Huge Adrenocortical Carcinoma: Case Report and Literature Review

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Abstract

Adrenocortical carcinoma is a rare entity. We report a case of a 47-year-old male with abdominal pain. without classical tumor symptoms. Computed tomography revealed a left adrenal mass measuring 17 cm. Hormonal evaluation was negative. Open left adrenalectomy was performed by sub-costal approach. Diagnosis of adrenocortical carcinoma was established by pathology. Metastatic workup was negative. The patient has been followed regularly with no recurrence for one year. Adrenocortical carcinoma is a challenge. Surgery is the main treatment.

Keywords: Adrenocortical Carcinoma; Adrenalectomy; Mitotane

Introduction

Regarding the age of diagnostic patients, there is a bimodal distribution with a predilection for patients aged 5 and 20 years on the one hand and between 40 and 50 years on the other hand [1]. 60% of adrenocortical carcinoma were hormone-secreting [2]. Non-secreting masses were associated with a poorer prognosis because they were diagnosed at an evolved or metastatic stage, because of their insidious character [3].

In this study, we report a patient diagnosed as huge adrenocortical carcinoma treated by open adrenalectomy.

Case Presentation

Our patient is a 47-year-old male with isolated abdominal pain. Computed tomography showed a 17cm enhancing mass, originated from left adrenal gland, with 30 Hounsfield units density, well encapsulated, and a wash-out less than 60%, as showed in figure 1.

MRI revealed a tricompartmental mass arising from the left adrenal gland, with an area of hyperintensity in the centre and hyper-

Figure 1: Intravenous contrast CT scan shows a 17 cm retroperitoneal mass in the left suprarenal area, with heterogeneous enhancement, Regions of non-enhancing tissue (arrow) are consistent with necrosis.

intensity and the upper and lower pole suggesting haemorrhage and necrosis as showed in figure 2.

Figure 2: Coronal section of MRI showing the heterogeneous tricompartmental mass arising from the left adrenal gland.

Hormonal evaluation was negative, with normal ACTH and cortisol level. Suppression test was negative. Measurement of plasma renin activity and serum aldosterone levels showed no abnormalities. Plasmatic metanephrine and normetanephrine was also normal. Metastatic workup was negative.

A left subcostal adrenalectomy was performed. The mass was pushing the pancreas, spleen and left kidney with no evidence of organ invasion. No lymph nodes were found. After ligation of the adrenal vein, the mass was dissected and removed (Figure 3).

Figure 3: Surgical specimen showing a 17 cm well-encapsulated mass.

Histological examination showed 17x15x9 cm well-encapsulated, mass. The cross-section of the tumor (Figure 4) was tan-yellow and showed a tricompartmental aspect recalling the MRI findings.

Figure 4: The cross-section of the 17 cm adrenal mass recalls the tricompartmental aspect noted in the MRI findings with haemorrhage and necrosis areas.

Histological examination of the tumor (Figure 5) showed overt features of malignancy.

Figure 5: (A) Hematoxylin and eosin stain of the adrenal gland(x10) showing tumor cells (yellow arrow) and residual rim of normal adrenal gland (red arrow). (B) Hematoxylin and eosin stain of the tumor (x20), where the neoplastic cells show nuclear pleomorphism with multiple nuclei.

The diagnosis of adrenocortical carcinoma was confirmed. Post-operative recovery was uneventful. Our patient was discharged 4 days after surgery. The patient has been followed for one year with no recurrence.

Discussion

Adrenocortical carcinoma is a rare entity with a relatively bad prognosis [4]. A female predominance has been noticed [4]. Most patients with non-functional adrenocortical carcinoma present with advanced disease that is characterized by abdominal or extra-abdominal metastatic masses, because they are diagnosed incidentally [5]. Approximately 60% of cases present clinical symptoms of hormonal secretion. Hormone secretion may help to orient between benign and malignant adrenocortical masses [2].

Hormonal evaluation helps to define the functional character of the tumor and its origin. Imaging and especially computed tomography is sufficient to define the size of the mass and metastatic work-up [6]. Tumors more than 4 cm must raise suspicion of malignancy. Surgery is the mainstream of treatment for localized stages and as soon as the tumor is extirpable [7]. As in our cases, we performed open radical adrenalectomy.

Postoperative surveillance for recurrence is primordial to early detect recurrence. It should be performed regularly for the first 2 years (every 3 months) and then for 5 years every 6 months. Contrast tomography is the standard imaging for follow-up [8].

In case of metastatic disease or recurrence after surgical exercise, the only treatment that can be used is mitotane [9].

Apart from the standard radiological examinations used to detect recurrence after surgery, certain biomarkers such as microRNAs, found in adrenal tissue, where its presence is considered by some authors as a biomarker of malignancy and/or recurrence [10].

Conclusion

Adrenocortical carcinoma is a rare malignancy. Complete surgical excision is the mainstay of treatment especially in early stages. regular follow up plan should be established due to its high recurrence rate.

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