Laparoscopic adrenalectomy for giant pheochromocytoma.
What is the size limit ?

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AIM: *This case report describes a giant pheochromocytoma in a young female, experienced cardiac symptoms, treated by a transperitoneal laparoscopic right adrenalectomy.*
Material and methods: A 29 years old female with Taki-tsubo syndrome, consequent to the chronic release of catecholamines, with a palpable abdominal mass and vague abdominal symptoms was referred to our department.
Abdominal CT scan has demonstrated a solid mass of 13 cm in the right adrenal space so, after pre-operative management with alpha-adrenergic receptor and beta blockade and a 3D CT scan reconstruction a right adrenalectomy laparoscopic approach was performed.
RESULTS: *Our result underlines that 13 cm in size for a giant pheochromocytoma is not an absolute contraindication to perform a minimally invasive approach in expert hands, with optimal surgical, oncological and cosmetic results.*
DISCUSSION: *The only curative option for non-metastatic pheochromocytomas disease is surgical resection. Laparoscopic adrenalectomy is the treatment of choice but the limit size for a safe and feasible minimally invasive approach is not yet defined.*
CONCLUSIONS: *This case report could help to better define more solid recommendations in the next future and also provide landmarks and key steps for laparoscopic surgeons.*

KEY WORDS: Giant Pheochromocytoma, Laparoscopic Adrenalectomy, Pheochromocytoma Management

Introduction

Giant pheochromocytomas are rare tumours larger than 7cm in size. Surgery is the chosen treatment but the limit size for a minimally invasive approach is not yet defined. The standard approach for giant pheochromocytomas, with no pre- or intraoperative evidence of malignancy, is a current dilemma for surgeons [1-2] and it is still considered technically demanding operation. SAGES guidelines for minimally invasive treatment of adrenal pathology recommends a laparoscopic adrenalectomy, emphasizing that a tumour size of 7.5 cm or larger is an independent risk factor for a longer operating room time, more blood loss and a more frequent subsequent use of open surgery [3]. This case report shows feasibility and outcomes of transperitoneal laparoscopic right adrenalectomy for a giant adrenal tumor of 13 cm. This paper was written according to CARE guideline.

Case Report

A 29 years old female with Tako-tsubo syndrome, consequent to the chronic release of catecholamines, experienced episodes of tachycardia and palpitations associated with nausea and chest pain was referred to our department.
The patient presented with a palpable abdominal mass with vague abdominal symptoms. Endocrine counselling,

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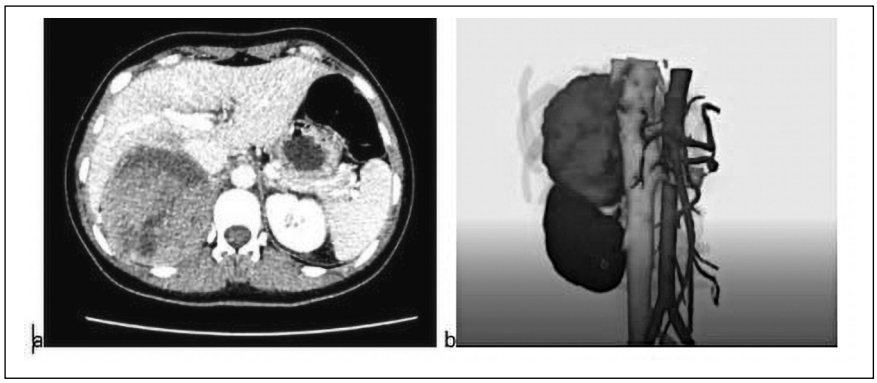

Fig. 1

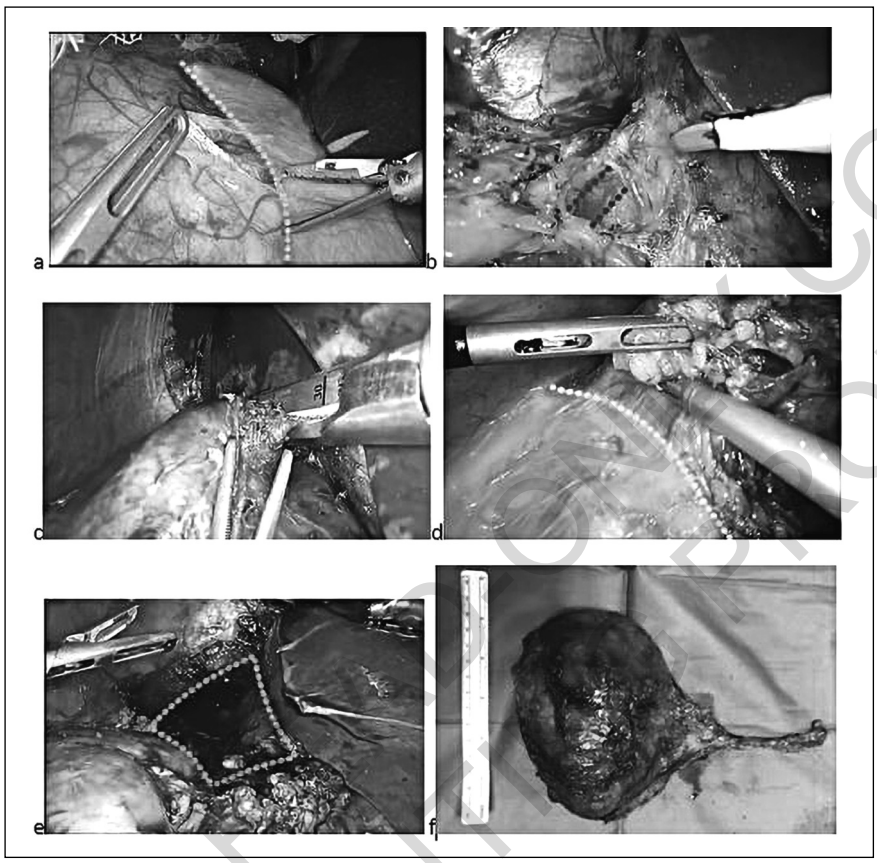

Fig. 2

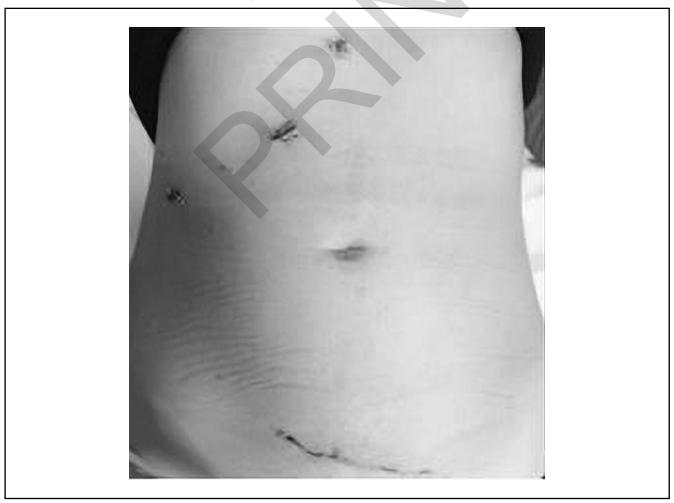

Fig. 3

biochemical studies and 24-hours urine collection were suspected for pheochromocytoma. Abdominal CT scan demonstrated a solid mass of 13 cm in the right adrenal space (Fig. 1A), with solid and necrotic-colliquative component. 3D CT scan reconstruction was performed (Fig. 1B).

Imaging showed the relationship of the pheochromocytoma with its surrounding structures. The normal configuration of the right adrenal gland was indistinguishable from the mass which compresses the vena cava in its subhepatic course. After pre-operative management with alpha-adrenergic receptor and beta blockade, a transperitoneal laparoscopic right adrenalectomy was proposed as treatment of choice.

Under general anesthesia, patient was positioned in left decubitus and four trocars were placed. Laparoscopic

entry was performed by Open Veress-assisted technique [4]. The operation begins by identifying the first landmark: the inferior vena cava with the incision of the peritoneum. The voluminous mass altered the normal anatomy of the area making this step more difficult and the correct identification of quadrilateral of the Albaran-Chatelin which corresponds to the adrenal lodge resulted not easy. In order to minimize hemodynamic instability due to catecholamine release, during the vascular time was necessary to reduce the direct manipulation or compression of the adrenal gland as well as the control of CO_2 insufflation.

The operation begins the identification of the right renal vein and such sequently of the right adrenal artery The diameter of adrenal vein was increased by the larger tumour size (Figs. 2A-2B). Hence, the section of the adrenal vein was performed with a mechanical suturing machine with vascular load (Fig. 2C). Finally, the liberation of the pheochromocytoma was completed (Figs. 2D-2E), the mass was placed in the endo-bag to be extracted by minimal Pfannestiel incision (Fig. 2F). The mean time for adrenalectomy was 155 minutes. There were no postoperative complications and the patient was discharged well on postoperative day 4. The final histopathological diagnosis for the patient turned out to be pheochromocytoma 13 cm x 10 cm x 5 cm. At one and six -months follow-up patient was completely asymptomatic, requiring no pharmacological treatment and presented an optimal cosmetic result (Fig. 3).

Discussion

Many doubts still concern the use of minimally invasive approach for giant pheochromocytoma [1]. The surgical method depends on the tumour size, signs of malignancy, presence of local invasion and surgeon expertise with a good team effort. This case report suggests that a 13 cm diameter is not an absolute contraindication to laparoscopic approach if performed by expert laparoscopic surgeons. Pheochromocytomas are rare neuroendocrine tumours arising from chromaffin cells of the adrenal gland, the classic triad occurs with headaches, palpitations and hypertension due to the plasmatic metanephrine release. If this tumour exceeded 7.5 cm in size are defined as giant pheochromocytoma, and in this case, they usually manifest themselves with vague abdominal symptoms without the classic triad. Biochemical analysis and CT scan are fundamental and considered as the gold standard for the diagnosis. Complete resection is currently the only treatment for non-metastatic pheochromocytoma after medical premedication with alpha- followed by beta-adrenergic blockage.

Laparoscopic adrenalectomy (LA), is considered the gold standard for adrenal pathology and there are multiple minimally invasive approaches to remove the adrenal gland: lateral or anterior transperitoneal and lateral or posterior retroperitoneal. Transperitoneal approach with patient in lateral decubitus offer a direct and great exposition of quadrilateral of the Albaran-Chatelin space limiting the adrenal gland and provides an adequate working space. SAGES guidelines for minimally invasive treatment of adrenal pathology recommends a laparoscopic adrenalectomy, emphasizing that a tumour size of 7.5 cm or larger is an independent risk factor for a longer operating room time, more blood loss, and a more frequent subsequent use of open surgery [3].

Economopolos et al [1] describe a prolonged operative time for tumor size at least 4 cm and an increase of postoperative complications ad conversion rate by tumor size greater than 8 cm. Carter [4] compared laparoscopic adrenalectomy for pheochromocytomas disease <6cm vs > 6 cm in size. They concluded that LA is equally safe and feasible however the larger adrenal tumour of this series was only 9 cm in size. Clement [5] in a recent literature review reported three cases of laparoscopic excision of pheochromocytoma > 10 cm in size. Despite the results of the literature not encouraging for the size of this case hemodynamic stability, full alfa-blockade and optimal communication between the surgical and anesthetic team were achieved. Landmarks are fundamental in laparoscopic surgery [6] and surgery was completed without complications and with good oncological and cosmetic outcomes [7]. New technologies improve surgical practice and outcomes [8,9]. In the present study, 3D imaging allowed us to better visualize adrenal tumor and anatomical surrounding organs.

Conclusion

Our result underline that LA is safe and feasible for giant pheochromocytoma, up to the size of 13 cm, when performed by expert surgeons and this could help to better define more solid recommendations in the next future.

Riassunto

L'intervento per via laparoscopica di grandi masse surrenaliche è ormai da anni una procedura standardizzata. Rifacendoci alle linee guide SAGES (citate nel manoscritto con relativo riferimento bibliografico), l'approccio laparoscopico è considerato sicuro e fattibile in caso di lesioni, prive di caratteri di malignità, inferiori a 7.5 cm, descrivendo per casi di dimensioni superiori maggiori complicanze intraoperatorie e aumentato rischio di conversione in open senza tuttavia fissare un limite per la scelta della chirurgia mininvasiva come primo approccio. Nel nostro case report, riportiamo un caso di feocromocitoma gigante (dimensioni pari a 13 cm) trattato completamente con approccio laparoscopico mininvasivo per via anteriore transperitoneale dimostrandone la sua

fattibilità priva di complicanze intra operatorie e perioperatorie. L'approccio mininvasivo ha assicurato una breve degenza, assenza di complicanze post-operatorie e un ottimo risultato estetico con minima incisione di servizio tipo Pfannestiel. Il nostro case report potrebbe in futuro, contribuire a modificare il limite fissato per l'approccio laparoscopico nella chirurgia surrenalica.

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