Laparoscopic adrenalectomy in a case of congenital duplication of the inferior vena cava
Case report

Giuseppe Pisano*, Pietro Giorgio Calò*, Stefano Piras*, Umberto Pisano**, Angelo Nicolosi*

*Department of Surgical Sciences, University of Cagliari, Cagliari, Italy
**Department of General Surgery, Aberdeen Royal Infirmary, Aberdeen, United Kingdom

Laparoscopic adrenalectomy in a case of congenital duplication of the inferior vena cava. Case report

INTRODUCTION: Congenital duplication of the inferior vena cava is frequently associated with other vascular anomalies of its venous tributaries. Awareness of such occurrence facilitates laparoscopic surgery and avoids inadvertent vascular injuries.

CASE REPORT: An adrenal mass of increasing size was discovered in a 39 years old lady previously submitted to restorative proctocolectomy for Familial Polyposis Coli. Since the first preoperative work up, Computerized Tomography showed a duplicated inferior vena cava as well as other visceral and vascular anomalies. During laparoscopic adrenalectomy a double adrenal vein was discovered: the first one draining normally into the vena cava and the second one into the right renal vein. Both of them were clipped and divided and surgical outcome was successful.

DISCUSSION: The progress in cross-sectional imaging made easily recognisable congenital anomalies of the inferior vena cava in patients otherwise asymptomatic. Its occurrence has been evaluated through previous reports on venous anomalies during adrenal and renal surgery as well as through angiographic studies. Knowledge of these anomalies is very important for interventional radiologists, urologists and for general surgeons. However when facing adrenal surgery the operator should be aware that a double vein can be found in up to 10% of the cases and such occurrence is more predictable in case of pheochromocytoma and of large adrenal mass. Surgeons should rely both on preoperative diagnostic imaging and careful dissection through laparoscopic magnified view to avoid harmful bleeding complications.

KEY WORDS: Inferior vena cava duplication, Laparoscopic adrenalectomy, Vascular anomalies.

Introduction

Laparoscopic adrenalectomy (LA) requires a thorough knowledge of gland’s blood supply both of normal anatomy and of its variation 1. One of the most dreaded complication in this type of surgery is massive haemorrhage from the right adrenal vein as it enters the inferior vena cava (IVC) or from the left adrenal vein when it merges into the renal vein 2,3. Knowledge of possible anatomical variations of venous supply is mandatory in order to prevent serious bleeding complications 4-6. The present work originates from a case of laparoscopic right adrenalectomy where IVC duplication and multiple venous and arterial anomalies were encountered. We reviewed vascular embryogenesis and previous reports of traditional surgery and laparoscopy with regards to IVC anomalies. We also analysed if preoperative work up can obviate possible surgical lesions and make laparoscopy safer.
Case report

A 39 years old lady was referred to our Institution by the Oncology Department following a right adrenal mass. Eight years before she underwent a restorative proctocolectomy for a Familial Polyposis Coli (FPC), already in malignant transformation with multiple regional node metastases. For this reason, a few weeks after surgery, she also underwent chemotherapy and pelvic irradiation. During the follow up, at a distance of 6 years from the previous surgery, a CT scan showed for the first time a 23 mm adrenal adenoma, that reached the size of 34 mm in the following two years (Fig. 1). The adrenal adenoma had a slight metabolic activity at the PET scan without features of malignancy (Fig. 2); in a previous PET scan, however, the same gland had no metabolic activity at all. Endocrine assessment showed normal level of serum and urinary cortisol, normal low dose suppression test as well as normal plasma aldosterone and urinary cathecolamines. The indication to LA arose from a steady increasing size of the gland together with a mild suspicion of malignancy. Before surgery we carefully evaluated all the previous medical records and CT scan performed on the patient during the previous hospital...
admissions. We realized in this way that the gastrointestinal surgeons responsible of the first operation of procto-colectomy were aware of a complex vascular and visceral anomalies that included polysplenia, absence of pancreatic tail and IVC duplication (Fig. 1): in particular both iliac veins merged in the left IVC trunk which abnormally ended in the right atrium from above, whereas a small right IVC normally entered the right atrium from below (Figg. 3, 4). The right renal vein drained into the small right IVC. Other arterial anomalies included a splenic artery originating directly from the aorta and common hepatic artery originating from the superior mesenteric artery.

LA appeared more challenging than usual for all these anomalies and at the same time because of the previous extensive surgery. The procedure started according to the usual flank transperitoneal approach with the patient lying on her left side. An extensive adhesiolysis was necessary before proceeding with the usual mobilisation of the liver from the posterior peritoneal fascia. The small sized right IVC was exposed and the adrenal vein clipped and divided at its confluence (Fig. 5). Then exposure of the adrenal gland continued from the surrounding tissues down towards the renal vein: it was at this point that another adrenal vein was discovered entering the right renal vein (Fig. 6). Also this branch was clipped and divided and the intervention was finally concluded. Postoperative course was uneventful and the patient discharged in day 3. The surgical specimen proved to be a cortical adenoma at the histological examination. At one year of follow up the patient is well and continuing her surveillance checks, on account of FPC.

Discussion

Since its introduction by Gagner in 1992 LA proved to be a very safe and effective procedure in the surgical treatment of many adrenal diseases; in spite of few exceptions, most Authors consider LA as the gold standard of all functioning and non functioning benign tumors provided their size is 6-8 cm large. Early clipping and division of adrenal veins is considered the mainstay of LA since venous bleeding is much more difficult to be handled compared with the hemorrhage of the small adrenal arteries usually amenable to energy based coagulation.

Mainly for this reason a preoperative knowledge of vascular anomalies involving both the IVC, renal and adrenal veins, can be very helpful for a more careful surgical dissection.

Anomalies in the development of IVC and renal veins are extensively reported since the introduction of non invasive imaging tools among which CT angiography is the most frequently adopted. From embryology we know that normal adult right-sided IVC is complete by the 8th week of gestation; its development results from the coordinated growth and regression of a series of veins, the so called posterior, subcardinal and supracardinal veins. Final result comprises a spectrum of anatomical conditions that extends from a normal right-sided IVC to a single left IVC with a double IVC standing in between. Together with IVC also renal veins anomalies are described such as circumaortic or retroaortic renal veins.

Most of these anomalies are asymptomatic, although few reports describe venous dilation of the abdominal walls. In rare cases congenital anomalies of the IVC have been recognised as a possible etiologic factors of unexplained venous thromboemolism in young people. Clinical implications of such anomalies are specially important for radiologists in case of interventional procedures such as insertion of a caval filter, embolization of a spermatic vein and moreover in the sampling of...
adrenal veins in the diagnosis of primary hyperaldosteronism. General surgeons and urologists are also involved in case of IVC anomalies: approach to vascular structures during retroperitoneal surgery in case of anomalous vessels is a potential hazard both in open as well as in laparoscopic surgery. Even if a left IVC can be misdiagnosed as a paracaval lymphadenopathy, the main risk factors are linked with anomalous drainage of renal and adrenal veins. Surgical reports in these cases draw attention to the possible bleeding caused by a lesion of a vein not expected to be in that particular site. IVC anomalies are more frequently observed during renal surgery or in the evaluation of living related renal donors. In the majority of cases each adrenal gland has a single venous outflow, but surgeons should be aware that mainly in the right side two adrenal veins can be encountered at rate around 5-10%. With regards to abnormal drainage of adrenal veins, reports in the literature are not so many; they comprise works based upon studies of radiological imaging where surgery was not necessarily contemplated, as well as surgical experiences describing the very laparoscopic adrenalectomy. Reports on anatomical variations of adrenal veins are not necessarily linked with a congenital IVC anomaly. MacGillivray et al. describe a case of confluence of right adrenal vein into an accessory hepatic vein; Parnaby et al., in a series of 162 laparoscopic adrenalectomies found 5 cases of abnormal venous drainage: 3 patients had two left veins draining separately into the left renal vein, one patient had two right veins draining separately into the IVC and one patient had had two right veins draining into the IVC and right renal vein. The latter case presents the same outline of the one we report, with the only difference that in the present case a double IVC was associated.

It is worth to note that in Parnaby’s experience adrenal vein variants were present in 4 cases of pheochromocytomas and in a single case of adrenocortical carcinoma. A case of adrenal ganglioneuroma with a left IVC is reported by Qureshi and Medhi: these Authors stress the importance of preoperative imaging that facilitates surgical resection and avoided catastrophic vascular injury. In spite of some difference, this very consideration can be made also in our experience: awareness of a double (or left) IVC made us suspicious of other anomalies even if CT could not prove the existence of a double adrenal venous drainage. As a matter of fact a clear demonstration of adrenal veins anomalies can not be obtained by the commonly employed angioCT and MNR except in case of a huge adrenal mass. A sure prove of course and flow of adrenal veins is reported only by selective angiographic studies such as that from Sebe et al., who in 88 venograms found anatomical variations in the drainage of adrenal vein with a frequency of 5% on the right and 6% on the left. For these reasons a surgeon facing laparoscopic surgery of adrenal gland must be aware that up to 10% of the cases a double vein can be found; such anomaly should be expected in relation to a large size of the gland, in case of pheochromocytomas and when other concomitant vascular anomalies are also present. Even if preoperative detailed images of a double adrenal vessel are not always available, the very laparoscopic approach with its magnified view makes dissection and recognition of anatomical structures easier and safer. According with our experience, in spite of the well known IVC duplication we could only suspect a further anomaly: double adrenal vein was so identified thanks to the careful dissection made possible by the fine laparoscopic vision.

**Conclusion**

In the present report we observed a double variation from normal anatomy, that is a duplicated IVC and a double adrenal vein. While caval anatomy was already known at the time of surgery, the other finding, even if highly suspected prior to surgery, was detected only during laparoscopy. Accurate surgical dissection was so required in order to avoid vascular damage. Updated imaging techniques are very important to evaluate vascular anomalies in the setting of surgery but can not obviate the excellent view available during laparoscopy.

**Riassunto**

L’eventualità di un’emorragia intraoperatoria è la complicanza più temuta nel corso di una surreneectomia laparoscopica. Tale rischio diventa ancora più importante nel caso in cui siano presenti anomalie anatomiche rilevate da duplicità della Vena Cava Inferiore (VCI) e da anomalie confluenze delle vene surrenali. Tale riscontro, osservato nel caso descritto, ha fornito lo spunto per uno studio sull’incidenza di dette anomalie e sulle metodiche utili a prevenire la complicanza emorragica.

Una donna di 39 anni è giunta alla nostra osservazione a causa di una neoplasia non funzionante del surrene destro andata incontro ad un lento ma progressivo accrescimento volumetrico. Tale neoformazione era stata riconosciuta a distanza di tempo nel corso di controlli clinicamente utili a prevenire la complicanza emorragica. 

La neoplasia è stata sottoposta in altra sede a Proctocolectomia Restorativa circa 8 anni prima. Già in quell’epoca era stata documentata una duplicità congenita della VCI associata ad altre anomalie vascolari e viscerali. Nel corso della surreneectomia laparoscopica, dopo aver cliippato e sezionato la vena surrenalenale alla sua confluenza cavale, è stata riconosciuta una seconda vena con sbocco nella vena renale destra; anch’essa trattata in modo analogo e con esito favorevole.
La presenza di una duplice VCI, pur essendo un reper-to piuttosto raro, è divenuta di più frequente riscontro con la diffusione di tecniche di diagnostica per immagini sempre più elaborate, in particolare l’AngioTC. La presenza di tali anomalie può rendere problematiche alcune manove di radiologia interventistica quali il posizionamento di un filtro cavale o il cateterismo selettivo di una vena surrenali. È però in ambito chirurgico che la duplicità della VCI può causare i maggiori inconvenienti, rappresentati da un possibile danno vascolare. La maggior parte degli Autori ritiene in proposito indispensabile un valido studio pre-operatorio quale strumento atto a prevenire tali lesioni. Nell’esperienza personale lo studio pre-operatorio, pur confermando l’anormalità della VCI non aveva consentito il riconoscimento della doppia vena surrenali. I dati della letteratura riportano la presenza tale anomalia fino al 10% dei casi, soprattutto in caso di feocromocitoma e di masse surrenali di maggiori dimensioni. La diagnosi per immagini pre-operatoria pur importante, deve affiancarsi a una scrupolosa dissezione chirurgica, quest’ultima è resa possibile grazie all’eccellente visione fornita dalle immagini laparoscopiche e al più facile riconoscimento dell’anatomia venosa.

References


