

Open adrenalectomy surgery: an obsolete technique?

About a case of Conn's syndrome



Ann. Ital. Chir., 2008; 79: 47-52

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Open adrenalectomy surgery: an obsolete technique? About a case of Conn's syndrome

The authors report a case of an adenoma of Conn brought to their attention and treated by them in open surgery. The forms of primary hyperaldosteronism are essentially referable: 1) to the adenoma producing aldosterone, responsive to the ACTH, which represents the most common form (60-70%), responsive to surgery and with full recovery (70%) of cases. 2) to the bilateral hyperplasia, responsive to the angiotensine II, 25-30% of the cases, susceptible to medical therapy with receptorial antagonists of aldosterone. The diagnosis, based on the suspected clinical symptomatology, is determined by laboratory and imaging studies. Laparoscopic surgery is the orientation for this pathology. Comparative studies have shown that laparoscopic surgery adopted by experienced surgeons can treat the adrenal pathology in a miniminvasive way, with good results for effectiveness and safety, and for these reasons the laparoscopic treatment is considered the gold standard for this pathology. But adrenal pathology is rare, 4% in people suffering hypertension, and from 0.35 to 4.4% in tumours. There are, however, only a few medical centres which have the possibility of observing a high number of such a pathology with a consequent accepted learning curve for laparoscopic treatment. For this reason the authors believe that "open" surgery in adrenal pathology is not obsolete and that surgical teams which have experience in retroperitoneal pathology must intervene adopting a laparotomic approach, with the aim to operate the sick person, who totally confides himself to the surgeon, in an appropriate way.

KEY WORDS: Conn's syndrome, Laparoscopic adrenalectomy, Laparotomic adrenalectomy.

Introduction

Conn's Syndrome is a benign and rare neoplasia of the adrenal gland, functioning under the secretion of aldosterone. Frequently unilateral, it was described the first time by Jerome W. Conn in 1955¹, who underlined it in a young woman suffering from hypertension and hypokalemia and recovered after the removal of an adrenal tumour of 4 cm, secreting aldosterone.

Clinical case

A 54 year old female patient, in therapy with antihypertensive drugs for about 10 years, is taken to First Aid. She complained intense asthenia, nausea, atypical

thorax aches and palpitations. The analyses of First Aid underlined kalemia of 1.87 mmol/l, CK 2659 U/l, LDH 519 U/l, urea 89 mg/dl, creatinina 2.68 mg/dl. The admission diagnosis was hypokalemia and she was then

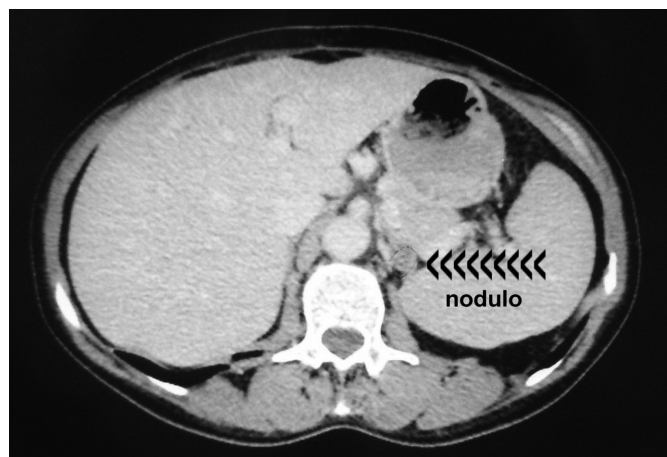


Fig. 1: CT scan: nodule hypodense to level of the side arm of the adrenal left

Pervenuto in Redazione Novembre 2006. Accettato per la pubblicazione Marzo 2007.

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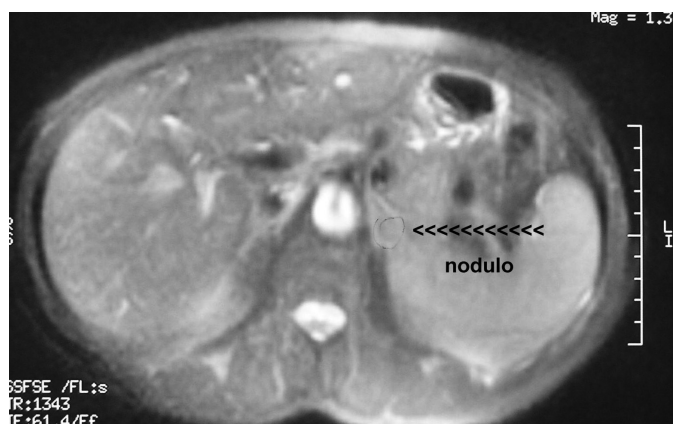


Fig. 2a: MRI scan in sequence SSFSE T2 w: nodule isointense to level of the side arm of the adrenal left.



Fig. 2b: MRI scan in sequence in opposition of phase: absence of signal.

hospitalized in the Division of Medicine. A nephrologic consultation set for diagnostic “excess syndrome of mineral corticoids”, with initial therapy of KCl 140 mEq in physiological to 30 ml/h, application of dosing urinary electrolytes, renin, angiotensine, aldosterone and ACTH, besides the thyroid hormones and proteinuria of Bence-Jones for investigations of advanced imaging. The U.S. didn't underline pathology on behalf of the retroperitoneals organs. The dosing of the aldosterone with RIA method was 209 pg/ml, the renin 5 pg/ml and the ACTH 21 pg/ml. The C.T. scan with contrast medium showed a roundish solid lesion of the left adrenal gland, with a 11 mms diameter, clean contours, hypodense in direct examination, with some impregnation of contrast medium, it all suggested adrenal adenoma (Fig. 1).

The MRI scan of abdomen, focused on the adrenal glands, which integrated the CT scan, performed with SSFSE and FSPGR sequences in phase and opposition of phase, before and after contrast medium, all made on the axial plan, underlined a nodule of around 1 cm, whose irregular behaviour, in the sequences in opposition of phase it reduced, was diagnostic for adenoma. After contrast medium, enhancement was not observed.

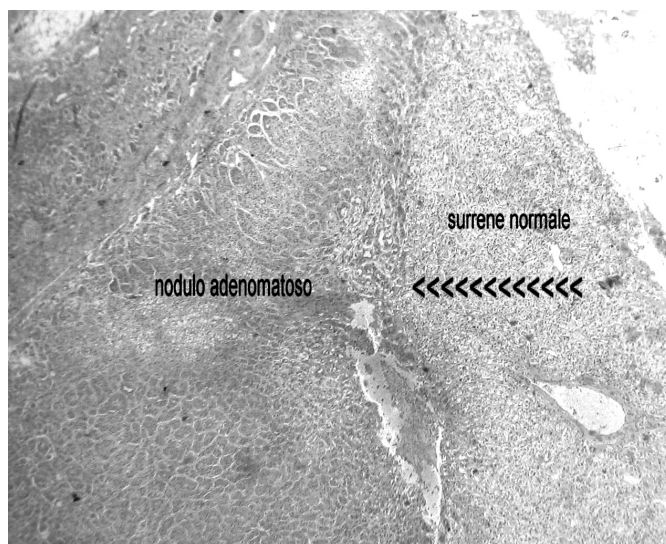


Fig. 3: Coloration E.E., enlargement 4x.

The dimension of the nodule in association with the clinical-laboratoristic descriptions accorded with the suspect of adenoma of Conn (Figs. 2a-2b).

The patient was taken to surgery for treatment of her case. Submitted to open adrenalectomy with left sub costal incision and access to the adrenal loggia through omental retro cavity through section in toto of the gastrocolic ligament. Discharged 10th day, she recovered with normalization of the biochemical kalemic situation. The histological examination showed a centre-nodular area of 2.5 cm contained in the thickness of the capsule, constituted by cellular elements with low nuclear grading, in absence of areas of necrosis and irrelevant mitotic index, the histological picture was compatible with adrenal adenoma (Fig. 3). After two years the biochemical follow-up is in the norm.

Discussion

The aim of this study is not only to introduce a clinical case, but from this case open a debate on the laparoscopic or the traditional procedures that are daily used in approaching surgical pathology in our hospitals, naturally having matured a sound experience in this field and give a clear indication on the matter.

The forms of primary hyperaldosteronism are essentially referable:

- to the adenoma, producing aldosterone, responsive to the ACTH ²,
- to the bilateral hyperplasia, responsive to the angiotensine II ³.

The incidence of this pathology constitutes the 4% of the whole population of the hypertensive ⁴ and 10% in selected populations ⁵.

The primitive hyperaldosteronism from adenoma represents the most common form, 60-70% of all cases, and

is more frequent in women between the third and fourth decades of life. The idiopathic form, sustained by bilateral hyperplasia, is seen in 25% - 30% of the cases, and affect, with a higher frequency, the male sex between the fourth and fifth decades.

The symptomatology is predominantly constituted by sist-diastolic arterial hypertension, of middle or even elevated degree, accompanied by symptoms due to the hypokalemia, such as asthenia, tiredness, muscular weakness, polyuria, above all nycturia, with elevated levels of aldosterone and low or normal levels of renin.

Of fundamental importance is the differentiation between the primitive form, sustained by adenoma, responsive to the surgical therapy and subject to possible recovery in the 70% of the cases ³, and the idiopathic form, sustained by bilateral hyperplasia, susceptible of medical therapy with receptorial antagonists of the aldosterone.

In more recent years, diagnostic advanced imaging (US, CT scan, MRI scan) has shown, in a non aggressive way, the functioning adrenal formations, otherwise diagnosed only with clinical-laboratoristic data or with invasive investigations such as the selective venous catheterism of adrenal veins ⁵⁻⁷.

The differentiation of subtype can require more use of instrumental investigations, firstly high resolution CT scan and with thin layer, which is the technique with the best sensibility in identifying the presence of adrenal nodules, esteemed from 58% to 75% ^{5,6}.

The MRI scan has a smaller sensibility in comparison to the CT scan ⁸, but it is useful in patients that have side effects to the use of the contrast medium.

Such methods have totally supplanted the selective venous catheterism of adrenal veins, that represented the gold standard in the diagnosis of subtype of the primitive hyperaldosteronism, due to the fact that such a procedure is difficult to carry out because the adrenal veins are small, cannulation is difficult, and while to the left it flows to a straight angle in the renal, to the rights it flows to acute angle in the cave and becomes difficult to cannulate.

The treatment is surgical. Such indication is extended to all the functioning adrenal neoformations, Conn's Syndrome from secreting adenoma, Syndrome of Cushing, disease of recidivous Cushing after hypophysial surgery, pheochromocytoma, incidentalomas or not functioning adrenal masses of diameter superior of 3-4 cm that show progressive enlargement in the CT scan or MRI scan at intervals of 6 months.

Since Gagner, in 1992, performed the first removal of the adrenal gland by laparoscopic transperitoneale access for hyperacting illness ⁹, the treatment of such pathologies by the same way of access or different ways of access, retroperitoneal lointomic according to Gaur ¹⁰, or with completely back access with patients prone on the operating table ¹¹⁻¹³, has become principally laparoscopic, relegating to an ancillary position the "open" surgical access and only and exclusively for the removal of adrenal

malignant neoplasias, the removal of retroperitoneal masses with a diameter > 7-8 cm and with previous operations on the superior quadrants of the abdomen, or in presence of possible pathologies associated with the kidney, or associated with severe obesity.

Indeed different comparative studies have been completed, which have shown how laparoscopy allows the treatment of the greater part of adrenal pathology with a very less invasive approach in comparison to the traditional surgery, but of identical effectiveness and safety, to such point that today, in all the scientific congresses and in literature, laparoscopic adrenalectomy is considered the "gold standard" for the treatment of non neoplastic adrenal masses ^{14,15}.

But what is a "gold standard" intervention?

From our point of view, a gold standard is a surgical procedure which must be easily reproduced and extended to the majority of the surgeons; in such a light laparoscopic colecistectomy is a "gold standard", because it is practiced today by the large majority of the surgeons because of the high frequency of such a pathology in every hospital.

Adrenal pathology is a rare pathology, 4% in the hypertensive people ⁴, from 0.35% to 4.4% in the tumors ¹⁶, therefore, there are only a few medical centres that have the opportunity of having a conspicuous amount of such a pathology that allow a learning curve of laparoscopic treatment. From the data of literature and from the congressional communications available, in fact, there are not so many centres in Italy which treat such a pathology. In Campania there are only 2 or 3 centres, and all these are situated in the main town of the Region.

What must be done then, if such an eventuality occurs? Must we send the patients to a reference hospital without regards as to the inevitable waiting list, and the obvious problems in transferring patients, which is not always easy? Must we consider the fact that patients in doing so will not have the family nearby? Must we consider the increase of expenses for the family and for the local ASL, with the transfer of financial resources to other hospital structures? Or face the problem in a traditional surgical way, if one has experience of retroperitoneal surgery? Personally we believe that the surgical techniques are never obsolete, and every surgeon has to practise, with precision and correctness, in respect to patients generally, the best surgery that he/she has learned to do, telling them that different techniques do exist and can be used, but in other hospital structures and bearing in mind the questions previously indicated.

But if the experimentation of new techniques is correct in Centres of Excellence, and correct also the finding of new ways and new procedures to improve the status of the individual in his relationship with his/her illness, it is also correct, according to us, not to impose the capillary diffusion of new procedures that necessarily need medical training to be correctly acquired, and last but not least the possibility of having the right amount of

such a pathology to facilitate this medical training; the right amount that not all the hospital structures have. Perhaps it would be far more correct that the Scientific Societies, during the various congresses say a word in such a direction, indicating the good and positive things that are being achieved, but also complications and failures, and abstain themselves from labelling as a gold standard what cannot be practiced in all hospital structures.

Conclusion

The enormous technological and innovative progress sets, every day, a new way in not only treating the individual patient but also the related pathology. This certainly determines a different and new strategy in dealing with ill patients, and obliges surgeons to a continuous run in technological updating and to a constant apprenticeship. Today it is the patient who asks the surgeon to be "operated with keyholes" to have a sick gallbladder removed. The same thing will happen for other pathologies, which means that the surgeon will need constant medical training to acquire the confidence and the skills which are requested when employing new technical techniques. Congress Meetings declare a minimum of 50 operations per year per pathology as being the basic requirement for an accepted learning curve for the acquisition of a new technique. In Medical Centres where this minimum number of operations is not reached, the surgeon must follow and undertake periodical stages in training in more experienced centres, hoping that the financial resources will allow the updating of the medical diagnostic equipment where he/she works. Nevertheless, the quick run towards new procedures and methods doesn't necessarily mean that we put apart what has been taught us by the great surgeons of the past. We must be fully aware that our aim is to operate well, in an honest way, therefore respect a patient who trusts us and confides in our capability, no matter what technique we adopt, and asks us to be treated in a general caring way, and naturally asks us not to be considered as single anatomic pieces.

Riassunto

Gli autori riportano il caso di un adenoma di Conn occorso alla loro osservazione e da essi trattato chirurgicamente in via open.

Le forme di iperaldosteronismo primario sono riconducibili:

- all'adenoma produttore aldosterone, responsivo all'ACTH, che rappresenta la forma più comune (60-70%), frequente nella donna tra la III^a e IV^a decade di vita, responsiva alla terapia chirurgica e suscettibile di guarigione (70%) dei casi;
- all'iperplasia bilaterale responsiva all'angiotensina II ³, compresa tra il 25-30% dei casi, colpisce maggior-

mente il sesso maschile tra la IV^a-V^a decade, suscettibile di terapia medica con antagonisti recettoriali all'aldosterone.

La sintomatologia è costituita da ipertensione arteriosa accompagnata da sintomi legati all'ipokaliemia quali astenia, stanchezza, debolezza muscolare, poliuria soprattutto notturna, con elevati livelli di aldosterone e bassi o normali livelli di renina.

La diagnosi si basa sul sospetto clinico sintomatologico, ma è laboratoristica e strumentale. Negli ultimi anni l'imaging avanzata (US, TAC, RMN) ha permesso, in maniera non cruenta, la evidenziazione di formazioni surrenaliche funzionanti, altrimenti diagnosticate solo con dati clinico-laboratoristici e con indagini cruente quali il cateterismo venoso selettivo delle vene surrenaliche, peraltro, di non facile esecuzione perché le vene surrenaliche sono di piccole dimensioni e non facilmente incannulabili.

L'orientamento attuale è di operare in via laparoscopica la patologia di tale organo. Da quando Gagner, nel '92, ha eseguito la prima surrenalectomia laparoscopica, diversi Centri hanno sperimentato tale strada e da allora la patologia del surrene viene affrontata con approccio esclusivamente laparoscopico, riservando la via open solo per l'asportazione di masse surrenaliche con diametro > 7-8 cm, o con patologie renali associate o con precedenti interventi addominali od obesità grave. Diversi studi comparativi hanno dimostrato come la laparoscopia permetta di trattare la patologia surrenalica in via mininvasiva con risultati analoghi per efficacia e sicurezza, in mani esperte. Effettivamente oggi, in tutti i Congressi Scientifici ed in letteratura, la surrenalectomia laparoscopica è considerata il gold standard per il trattamento delle masse surrenaliche non neoplastiche.

Ma che cosa è un intervento gold standard?

A nostro avviso, gold standard deve essere considerata una tecnica chirurgica facilmente riproducibile ed esportabile alla maggioranza degli operatori, in tale ottica consideriamo la colecistectomia laparoscopica "gold standard", in quanto praticata oggi dalla maggioranza dei chirurghi, per l'alto volume di tale patologia afferente presso ogni plesso ospedaliero. La patologia surrenalica, di contro, è patologia rara, 4% negli ipertesi ⁴ e dallo 0,35% al 4,4% nei tumori ¹⁶, pertanto, i Centri che hanno l'opportunità di osservare un volume cospicuo di tale patologia che permetta di affrontare una curva di apprendimento del trattamento laparoscopico sono pochi. Cosa fare, se si verifica una tale evenienza?

Dirottare il paziente verso un centro di riferimento, con inevitabile lista di attesa, disagio per il trasferimento non sempre agevole, impossibilità di avere i familiari vicino, aggravio di spese per la famiglia ed a carico della propria ASL con trasferimento di risorse a favore di altre strutture? affrontare il problema in modo chirurgico tradizionale se si ha la necessaria esperienza chirurgica? Noi riteniamo che le tecniche chirurgiche non siano mai obsolete e ciascuno deve praticare la chirurgia migliore che ha imparato, avvertendo che si possono praticare

anche tecniche diverse presso altre strutture ma con i disagi di cui sopra. Oggi è il paziente a chiedere spesso di essere "operato con i buchi", ma indipendentemente dalle tecniche utilizzate, chiede di essere visto nella sua globalità di essere umano e non parcellizzato in singoli pezzi anatomici. Il nostro compito resta la precisione e la correttezza nei suoi confronti ed un continuo apprendistato per acquisire confidenza e manualità di padronanza nelle nuove tecnologie.

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