Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Digestive Surgery

Adrenocortical Tumor: With Special Reference to A Case Report

Moufid Abdellah[1*], Dahbi Skali Laila[1], Iliass Maouni[1], Abdellatif Settaf[1]

[1]Digestive Surgical Department B, Ibn Sina University Hospital, Rabat, Morocco

DOI: 10.36347/sjmcr.2022.v10i09.001 | **Received:** 18.07.2022 | **Accepted:** 27.08.2022 | **Published:** 02.09.2022

***Corresponding author:** Moufid Abdellah
Digestive Surgical Department B, Ibn Sina University Hospital, Rabat, Morocco

Abstract **Case Report**

Adrenocortical tumor (ACT) is a rare tumor, the annual incidence of which is estimated to be 0.5 to 2 cases per million [1], leading to 0.04 and 0.2% of causes of cancer death.We report here the case of a patient admitted in the surgery department B at Ibn Sina hospital in Rabat.This is a 50-year-old man, a chronic smoker with 33 smoking years, who underwent an operation a month ago for suspicion of a Hydatid Cyst of the right lobe of the liver, with intraoperative finding of a right adrenal mass and a normal liver. Biopsy later on reported a malignant adrenocortical tumor, so the patient was referred for subsequent care and management. The patient subsequently underwent surgery by laparotomy, and complete resection of the adrenal tumor mass was successful, followed by simple postoperative consequences.

Keywords: adrenal neoplasms, adrenocortical carcinoma, endocrine disorders, pheochromocytoma, tumor surgery.

INTRODUCTION

Adrenocortical tumor (ACT) is a rare tumor, the annual incidence of which is estimated to be 0.5 to 2 cases per million [1], leading to 0.04 and 0.2% of causes of cancer death.

ACT is one of the most serious malignant endocrine tumors with metastases at the time of diagnosis occurring in 20 to 40% of the cases, most often to the liver, lungs or bones [2].

There are two frequency, a first in childhood, and the second is between 40 and 50 years old, with a slight female predominance (sex ratio 1.5) [2].

On the clinical level, we can have 3 tables:

- Nearly half of ACT s is discovered due to clinical signs of hormonal hypersecretion. Examples include, from the most common to the least common, mixed hypersecretion of cortisol and androgens, isolated hypersecretion of cortisol, virilization by androgenic hypersecretion or different associations of steroid hypesecretion [2].
- A mass syndrome is frequently present in the case of non-secreting ACT. In this case, the ACT s have an average weight and height on anatomo-pathological examination of more than 500 grams and 12-15 cm [2, 4]. Symptoms may include abdominal or lumbar pain, a palpable mass, weight loss, tumor fever, or signs of inferior vena cava compression. It should be noted that the general condition of patients with ACT is asymptomatic for a long period of time, explaining the often late diagnosis of these tumors.
- In much more rare cases, the diagnosis of ACT is incidentally discovered during an imaging performed for another indication. Approximately 10% of adrenocortical tumors are discovered incidentally, most often at early stages [2]. It is important to note that the probability that an adrenal mass is malignant increases with its size [5]: it is extremely low for lesions of less than 4 cm and around 15 to 20% for lesions of more than 6cm.
- Recently, an incidental finding of ACT is increasing [6], although this observation is not always confirmed, more particularly across the Atlantic [7]. The discovery of these malignant lesions at an early stage is a major challenge for management.
- Finally, it is not uncommon for a ACT to be discovered as part of the assessment of its metastases.

On the biological level: a complete hormonal and biochemical assessment is necessary to avoid

missing a particularly important infraclinical hypersecretion for appropriate management: subsequent follow-up (tumor markers) and immediate postoperative hormonal substitution.

In addition to the complete standard biochemical assessment, a hormonal assessment is necessary with: a cortisol level at 8 a.m., urinary free cortisol (FLU), serum or salivary nocturnal cycle of cortisol (the most important of which is the midnight level), ACTH levels, dexamethasone suppression test, plasma aldosterone and renin activity, testosterone levels, dehydroepiandrosterone sulfate, Delta4 androstenedione, estradiol, intermediate derivatives (17-OH progesterone, deoxycorticosterone, compound S), and urinary methylated derivatives [2].

In case of ACT suspicion, required imaging modalities include: a thoraco-abdomino-pelvic CT [8], an MRI [9, 10], and a PET scan [11, 12]. In case of mets to the inferior vena cava, these examinations must include sufficient contrast material with time delay to clearly delineate the relationship between the lesion and the vena cava.

Management: Currently, radical surgery is the only curative therapeutic option [13], but there is a high risk of postoperative recurrence (more than 85%).

There are few options for adjuvant treatments and their effectiveness is uncertain.

Radical surgery is the only hope for a cure, but the overall 5-year survival for all stages varies between 30 and 40% [4] and depends mainly on the stage of the tumor, which is now staged according to the European Network classification for the Study of Adrenal Tumors (ENSAT) [14]

CASE REPORT

We report here the case of a patient admitted in the surgery department B at Ibn Sina hospital in Rabat.

This is a 50-year-old man, a chronic smoker with 33 smoking years, who underwent an operation a month ago for suspicion of a Hydatid Cyst of the right lobe of the liver, with intraoperative finding of a right adrenal mass and a normal liver. Biopsy later on reported a malignant adrenocortical tumor, so the patient was referred to surgery B unit for subsequent care and management.

The patient had Right Upper Quadrant pain upon physical examination, as well as pain at the level of the epigastrium, associated with a fixed palpable mass.

Laboratory examination was normal except for mild anemia with a Hb level of 10g/dl.

Normal hormonal balance (a cortisol cycle levels, methylated drifts, etc.).

Abdominal CT scan: a hypodense right adrenal mass heterogeneously enhanced after injection of contrast product measuring 15.5cm, exerting a mass effect on the liver and the right kidney. This mass touches the inferior vena cava and the duodenopancreatic junction (Figure 1).

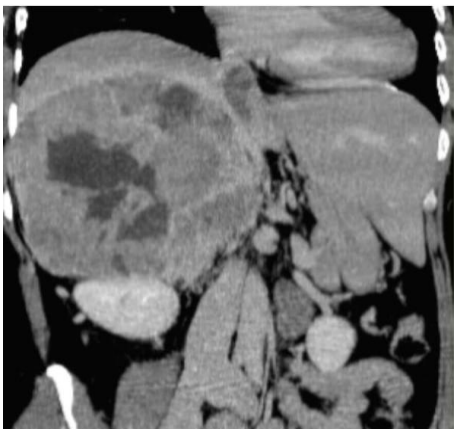

Figure 1

The patient subsequently underwent surgery by laparotomy, and complete resection of the adrenal tumor mass was successful, followed by simple postoperative consequences (Figure 2 and 3).

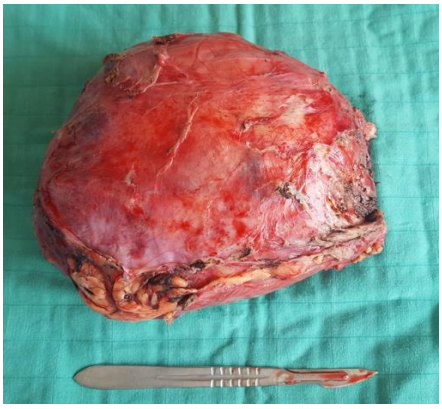

Figure 2

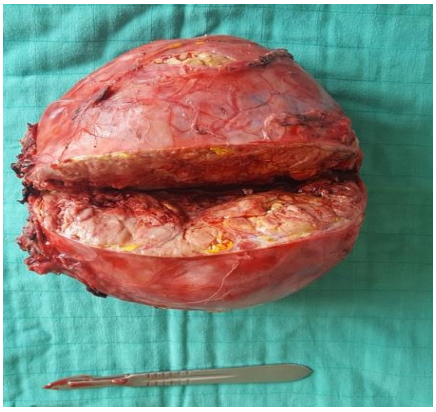

Figure 3

Pathological examination revealed an adrenocortical tumor.

Biopsies from the organs in contact with the tumor (kidney, liver, duodenum colon, etc…) revealed no tumor metastases.

DISCUSSION AND ANALYSIS

The controversy remains the surgical excision of any potentially resectable ACT [16, 17],based on the ENSAT classification, which is now more widely used.

For stage I and II tumours, excision of the adrenal gland and the cellulo-nodular compartment is the norm. Excision is also indicated in stage III of the ENSAT classification, with loco-regional invasion, including lymph node extension to neighboring organs or vena cava or in loco-regional recurrences.

The resection of ACT is in the majority of cases is a complex open surgery requiring the widening of the excision to neighboring organs that can be invaded. However, ACT surgery can, in expert centers, for lesions of less than 6 cm, be made by laparoscopy.

Excision is more controversial for metastatic stage IV for which recurrence is normally very common [18]. Metastases to close organs are present in 17–42% [18, 19], mostly affecting the liver or lung.

The efficacy of palliative chemotherapy in stage IV ACT is very moderate. The first-line gold standard, determined by a prospective international randomized trial, combines chemotherapy based on etoposide, doxorubicin, cisplatin and mitotane [20].

In an Italian phase II trial, 10 (13%) of the 72 patients with unresectable metastatic ACT successfully underwent surgery after an objective response to chemotherapy combined with mitotane [21].

Given the scarcity of ACTs, it is preferred that they are managed in referral centers. However, the definition of a referral center is not well established [22-24].

Beyond the technical expertise, it is necessary to highlight the importance of multidisciplinary team management of these patients, with teams consisting of surgeons, endocrinologists, oncologists, radiologists, pathologists, biologists and radiation therapy specialists.

In stage I and early stage II cases, the benefit of laparoscopy is still the subject of debate [25,26]. While the laparoscopic approach has become, based on experts' consensus, the reference technique for excision of benign adrenal tumors [28], this is more controversial for ACT. Studies are limited and no consensus has yet been found [25-27], but, for some teams, laparoscopy is associated with an increased risk of capsular rupture [25], loco-regional recurrence or peritoneal carcinomatosis.

The severity of capsular invasion and the bad prognosis of ACT should prioritize the importance of total excision over the choice of approach.

However, for stage I and early stage II, laparoscopic resection of a ACT can be proposed, on condition that a malignancy is already diagnosed preoperatively, a well experienced laparosocopic surgeon, and only for lesions of less than 6 cm [15]. In case of pre- or intraoperative doubt, conversion to laparotomy should be performed.

Most ACTs are diagnosed at later stages, as locally advanced stage II, III or IV tumors depending on the existence of locoregional or metastatic extension, with weight and size characteristics that prevent the laparoscopic approach. The main objective of ACT oncologic surgery is to obtain complete R0 excision, including a minimum of lymph node dissection of the renal, without tumor invasion, in order to minimize the risk of loco-regional recurrence. The risk of tumor invasion should not be underestimated and is favored by the size of the ACT s, their depth, the existence of a fragile capsule, the presence of hypervascular adhesions with the adjacent parenchymal organs and the fragility of the tissues. Cases of cortisol hypersecretion necessiatte an approach allowing good exposure and atraumatic tumor manipulation [29, 30].

The opening of invaded anatomical planes between the ACT and the neighboring organs (kidney, diaphragm, liver on the right, spleen, pancreas on the left) seems to be an important factor in loco-regional recurrence, by capsular rupture, hemorrhage, or insufficient margins [31, 32].

The intra operative determination of this invasion is often difficult. In 30% of cases, tumors initially classified as stage II are reclassified as stage III after anatomopathological examination has demonstrated invasion of adjacent tissues [33].

Cases of locally advanced tumors of the adrenal gland may therefore require surgery with complete resection removing of the tumour, the kidney, the invaded adjacent organs, and all the periadrenal and perirenal fat. Data showing a benefit of extensive lymph node dissection are few, but one study showed a 73% lymph node invasion rate in ACT and a recent study suggests a reduced risk of local recurrence and mortality related to ACT when dissection removes at least 5 lymph nodes [34]. Lymph node dissection also allows for more accurate disease staging, although its

influence on survival remains uncertain [35]. Thus, lymph node dissection must be performed and must include at least the periadrenal and perirenal fat by removing the renal hilar nodes. The benefit of latero-aortic and inter-aortic-cellar dissection is even more debated and this dissection should not be taken into consideration.

In case of hepatic invasion, resection may require right hepatectomy. On the left, it may require dissection of the pancreatic isthmus as a primary approach to the spleno-mesaraic venous confluence and the visceral aortic junction.

In more favorable situations (voluminous stage II), an extensive adrenalectomy removing all the adrenal and perirenal fat and carrying out a complete loco-regional lymph node dissection and of the renal hilum makes it possible to preserve the ipsilateral kidney. The excision of a major part of the ipsilateral kidney should be avoided, except if it is necessary to obtain an R0 margin, in particular for tumors placed on the renal pedicle, regardless of their size.

Vena Cava extension, present in less than 10% of patients, and linked to the venous extension of a tumoral thrombus most often progressing through the main adrenal vein, is more frequent for right ACT.

It is more often a vena cava thrombus tumor than tumor extension to the vena cava wall, with a high risk of hepatic and/or pulmonary metastases [36].

Depending on the upper level of the thrombus (sub-hepatic VCI, retro-hepatic VCI, supra-hepatic VCI), thrombectomy may require sub-hepatic supra-renal vena cava clamping, vascular exclusion with sub-diaphragmatic or intra-pericardial supra-hepatic vena cava clamping, or, exceptionally, extracorporeal venous circulation in the event of intra-auricular extension [36].

Venous invasion of the vena cava wall is often limited and accessible to partial resection with direct closure or with the help of a prosthetic, autologous (peritoneum), or heterologous (bovine pericardium) patch.

Post op Monitoring

There is no study validating a monitoring protocol, a certain number of recommendations exist [15]:

Immediate postoperative

For secreting tumours, hormonal assays must be carried out following surgery, to ensure that the excision has been complete.

From a distance

Biology

For any tumor, a complete hormonal assessment must be carried out every 3 months, including cortisol (urinary cortisol, low braking test), androgens (S-DHEA, androstenedione, testosterone), and precursors.

The elevation of these markers indicates tumor recurrence.

Imaging

A thoraco-abdomino-pelvic CT scan, without and with injection of contrast product, must be performed in search of loco-regional recurrence or distant metastases every 3 months for 2 years, then every 3 to 6 months for 3 years, then annually.

The scanner is combined with an 18F-fluoro-deoxyglucose PET-scan every 3 months.

Depending on the call points, a cerebral MRI and a bone scan are performed.

IN CONCLUSION

CS is a rare tumor of the adrenal gland which develops at the expense of the adrenal cortex, the diagnosis of which is often late with a more or less poor prognosis, the reference treatment is surgery, and the quality of the initial excision represents one of the factors major predictions. However, the quality criteria for this surgery are still poorly defined, due to the very low incidence of the disease.

BIBLIOGRAPHY

1. Lindholm, J., Juul, S., Jørgensen, J. O. L., Astrup, J., Bjerre, P., Feldt-Rasmussen, U., ... & Weeke, J. (2001). Incidence and late prognosis of Cushing's syndrome: a population-based study. *The Journal of Clinical Endocrinology & Metabolism*, *86*(1), 117-123.
2. Abiven, G., Coste, J., Groussin, L., Anract, P., Tissier, F., Legmann, P., ... & Bertherat, J. (2006). Clinical and biological features in the prognosis of adrenocortical cancer: poor outcome of cortisol-secreting tumors in a series of 202 consecutive patients. *The Journal of Clinical Endocrinology & Metabolism*, *91*(7), 2650-2655.
3. Luton, J. P., Cerdas, S., Billaud, L., Thomas, G., Guilhaume, B., Bertagna, X., ... & Bricaire, H. (1990). Clinical features of adrenocortical carcinoma, prognostic factors, and the effect of mitotane therapy. *New England Journal of Medicine*, *322*(17), 1195-1201.
4. Icard, P., Goudet, P., Charpenay, C., Andreassian, B., Carnaille, B., Chapuis, Y., ... & Proye, C. (2001). Adrenocortical carcinomas: surgical trends and results of a 253-patient series from the French Association of Endocrine Surgeons study group. *World journal of surgery*, *25*(7), 891-897.

5. O'Neill, C. J., Spence, A., Logan, B., Suliburk, J. W., Soon, P. S., Learoyd, D. L., ... & Sywak, M. S. (2010). Adrenal incidentalomas: risk of adrenocortical carcinoma and clinical outcomes. *Journal of Surgical Oncology*, *102*(5), 450-453.
6. Icard, P., Chapuis, Y., Andreassian, B., Bernard, A., & Proye, C. (1992). Adrenocortical carcinoma in surgically treated patients: a retrospective study on 156 cases by the French Association of Endocrine Surgery. *Surgery*, *112*(6), 972-980.
7. Paton, B. L., Novitsky, Y. W., Zerey, M., Harrell, A. G., Norton, H. J., Asbun, H., ... & Heniford, B. T. (2006). Outcomes of adrenal cortical carcinoma in the United States. *Surgery*, *140*(6), 914-920.
8. Korobkin, M., Brodeur, F. J., Francis, I. R., Quint, L. E., Dunnick, N. R., & Londy, F. (1998). CT time-attenuation washout curves of adrenal adenomas and nonadenomas. *AJR. American journal of roentgenology*, *170*(3), 747-752.
9. Korobkin, M., Brodeur, F. J., Francis, I. R., Quint, L. E., Dunnick, N. R., & Goodsitt, M. (1996). Delayed enhanced CT for differentiation of benign from malignant adrenal masses. *Radiology*, *200*(3), 737-742.
10. Prager, G., Heinz-Peer, G., Passler, C., Kaczirek, K., Schindl, M., Scheuba, C., ... & Niederle, B. (2002). Can dynamic gadolinium-enhanced magnetic resonance imaging with chemical shift studies predict the status of adrenal masses?. *World journal of surgery*, *26*(8), 958-964.
11. Ardito, A., Massaglia, C., Pelosi, E., Zaggia, B., Basile, V., Brambilla, R., ... & Terzolo, M. (2015). 18F-FDG PET/CT in the post-operative monitoring of patients with adrenocortical carcinoma. *Eur J Endocrinol*, 173, 749-756.
12. Gust, L., Taieb, D., Beliard, A., Barlier, A., Morange, I., de Micco, C., ... & Sebag, F. (2012). Preoperative 18F-FDG uptake is strongly correlated with malignancy, Weiss score, and molecular markers of aggressiveness in adrenal cortical tumors. *World journal of surgery*, *36*(6), 1406-1410.
13. Schteingart, D. E., Doherty, G. M., Gauger, P. G., Giordano, T. J., Hammer, G. D., Korobkin, M., & Worden, F. P. (2005). Management of patients with adrenal cancer: recommendations of an international consensus conference. *Endocrine-related cancer*, *12*(3), 667-680.
14. Fassnacht, M., Johanssen, S., Quinkler, M., Bucsky, P., Willenberg, H. S., Beuschlein, F., ... & German Adrenocortical Carcinoma Registry Group and the European Network for the Study of Adrenal Tumors. (2009). Limited prognostic value of the 2004 International Union against Cancer staging classification for adrenocortical carcinoma: proposal for a Revised TNM Classification. *Cancer*, *115*(2), 243-250.
15. Gaujoux, S., Mihai, R., Carnaille, B., Dousset, B., Fiori, C., Porpiglia, F., ... & Zinzindohoue, F. (2017). European Society of Endocrine Surgeons (ESES) and European Network for the Study of Adrenal Tumours (ENSAT) recommendations for the surgical management of adrenocortical carcinoma. *Journal of British Surgery*, *104*(4), 358-376.
16. Henry, J. F., Peix, J. L., & Kraimps, J. L. (2012). Positional statement of the European Society of Endocrine Surgeons (ESES) on malignant adrenal tumors. *Langenbeck's archives of surgery*, *397*(2), 145-146.
17. Berruti, A., Baudin, E., Gelderblom, H., Haak, H. R., Porpiglia, F., Fassnacht, M., & Pentheroudakis, G. (2012). Adrenal cancer: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Annals of oncology*, *23*, vii131-vii138.
18. Dy, B. M., Strajina, V., Cayo, A. K., Richards, M. L., Farley, D. R., Grant, C. S., ... & Thompson, G. B. (2015). Surgical resection of synchronously metastatic adrenocortical cancer. *Annals of surgical oncology*, *22*(1), 146-151.
19. Didolkar, M. S., Bescher, R. A., Elias, E. G., & Moore, R. H. (1981). Natural history of adrenal cortical carcinoma: a clinicopathologic study of 42 patients. *Cancer*, *47*(9), 2153-2161.
20. Fassnacht, M., Terzolo, M., Allolio, B., Baudin, E., Haak, H., Berruti, A., ... & Skogseid, B. (2012). Combination chemotherapy in advanced adrenocortical carcinoma. *New England Journal of Medicine*, *366*(23), 2189-2197.
21. Berruti, A., Terzolo, M., Sperone, P., Pia, A., Della Casa, S., Gross, D. J., ... & Dogliotti, L. (2005). Etoposide, doxorubicin and cisplatin plus mitotane in the treatment of advanced adrenocortical carcinoma: a large prospective phase II trial. *Endocrine-related cancer*, *12*(3), 657-666.
22. Gratian, L., Pura, J., Dinan, M., Reed, S., Scheri, R., Roman, S., & Sosa, J. A. (2014). Treatment patterns and outcomes for patients with adrenocortical carcinoma associated with hospital case volume in the United States. *Annals of surgical oncology*, *21*(11), 3509-3514.
23. Lombardi, C. P., Raffaelli, M., Boniardi, M., De Toma, G., Marzano, L. A., Miccoli, P., ... & Bellantone, R. (2012). Adrenocortical carcinoma: effect of hospital volume on patient outcome. *Langenbeck's archives of surgery*, *397*(2), 201-207.
24. Greco, F., Hoda, M. R., Rassweiler, J., Fahlenkamp, D., Neisius, D. A., Kutta, A., ... & Fornara, P. (2011). Laparoscopic adrenalectomy in urological centres–the experience of the German Laparoscopic Working Group. *BJU international*, *108*(10), 1646-1651.
25. Kebebew, E., Siperstein, A. E., Clark, O. H., & Duh, Q. Y. (2002). Results of laparoscopic adrenalectomy for suspected and unsuspected

malignant adrenal neoplasms. *Archives of Surgery*, *137*(8), 948-953.
26. Iino, K., Oki, Y., & Sasano, H. (2000). A case of adrenocortical carcinoma associated with recurrence after laparoscopic surgery. *Clinical endocrinology*, *53*(2), 243-248.
27. Palazzo, F. F., Sebag, F., Sierra, M., Ippolito, G., Souteyrand, P., & Henry, J. F. (2006). Long-term outcome following laparoscopic adrenalectomy for large solid adrenal cortex tumors. *World journal of surgery*, *30*(5), 893-898.
28. Assalia, A., & Gagner, M. (2004). Laparoscopic adrenalectomy. *Br J Surg*, 91, 1259-1274.
29. Jany, T., Chevallier, C., & Pattou, F. (2007). Transthoracic transdiaphragmatic approach for resection or large right sided adrenal tumors. *Journal de Chirurgie*, *144*(6), 527-531.
30. Vanbrugghe, C., Lowery, A. J., Golffier, C., Taieb, D., & Sebag, F. (2016). Adrenocortical carcinoma surgery—Surgical extent and approach. *Langenbeck's archives of surgery*, *401*(7), 991-997.
31. Amini, N., Margonis, G. A., Kim, Y., Tran, T. B., Postlewait, L. M., Maithel, S. K., ... & Pawlik, T. M. (2016). Curative resection of adrenocortical carcinoma: rates and patterns of postoperative recurrence. *Annals of surgical oncology*, *23*(1), 126-133.
32. Margonis, G. A., Kim, Y., Prescott, J. D., Tran, T. B., Postlewait, L. M., Maithel, S. K., ... & Pawlik, T. M. (2016). Adrenocortical carcinoma: impact of surgical margin status on long-term outcomes. *Annals of surgical oncology*, *23*(1), 134-141.
33. Miller, B. S., Gauger, P. G., Hammer, G. D., & Doherty, G. M. (2012). Resection of adrenocortical carcinoma is less complete and local recurrence occurs sooner and more often after laparoscopic adrenalectomy than after open adrenalectomy. *Surgery*, *152*(6), 1150-1157.
34. Reibetanz, J., Jurowich, C., Erdogan, I., Nies, C., Rayes, N., Dralle, H., ... & German ACC Study Group. (2012). Impact of lymphadenectomy on the oncologic outcome of patients with adrenocortical carcinoma. *Annals of surgery*, *255*(2), 363-369.
35. Saade, N., Sadler, C., & Goldfarb, M. (2015). Impact of regional lymph node dissection on disease specific survival in adrenal cortical carcinoma. *Hormone and Metabolic Research*, *47*(11), 820-825.
36. Chiche, L., Dousset, B., Kieffer, E., & Chapuis, Y. (2006). Adrenocortical carcinoma extending into the inferior vena cava: presentation of a 15-patient series and review of the literature. *Surgery*, *139*(1), 15-27.