

Oncological surgery

Elixir Oncological surgery 170 (2022) 56427 - 56431

Elixir
ISSN: 2229-712X

Metastatic Giant Adrenocortical Tumor in the Liver

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ARTICLE INFO

Article history:

Received: 21 July 2022;

Received in revised form:

2 September 2022;

Accepted: 16 September 2022;

Keywords

Adrenocortical Tumor,
Carcinoma,
Metastasis,
Nephrectomy,
Hepatectomy.

ABSTRACT

Adrenocortical tumor is a rare tumor of the adrenal developing depending on the adrenal cortex; most often responsible for an endocrine syndrome. We report a case of a 41-year-old woman who presented with treatment-resistant chronic low back pain, whose radiological exploration revealed a retroperitoneal mass dependent on the adrenal gland measuring 108 x 84 x 88 mm (APxTxH) metastatic of the liver of segment VII and segment VIII, the management of which was surgical in two stages.

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Introduction

The adrenal is an endocrine gland, which can be the site of many benign or malignant tumor pathologies.

The adrenal cortex or carcinoma of the adrenal gland is a very rare malignant tumor, it develops within the adrenal cortex. Its incidence is 0.7 – 2 cases per million inhabitants, more frequent in women with a sex ratio of 1.5, and two incidence peaks: in childhood before 5 years old, and between 40-50 years old(1).

Most often, these are secreting tumors, hormonal hypersecretion is made of cortisol, androgens, estrogens, and more rarely, aldosterone. Sometimes the diagnosis is fortuitous, following abdominal pain associated with a mass discovered on clinical examination or imaging.

In addition, the malignant adrenal cortex has a very significant loco-regional and vascular invasiveness that can lead to thromboses of the inferior vena cava, remote tumor dissemination most often in the liver and lungs. The 5-year survival rate is 35%(2) and the a very high locoregional recurrence rate(3).

Surgical management is the cornerstone in the treatment of malignant adrenocortical tumors.

In this article, we present a case of a metastatic malignant adrenocortical tumor in the liver about a case whose management was surgical.

Clinical case

Mrs. NS aged 41, OMS 1 ASA 1 GN 2, was admitted for exploration of chronic low back pain evolving since 6 months, without other signs associated, clinical examination was unremarkable. Abdominal ultrasound revealed an intraperitoneal formation of heterogeneous tissue 105 mm x 95 mm x 96 mm, containing necrotic tissue. Abdominal CT confirmed the presence of this intraperitoneal formation, well limited, heterogeneous hypodense strongly enhanced after injection of PDC, containing calcifications and areas of necrosis measuring: 108 x 84 x 88 mm (APxTxH). High, she comes into intimate contact with the antropylic region of

the stomach with loss of border of separation by place and pushes back the spleen with respect for the border of separation. Below it pushes back the jejunal loops, the body and the tail of the pancreas, spléno-mesaraïque vein, Down and back she comes in contact with the pole interior of the left kidney with loss of border of separation by place. Outside, she comes into contact with the left colon. Direct and indirect splenorenal bypass route, liver of normal size and morphology, regular contours, the seat of a hypodense lesion straddling segment VII and segment VIII measuring: 8x9cm. VB, VB1H/EH unremarkable. TP, hepatic veins, and midline abdominal vessels are unremarkable.

An echo-guided liver biopsy with an immunohistochemical supplement was performed, showing: Hepatic localization of a poorly differentiated tumoral process whose immunohistochemical profile points towards a cortico-adrenal carcinomatous process. The decision of management was made in CPR: two-stage adrenal and hepatic mass resection surgery.

The surgery (07/10/2020) of primary resection was performed under AG + ALR and with a median approach, to exploration: a tumor of the left adrenal without invasion of the spleno-pancreatic block with synchronous right uni-lobar metastasis not visible on the associated surface has a spontaneous hematoma between the tip of the left lobe and the anterior part of the stomach. The monobloc resection was performed: Extended left adrenal nephrectomy. As for the post-opérative period has been marked by an asymptomatic biological pancreatic fistula on D5 postoperative, revealed by a lipase level in the drains at 3025 ui and a wall infection treated by local care.

The anatomopathological study (22/10/2020) had concluded:

- Carcinoma adrenal cortex (Weiss grade >3) of 13cm major axis.
- classified pT2NoM1
- Stage IV of ENSAT Staging system

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A monitoring consultation (11/04/2020) was unremarkable. Subsequently, a resection of the liver metastasis was scheduled for (07/12/2020) under AG + ALR: the surgical approach was made by a J incision and exposure with 3 metal valves, at intraoperative ultrasound exploration: the hepatic lesion drives back the median hepatic vein and the inferior vena cava without invading them, the operative gesture consisted of a right hepatectomy carrying the median hepatic vein by an anterior approach and whose postoperative course was simple.

The anatomopathological study (28/12/2020) concluded:

- Right hepatectomy specimen of a known adrenocortical metastasis.
- Healthy resection margin 0.1cm

Discussion

1-Epidemiology

Corticosurrenaloma is a rare tumor, the incidence of which is 1 to 2 cases per million individuals (4), with a sex ratio of 1.5 in women, and characterized by two peaks in early childhood or middle age, although it can affect people of all ages. Most cases appear to be sporadic; however, they may be associated with hereditary syndromes, namely multiple endocrine neoplasia type 1 or the syndrome of Li Fraumeni(5).

The overall survival of localized adrenal cortex is estimated at 5 years in 30% to 50% of cases, as for the metastatic form, the overall survival is estimated at 1 year (6).

2- Diagnosis

Clinic

The majority of adrenocortical tumors are tumors that secrete adrenal hormones, so there is hormonal hypersecretion (glucocorticoids and/or mineralocorticoids and/or androgens).

Tumor syndrome defined by pain in the hypochondrium, fortuitous discovery of a tumor mass on palpation of the hypochondrium, and more rarely discovery of distant metastases; can by itself be revealing of its tumors. Sometimes it is associated with a paraneoplastic syndrome: weight loss, asthenia, fever, pain, and deterioration in general condition (7).

Imaging

Imaging makes it possible to highlight the tumor mass with adrenal development, thus evaluating locoregional development (adipose tissue, mediastinal-pelvic abdominal lymph nodes, vena cava, renal vein, adjacent organs) or distant metastases (liver, lung, bone, peritoneum, most often) (8).

CT is the reference examination, the initial analysis of the tumor is based on the spontaneous density of the latter: any tumor with a density of less than 10 UH can be considered benign.

The adrenocortical tumor appears on CT as a heterogeneous mass, with irregular margins and a spontaneous density > 10 HU whose size exceeds 5 cm. It may present numerous calcifications, very often with necrotic or hemorrhagic areas. This tumor enhances heterogeneously after injection of the contrast product (8).

On MRI, the lesion appears hypointense on T1, and hyperintense on T2; sometimes there are very limited areas of intracytoplasmic fat. On the sequences in phase opposition, an increase in the signal is obtained. MRI can also supplement abdominal CT data to refine the assessment of locoregional, metastatic, vascular or ganglionic extension. the renal veins and the IVC to the right atrium and detects a

tumor extension, with an almost anatomical image, real pre-surgical cartography.

The PET-SCAN with 18 fluoro-deoxyglucose makes it possible to identify benign and malignant tumors as well as to complete the search for distant metastases which are sometimes silent(9).

Histological diagnosis:

The place of biopsy is limited in the diagnosis of adrenocortical tumors, due to the risk of tumor dissemination by capsular rupture, the only indication for a biopsy is the presence of distant metastasis to have histological proof(10).

4- Support

Treatment of localized stages: Stage I and II

>Surgical

Surgery is the reference treatment for adrenocortical tumors, the standard approach is laparotomy, in the case of Stage II it is wise to perform large surgery to reduce the rate of local recurrence, the resection must be in one piece without capsular rupture. Lymph node dissection would affect improving survival (11).

In the case of local recurrence, iterative surgery seems the best way to treat late recurrence > 12 months and initial R0 surgery (12).

>Adjuvant treatment

Is justified because of the strong nature of recurrence of adrenocortical tumors(11), it is based on adjuvant radiotherapy of the tumor bed, it is proposed for stages I – II R1, it must be started within 3 months following surgery for an At a dose of 50 to 60 Gy, treatment with mitotane seems justified despite the weak evidence in the literature (13).

The efficacy of mitotane has been demonstrated for stages I-II R0. The median recurrence-free survival was 42 months in the mitotane group versus 10 and 25 months in the 2 untreated control groups(14). The duration of adjuvant medical treatment is 2 years, with control of mitotane (11).

Treatment of locally advanced and metastatic stages: Stage III and IV

>Surgical

Surgery retains its place if complete resection extended to surrounding organs associated with adrenalectomy is possible.

> Medical treatment with mitotane:

May improve the quality of life of patients with secreting tumors, indicated as monotherapy in case of slowly progressive tumor (11).

> Chemotherapy

Indicated in the event of a rapidly evolving tumor, two first-line therapeutic strategies have been described: an association of etoposide, doxorubicin, cisplatin (EDP), and mitotane or streptozocin and mitotane, the results of which on overall survival are almost identical at 14.8 months for EDP-mitotane vs 12 months for streptozocine-mitotane, on the other hand, the response rate was better for EDP-mitotane (15).

5- Prognosis

The specific 5-year survival essentially depends on the tumor stage, the R0 resection, and the capsule rupture. For stage I it is described at 82%, Stage II 58%, stage III 55% and 18% for stage IV (16).

Conclusion

The diagnosis of adrenocortical tumors is difficult to establish and is based on several criteria, essentially clinical, biological, and morphological, including therapeutic management depending on the stage of the tumor, surgery is

the cornerstone of this management or resection must be extended to surrounding organs, without capsular rupture and

R0 to minimize the risk of recurrence and improve overall patient survival.

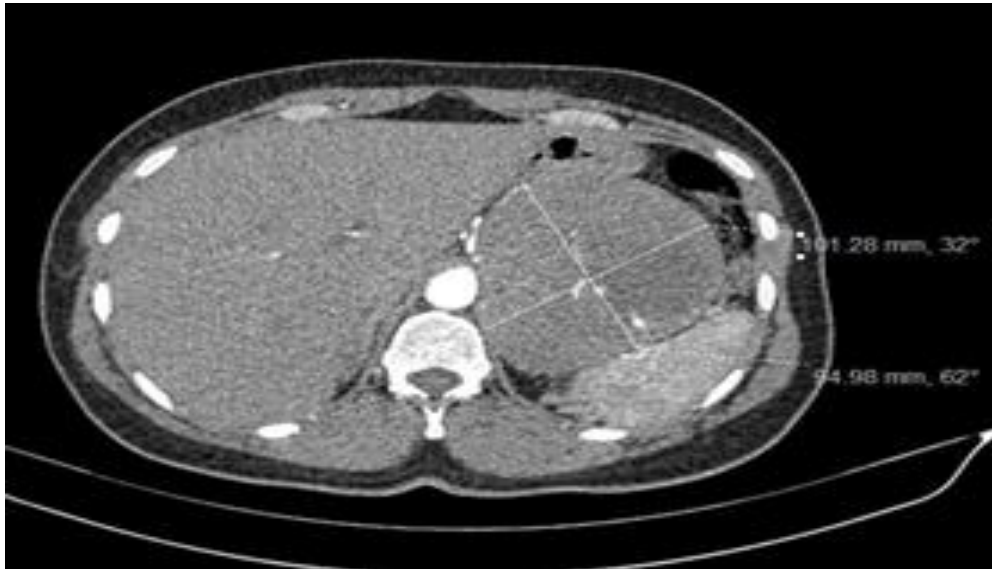


Figure 1. Adrenal mass with calcification and area of intra-tissue necrosis on cross-section.



Figure 2. Tissue mass depends on the left adrenal on sagittal section.

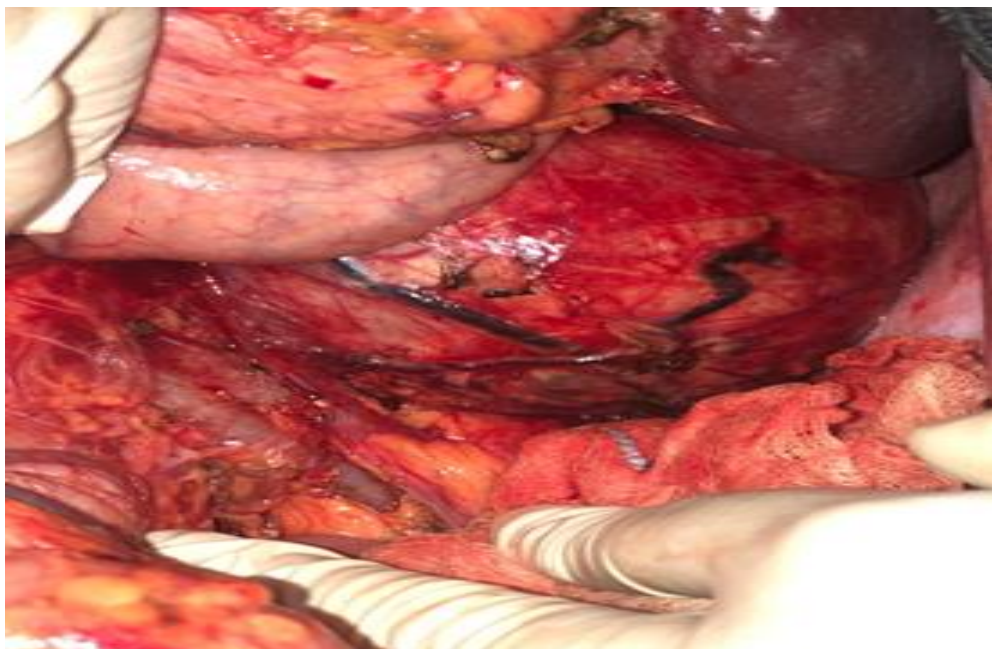


Figure 3. Intraoperative image of malignant adrenocortical tumor.



Figure 4. Synchronous liver metastasis from an adrenocortical tumor measuring 74.88 mm x 73.20 mm on cross section.

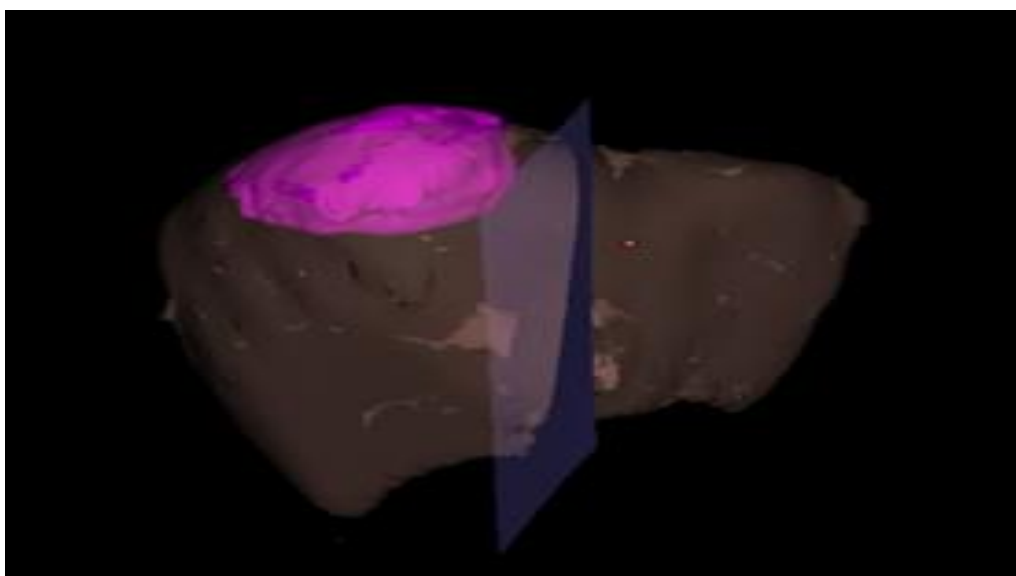


Figure 5. 3D model of hepatic metastasis from the adrenocortical tumor with a slice of section respecting healthy margins.

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