

Case Report

Adrenal Carcinoma with Cardiac Metastasis in a Child: Case Report

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ABSTRACT

Adrenal carcinoma is very rare in children, and cardiac metastasis from this tumor is even rarer. We present such a case and discuss its management. A ten year old boy presenting with hirsutism was referred to the radiology department of Mubarak Al-Kabeer Hospital for investigation. The investigations performed included US of the abdomen and CT examination of the chest and abdomen. In addition, the patient also had hormonal and

routine biochemical investigations. The Ultrasound and CT examinations revealed a solid mass arising from the right adrenal gland, infiltrating into the inferior vena cava and extending into the right atrium. Ultrasound guided fine needle aspiration of the mass, confirmed the diagnosis of adrenal carcinoma. The case is reported because of its rare nature.

KEYWORDS: adrenal carcinoma, cardiac metastasis, imaging

INTRODUCTION

Adrenocortical carcinoma is a rare malignancy in childhood^[1,2]. Extension of this tumor through the IVC into the right atrium is even rarer. The estimated incidence of this tumor is about 1 in 1,700,000^[1]. There are limited reports of cases with extension into the IVC and the right atrium. To our knowledge only ten such cases have been previously reported including one case with a tumor in the left adrenal gland^[3]. We report the case of a ten year old boy with a right adrenocortical carcinoma, extending into the IVC and the right atrium. To our knowledge this is the first case report from the middle east region.

CASE REPORT

A ten-year-old Omani boy presented in January 2001 with a three month history of weight gain and edema of both lower limbs in addition to features of precocious puberty (facial, axillary and pubic hair) for two years.

Physical examination revealed a short-statured and grossly overweight child with signs of precocious puberty. The genitalia appeared small and an adequate examination was not possible because of gross obesity. Examination of the cardio-vascular system at the time of admission revealed raised BP (165/95) and pansystolic murmur over the tricuspid area. On abdominal examination an ill defined mass was palpable in the right hypochondrium

extending to the right lumbar region.

Hormonal study revealed that testosterone (47 nmol/l), 17 OH progesterone (21 nmol/l), DHEAS (45 nmol/l) and androstendione (14 nmol/l) were all raised. The serum biochemistry, liver and renal functions were unremarkable.

Ultrasound examination of the abdomen showed a large solid mass arising from the right adrenal gland measuring 12 cm in diameter, infiltrating into the IVC (Fig 1a) and extending into the right atrium (Fig 1b). CT scan of the chest and abdomen was done for further workup. It confirmed the right adrenal mass (Fig 2a) extending into the IVC (Fig 2b) and the right atrium (Fig 2c). The lungs were clear and there was no evidence of bone metastasis of the scanned bones.

The intracardiac extension of the mass was also confirmed on echocardiogram.

Ultrasound guided fine needle aspiration biopsy confirmed adrenocortical carcinoma.

The child's parents refused any treatment and got discharged against medical advice.

The child was readmitted two weeks later with acute cardiac failure and died on the third day of admission.

DISCUSSION

Adrenal cortical carcinoma is a rare tumor in children comprising only 0.2 % of all childhood malignant tumors. It often presents with a large

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Fig 1a



Fig 1b

Fig 1: Ultrasound examination showing large tumor thrombus in the IVC (a) [arrows] extending into the right atrium (b)

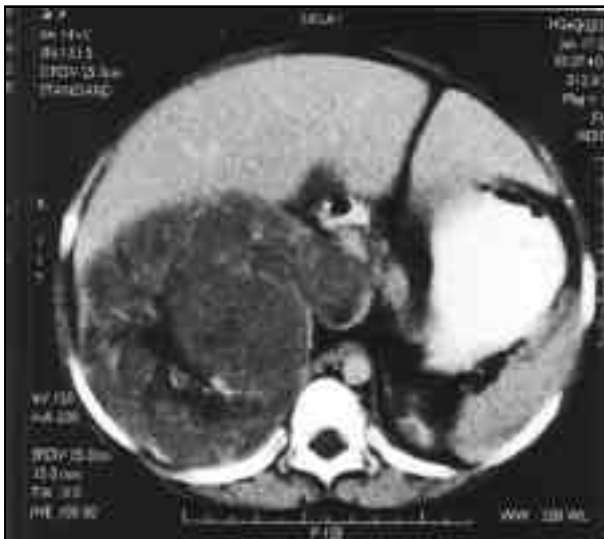


Fig 2a

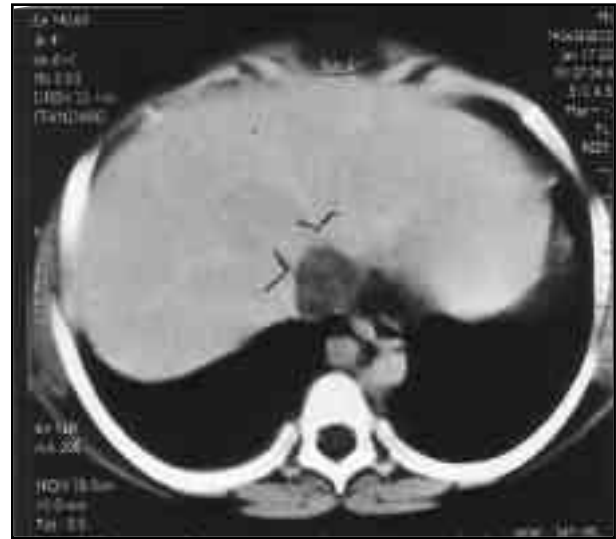


Fig 2b

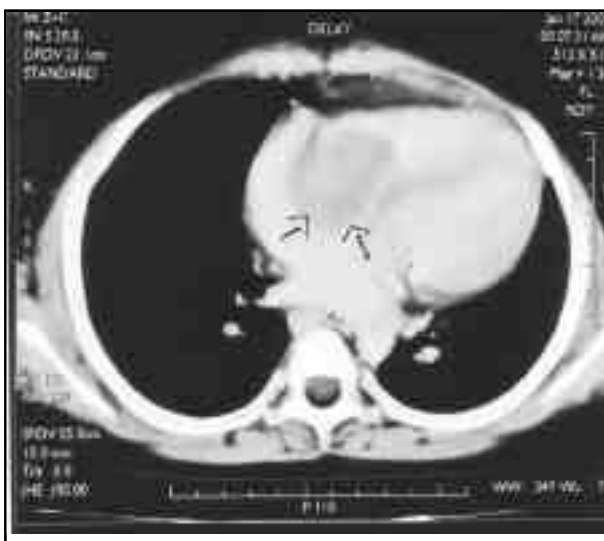


Fig 2c

Fig. 2: CT scan of upper abdomen showing the large adrenal mass infiltrating into the IVC (a & b [arrow heads]) and extending into the right atrium (c) [arrows]

abdominal mass and signs of precocious puberty^[1]. Despite its often dramatic presentation, there typically is long delay between the onset of symptoms and the time of diagnosis.

The clinical features of the tumor include those caused by the local mass effect, such as flank pain and abdominal fullness and of those caused by unregulated hormone production by the tumor^[4]. With cortical over production signs and symptoms of Cushing's syndrome are noted. Androgen secretion, which is less common, results in virilization, hirsutism and amenorrhea. The clinical features of right atrial involvement are due to central pulmonary embolization and cardiac and / or valve obstruction, often with associated congestive heart failure.

Ultrasound, CT and MRI studies are the main and effective imaging investigations in assessing the local as well as the IVC and intracardiac extension of the tumor^[5]. MRI is shown to be the

most sensitive method of diagnosing virilising adrenocortical tumors in children^[6]. The use of Ga-67 scintigraphy in detecting tumor recurrence and in assessing response to therapy has been reported^[1].

Due to the delay in diagnosis, adrenocortical carcinoma is a fatal disease with poor prognosis. Complete surgical resection is the only treatment that offers potentially long term disease free survival^[4]. Chemotherapy with Ortho Para DDD is recommended for treatment of residual or recurrent tumor after surgery or for patients with metastatic disease.

The mean survival without treatment is approximately three months and with treatment the five years survival is about 30%. The poor prognosis is mainly due to the long delay between the symptoms and the time of diagnosis.

In conclusion, it is imperative that the physician performs cardiovascular examination in suspected

or proven cases of adrenal carcinoma and recognize the endocrine manifestations of the tumor for prompt treatment.

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