Oncology

Suspicious adrenal incidentaloma in a patient with Congenital Adrenal Hyperplasia: A case report

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ARTICLE INFO

Keywords:
- Congenital adrenal hyperplasia
- Adrenal incidentaloma
- Laparoscopic adrenalectomy
- Adenocarcinoma

ABSTRACT

Congenital Adrenal Hyperplasia has been associated with an increased prevalence of adrenal masses. It is still unknown whether incidentalomas in CAH* patients are more frequent or if the risk of adrenal carcinoma is higher than the general population. Therefore, the management CAH subjects presenting with suspicious adrenal masses remains problematic.

We relate the case of a patient with CAH presenting for an adrenal incidentaloma with malignant features. The management of such cases is controversial as surgery is risky for large masses. Despite dimensions, a laparoscopic approach was used for resection. The patient remained disease free at 4 years post-operatively.

Introduction

Congenital adrenal hyperplasia has been shown to be associated with an increased prevalence of adrenal masses. Despite the fact that most incidentalomas are benign, the unfavorable prognosis associated with the misdiagnosis of a malignant tumor warrants a careful evaluation with a multidisciplinary expert team. It is still unclear whether the hyperplasia of the adrenal cortex found in subjects with CAH* contributes to an increase in tumorigenesis and adrenal malignancy. We present the case of a patient known to have CAH that presented to our clinic with the discovery of an adrenal incidentaloma presenting suspicious radiologic features.

Case presentation

IH is a 27 years old phenotypic male (46,XX) that was referred to our urology clinic by his endocrinologist following the finding of a left adrenal incidentaloma. IH was diagnosed late in his childhood with a non-salt losing or simple virilizing form of CAH due to 21 hydroxylase deficiency. The discovery of a suspicious adrenal mass on an abdominal ultrasound warranted further investigation by tomodensitometry (Fig. 1). The abdominopelvic CT-scan showed a 9 × 8 × 7cm mass on the left adrenal gland with calcifications and central necrosis measuring 5.5cm. The right adrenal gland was of normal size. The patient was clinically asymptomatic and had no new signs of hormonal excess.

Laboratory analysis demonstrated a normal complete blood count, electrolytes, glucose, renal and liver function tests. Metabolic investigations showed normal levels of blood cortisol at noon, ACTH, renin, aldosterone and plasma metanephrines. 17-OH progesterone was elevated. Urinary metanephrines and VIMA were in the normal range. No dexamethasone suppression test was performed at this time. Blood pressure was normal.

After discussion with his endocrinologist, a laparoscopic approach for left adrenalectomy and mass resection was suggested. The patient was informed of the benefits and risks of such a procedure, particularly the risks of bleeding and conversion to open surgery.

The patient was admitted 48 hours prior to surgery for preoperative preparation with a diet rich in sodium, and an intra-venous administration of 50mg of hydrocortisone every 6 hours.

The laparoscopic transperitoneal adrenalectomy went smoothly without any significant bleeding or collateral organ injuries. Carcinologic principles regarding surgical margins were thoroughly respected. Unusual adherences and peritumoral fibrosis were not encountered (Fig. 2A and 2.B.). Conversion to an open technique was not needed.

The specimen, consisting in the well-rounded shape mass and the left adrenal gland along with the surrounding fatty tissue and fascia...
was retrieved en-bloc from the abdomen in an endobag through the enlarged left iliac fossa trocar orifice (Fig. 2 D.).

The postoperative period was uneventful, and the patient remained well.

The pathology report described a 8.5 × 8x7.5 cm tumor originating from the adrenal cortex with partial coagulative necrosis (Fig. 3A). The tumor was composed of cells arranged in solid and trabecular architectural patterns with few islands of clear cells (<25%) (Fig. 3B.). Mitotic activity was low with no atypical mitotic figures. The internal part of the capsule was infiltrated but no capsular rupture was noted.
The modified Weiss scoring criteria was determined and revealed a score of 2, suggesting the tumor to have uncertain malignant potential. Follow-up by thoraco-abdominopelvic CT-scan for local recurrence and distant metastasis was carried out every 6 months for a period of 3 years. IH was disease free at 4 years post-operatively.

Discussion

An adrenal incidentaloma is an asymptomatic mass discovered on routine abdominal imaging not performed for an adrenal purpose. Patients with CAH, in particular, have been shown to present adrenal hyperplasia due to the hypersecretion of ACTH with the possibility of tumor formation secondary to this proliferation. It is not still established whether patients with CAH are more at risk of adrenal malignancy than the general population.

Our patient presented with a large heterogenous adrenal incidentaloma. He did not have any signs of hypersecretion on either physical examination or laboratory tests. The appropriate diagnostic approach was to assess the risk of malignancy and to decide whether surgical excision would be appropriate. The most agreed upon malignant radiologic criteria are large necrotic areas and highly irregular borders. In our patient, the CT-scan displayed features suggesting malignancy due to large size and to the presence of focal necrosis and hemorrhage. When malignancy cannot be excluded, the surgical resection and excision of the mass is advocated since it markedly improves the patient’s outcome. Since the possibility of malignancy could not be eliminated, resection was decided. It can be performed surgically either via an open or laparoscopic approach. Current evidence concerning the preferred technique is weak, and is based only on the heterogenous results of several cohort studies. It is largely agreed upon that small tumors without signs of local invasion should be removed laparoscopically and large malignant tumors via an open approach. This consensus leaves a large grey zone for tumors that do not fit in either category, such as in our patient. Some studies have reported successful laparoscopic removal of giant adrenal tumors. We opted for a laparoscopic approach with an experienced surgeon because the mass, although large in size, seemed well confined on imaging. We explained to the patient the risks of conversion to an open approach if there is an increased risk of spillage or capsular disruption.

Definitive diagnosis was obtained on histopathologic study that showed an adrenal mass of cortical origin. The modified Weiss score allows, through the identification of several criteria, to assess the probability of malignancy. When the modified Weiss score equals 3 or more, the tumor is most certainly malignant. In our patient, the modified Weiss score was 2, and as such was deemed to be of “uncertain malignant potential”. The patient remained disease free at 4 years post-operatively.

Conclusion

The management of adrenal incidentalomas can be challenging and controversial especially in patients with CAH. Indications for surgical resection should be discussed on a case-by-case basis in a multidisciplinary team of experts according to a radiological assessment for malignancy.

Consent

Written and informed consent for the publication of the case and images were obtained from the patient.

Funding information

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of competing interest

Declarations of interest: none.

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