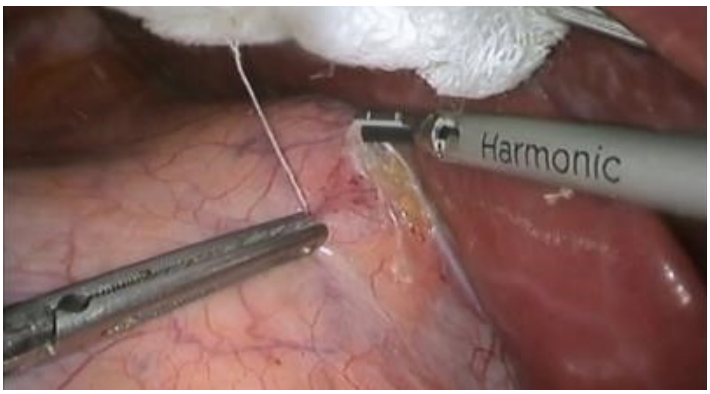

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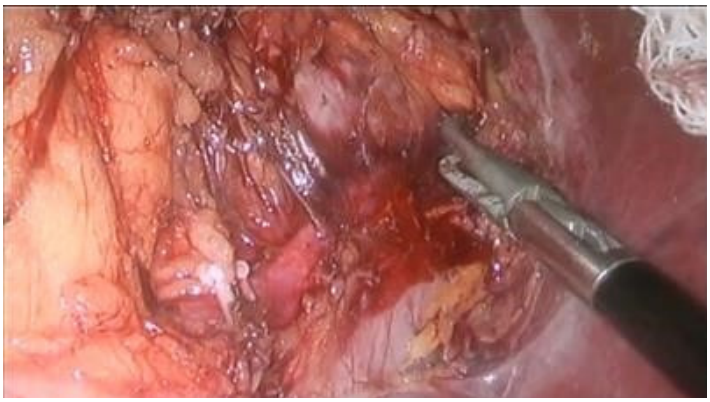

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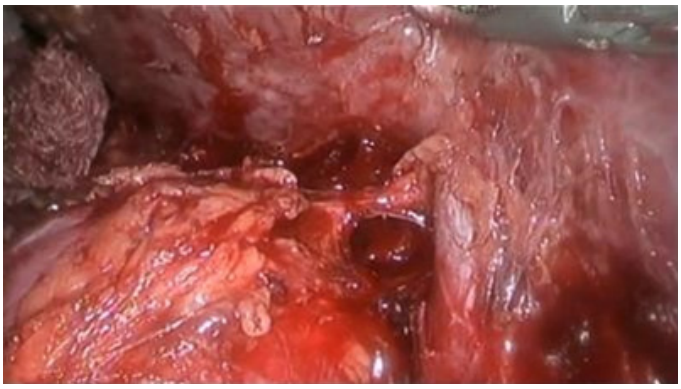

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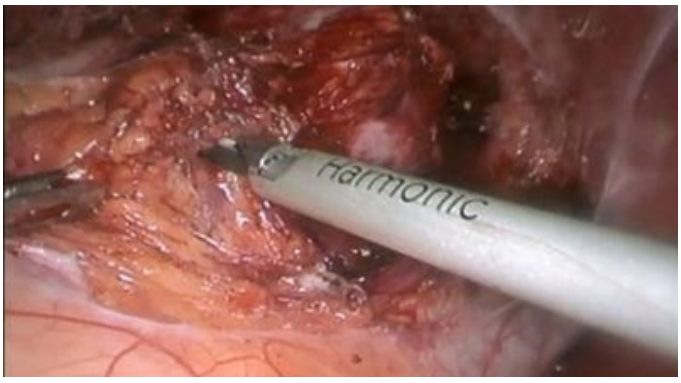

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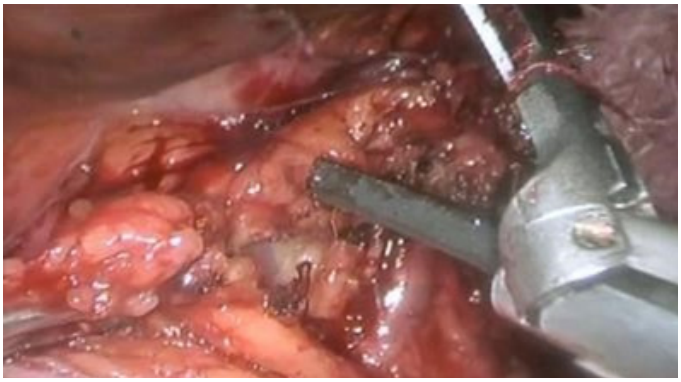

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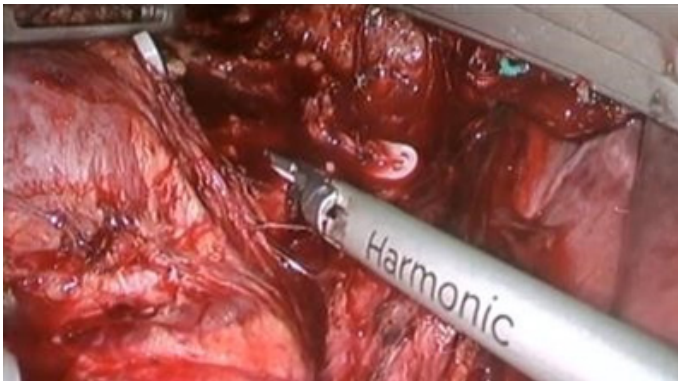

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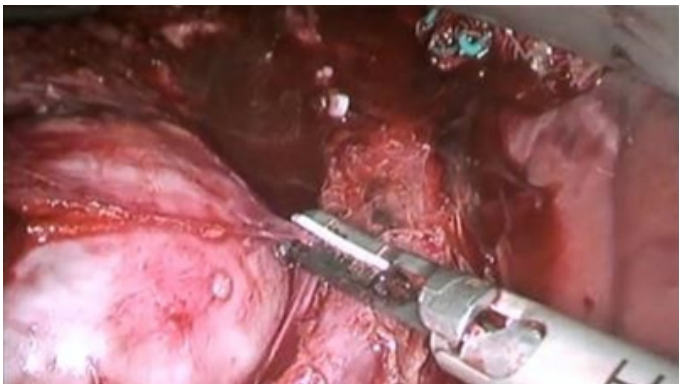

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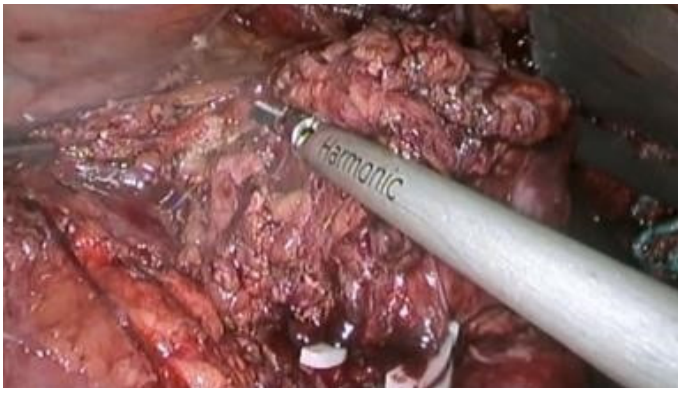

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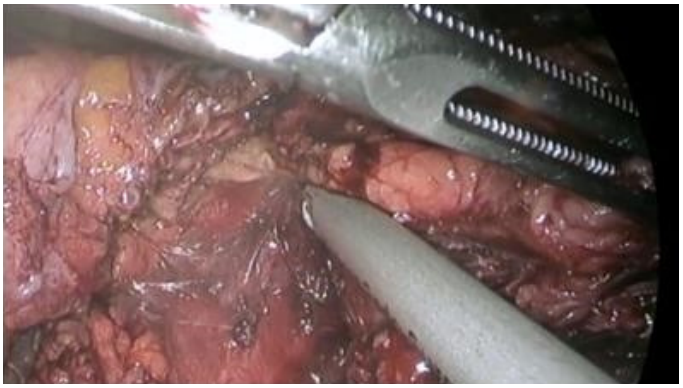

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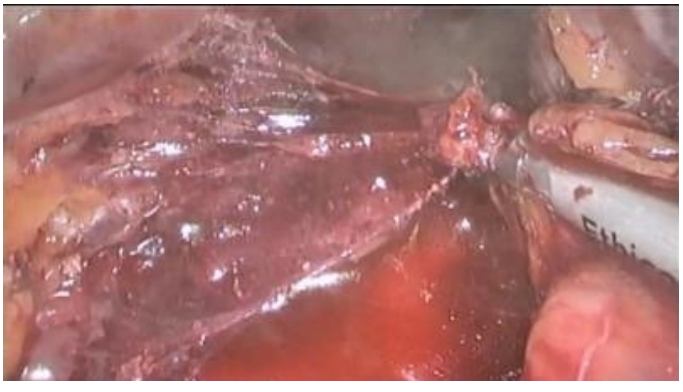
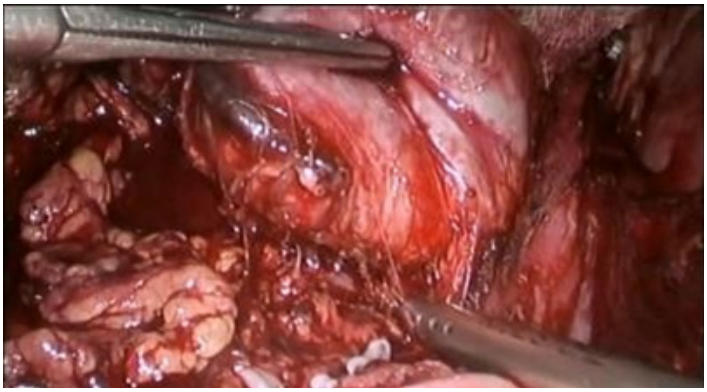


Figure 16

dissection with the harmonic scalpel between the paraganglioma and the vena cava (Figure 13). Here you can see how the vena cava folds with traction, so care must be taken not to confuse the planes (Figure 14). At this time we return to the posterolateral dissection since with the manipulation of the paraganglioma a hypertensive peak was presented. While the anesthesiologists controlled it, we continued with the dissection to completely separate the tumor and the periadrenal fat from the posterior muscles since no hypertensive spikes were seen when manipulating the gland (Figure 15). At this time, we finished with the dissection of the most superior and medial portion of the gland, above the paraganglioma, finding retroperitoneal vessels which were cut with harmonic (Figure 16). Once the pressure is controlled, we return to the dissection of the paraganglioma (Figure 17). In this shot you can see the two tumors perfectly, with the most medial and retrocaval portion of the paraganglioma pending dissection. To continue the blunt dissection of this portion, manipulation of the paraganglioma is unavoidable, which we did in communication with the anesthesiologists to stop manipulating when hypertensive peaks appeared. After the partial separation of the vena cava (Figure 18), we found several nonanatomical vessels coming from the aorta which were difficult to dissect, controlling them with hemolocks and harmonic scalpel. You can appreciate the great vascularity of the paraganglioma. Once the tumor block was released from the vena cava, we again tried to perform block traction through the adrenal tumor and not the paraganglioma to avoid hypertensive peaks. At this moment, the vena cava can be seen completely free of the tumor block and only these branches from the retroperitoneum and the aorta remain pending, which we control in the same way as the previous ones (Figure 19). You can see how difficult it was to control these branches, presenting bleeding that temporarily obscured the work field (Figure 20). In this final part of the dissection, in order to separately ligate each of the vessels that went to the gland, it was unavoidable to manipulate the paraganglioma, which is why hypertensive peaks were presented again (Figure 21). Once each of the vessels had been isolated, they were ligated proximally with hemolocks and cut with the harmonic (Figure 22). Finally, the tumor block was completely resected together with the periadrenal fat, leaving the posterior muscles of the abdomen completely free, as well as the vena cava and the multiple vessels originating from the retroperitoneum and the aorta (Figure 23). We covered the hemolocks with gelfoam to avoid contact with the posterior face of the vena cava and here you can see the block piece (Figure 24).



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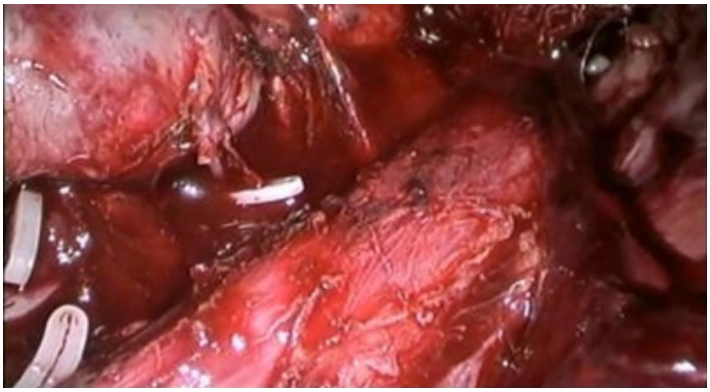

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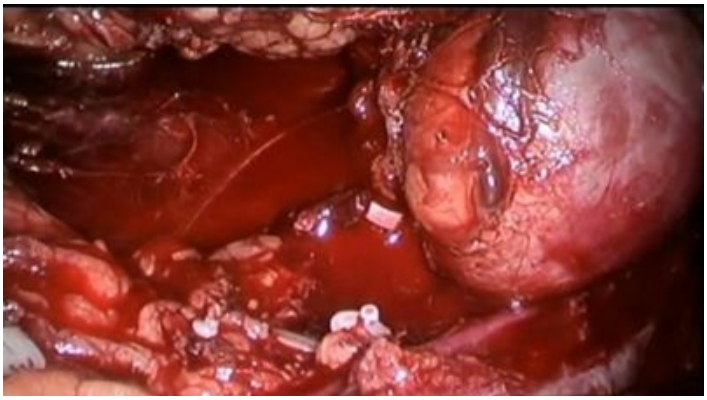

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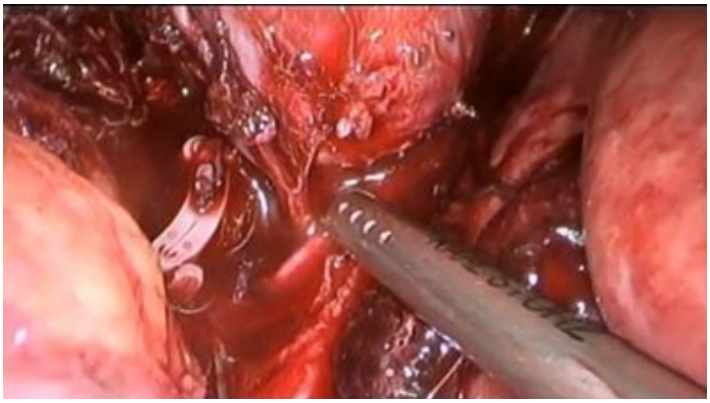

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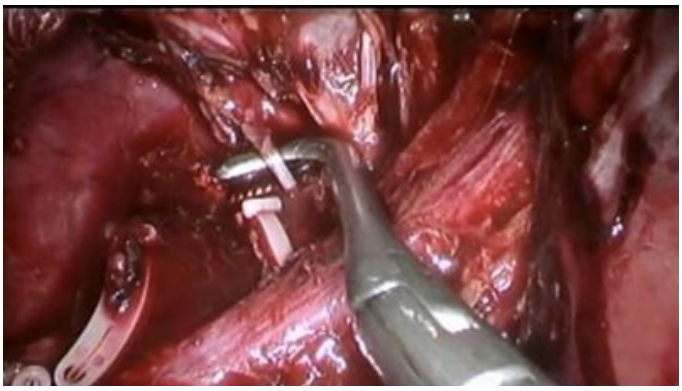

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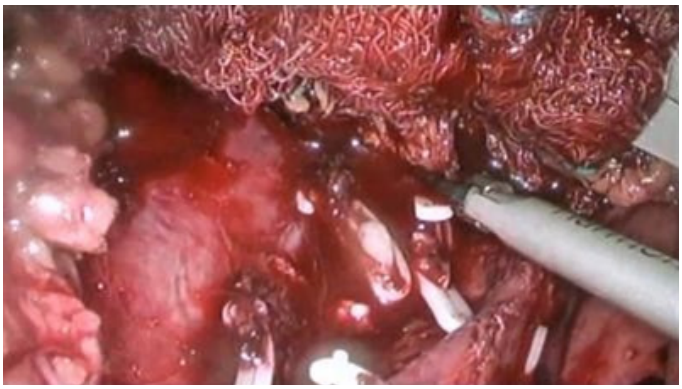

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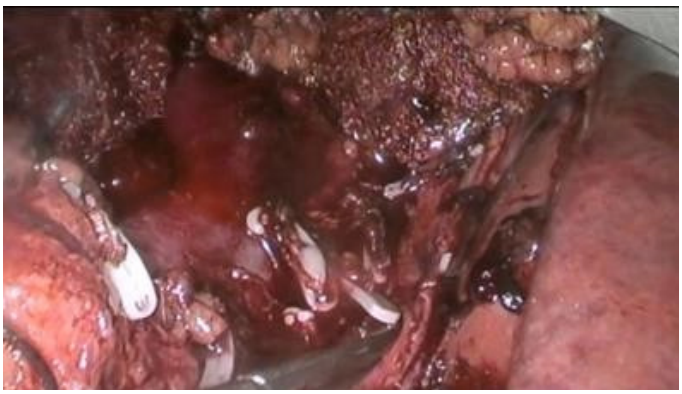

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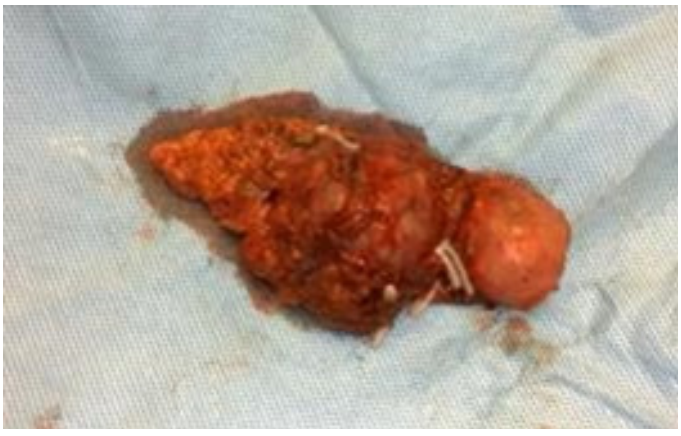

Figure 24

Discussion

To start the diagnostic protocol, measurement of metanephrines in plasma or urine should be performed, having a sensitivity of 97% and a specificity of 91% [3], the sample should be taken in the supine position, as this decreases the production of norepinephrine, reducing the number of false positives. For sample collection, patients must be in this position for at least 30 minutes before sample collection.

If there is elevation of both normetanephrines and metanephrines, there is a high probability that it is a pheochromocytoma, in the same way if there is an elevation of only normetanephrines or metanephrines above three times its value, it should be followed up to locate the tumor. However, when the values are at the upper limit despite performing the test in the supine position, two screening tests may be performed, the first is metanephrines in urine and the second is the clonidine test, which is positive despite the drug having a sensitivity of 96% with a specificity of 100% [4].

Regarding imaging studies, there is currently insufficient evidence to formulate guidelines about when and how to perform imaging studies in patients at risk, the initial study that should be performed is a contrast-enhanced CT scan, with sensitivity between 88 and 100%. The image that is displayed can be homogeneous or heterogeneous, necrotic with some calcifications, solid or cystic. MRI is recommended for cases in which recurrence, extra-adrenal or metastatic tumors are suspected; it has a sensitivity of 95-100% [5]. In some cases, a genetic study must be carried out.

Surgical Preparation

All patients with functioning paraganglioma or pheochromcytoma should undergo preoperative blockade to prevent perioperative cardiovascular complications. The clinical practice guideline of the endocrinology society suggests the use of α-adrenergic receptor blockers as the first option, it has been shown to significantly decrease diastolic pressure and heart rate [6], it should be administered from 7 to 14 days prior to surgery. A high-sodium diet and hydration should be included in the pre-surgical preparation to avoid hypotension after tumor removal [7].

Surgery

Laparoscopic resection is recommended for most tumors. However, open resection is recommended for tumors larger than 6 cm to avoid tumor rupture and local recurrence, ensuring complete resection. However, it is described that the laparoscopic approach is associated with less postoperative pain, less bleeding, fewer days of hospitalization and morbidity [8]. There are no data regarding any difference in recurrence rate after open vs laparoscopic surgery. The mortality rate is approximately 1%, and the conversion rate is approximately 5% depending on the experience of the surgeon.

The two most common approaches are the transperitoneal lateral abdominal approach (Gagner) and the retroperitoneal approach (Walz) [9]. The approach to choose depends on the characteristics of the tumor and the experience of the surgeon. The abdominal approach is preferred for performing bilateral adrenalectomies or

large tumors. Paragangliomas are generally found in anatomical areas that are difficult to approach laparoscopically, so the open approach is preferred.

Surgery must be performed carefully and the entire tumor must be removed, avoiding its rupture. The endocrinology society guideline suggests that partial adrenalectomy should be performed in patients with hereditary tumors, in patients with previous contralateral adrenalectomy to avoid permanent hypoadrenalism [10]. It has been observed in some studies that with the preservation of half of an adrenal gland and there is even a report that 15% of the adrenal gland is sufficient to preserve cortical function [11].

Conclusion

It is important to consider this pathology in order to be diagnosed, if the necessary studies for its approach are not available, we recommend referring the patient to a hospital that has the resources. We recommend the laparoscopic approach due to the lower morbidity of this procedure as well as the lower incidence of post-surgical complications, as long as the surgeon is trained to perform this procedure.

References

1. Lenders JW, Duh QY, Eisenhofer G, et al. Pheochromocytoma and paraganglioma: an endocrine society clinical practice guideline. The Journal of Clinical Endocrinology & Metabolism. 2014; 99: 1915-1942.
2. Lenders JW, Duh QY, Eisenhofer G, et al. Pheochromocytoma and paraganglioma: an endocrine society clinical practice guideline. J Clin Endocrinol Metab. 2014; 99: 1915-42.
3. Omura M, Saito J, Yamaguchi K, et al. Prospective study on the prevalence of secondary hypertension among hypertension patients visiting a general outpatient clinic in Japan. Hypertens Res. 2004; 27: 193-202.
4. Eisenhofer G, Goldstein DS, Walther MM, et al. Biochemical diagnosis of pheochromocytoma: how to distinguish true- from false-positive test results. J Clin Endocrinol Metab. 2003; 88: 2656-2666.
5. Giménez -Roqueplo AP, Caumont -Prim A, Houzard C, et al. Projection imaging of paraganglioma and pheochromocytoma in SDHX mutation carriers: a multicenter prospective study by investigators PGL.EVA. J Clin Endocrinol Metab. 2013; 98: E162-E173.
6. Kocak S, Aydintug S, Canakci N. Alpha blockade in the preoperative preparation of patients. With pheochromocytoma. int Surge. 2002; 87: 191-194.
7. Pacak K. Preoperative Management of the Pheochromocytoma Patient. J Clin Endocrinol Metab. 2007; 92: 4069-4079.
8. Walz MK, Alesina PF, Wenger FA, et al. Laparoscopic and retroperitoneoscopic treatment of pheochromocytomas and retroperitoneal paragangliomas: results of 161 tumors in 126 patients. World J Surg. 2006; 30: 899-908.
9. Dickson PV, Alex GC, Grubbs EG, et al. Posterior retroperitoneoscopic adrenalectomy is a safe and effective alternative to transabdominal laparoscopic adrenalectomy for heochromocytoma. Surgery. 2011; 150: 452-458.
10. Iihara M, Suzuki R, Kawamata A, et al. Adrenal- preserving laparoscopic surgery in selected patients with bilateral adrenal tumors. Surgery. 2003; 134: 1066-1072.
11. Brauckhoff M, Gimm O, Thanh PN, et al. Critical size of residual adrenal tissue and recovery desde impaired early postoperative adrenocortical function after subtotal bilateral adrenalectomy. Surgery. 2003; 134: 1020-1027.