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Case report

Late recurrent adrenocortical carcinoma presenting radiologically as a gastrointestinal stromal tumour: A case report



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Abstract

Introduction: Adrenocortical carcinoma (ACC) is a rare malignancy with an estimated incidence of 1–2 per million people. It may recur, after complete surgical removal by local or distant metastasis.

Observation: We report a case of late metastatic ACC presented as a mesenteric mass, 10 years post left adrenalectomy. Our case was initially misdiagnosed radiologically as gastrointestinal stromal tumour (GIST), and then the decision for exploration was made. The mass could be safely excised and confirmed pathologically to be an adrenocortical tumour.

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Introduction

Adrenal tumours are very common, affecting 3–10% of the human population, and the majority are small benign nonfunctional adrenocortical adenoma [1]. Adrenocortical carcinoma (ACC) is a rare

malignancy with an estimated incidence of 1–2 per million people [2]. There are 3 main clinical scenarios in which ACC patients present. For 40–60% of patients, the major presenting complaints are symptoms and signs of hormonal excess [3–5]. Another one-third present with non-specific symptoms due to local tumour growth, such as abdominal or flank pain, abdominal fullness, or early satiety [4,5]. Roughly, 20–30% of ACCs are incidentally diagnosed by imaging procedures for unrelated medical issues [6]. Patients with ACC only rarely present with classical tumour symptoms, such as cachexia or night sweats while paraneoplastic syndromes are uncommon [3,5].

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In patients with localized ACC, operative resection remains the mainstay of therapy. Patients with early-stage tumours who undergo a complete resection have a 40% 5-year survival rate whereas those with residual disease fare poorly [7]. Despite apparent complete microscopic operative resection, ACC recurs either locally or with distant metastasis in up to 50% of patients [7].

Gastrointestinal stromal tumours (GIST) present clinically with vague abdominal pain, abdominal fullness or early satiety, resembling one-third of cases suffering from ACC [8]. Radiologically, GIST appear in CT as masses with soft tissue density, variable in size and heterogeneity following contrast injection. In most of the cases, it is located within the mesentery or directly related to the bowel [8]. GIST resembles ACC in radiological appearance, except for the location. Most of the tumours that were reported to be wrongly diagnosed preoperatively as GIST were Schwannoma or Desmoid tumours [9,10].

We describe this very rare clinical presentation of metastatic ACC to the mesentery following previous two surgeries of left adrenal gland for ACC followed by local tumour recurrence, radiologically mimicking GIST.

Case report

A 24-year-old female patient presented to our urology clinic with vague upper abdominal discomfort. Her surgical history revealed that in 2005 she underwent left adrenalectomy and the histopathological examination described a mass 12 cm × 8 cm × 5 cm of 130 g in weight with microscopic features of tumour cells showing mild nuclear atypia and rare mitotic figures, minimal necrosis and no vascular invasion. The final pathologic diagnosis was left adrenocortical neoplasm of indeterminate malignant potential.

Five years, post adrenalectomy, the patient complained of recurrent attacks of left hypochondrial dull aching pain; when multiphasic CT was done, it revealed a soft tissue mass lesion in the anatomical site of left adrenal gland measuring 6 cm × 7.2 cm × 6.5 cm in dimension (Fig. 1). She underwent surgical exploration and excision of the mass. Left nephrectomy was done due to accidental injury of left renal vein during tumour resection. Histopathological examination

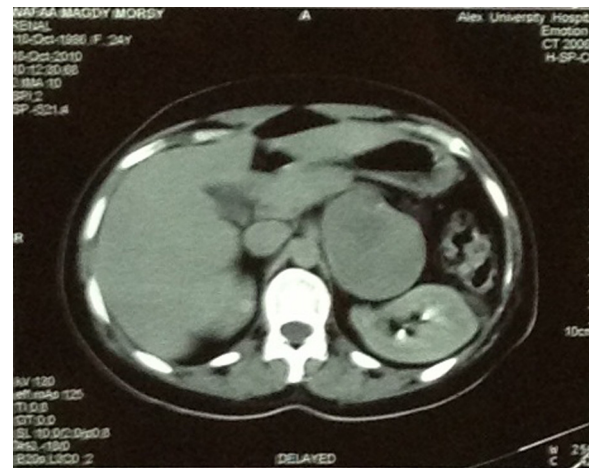


Figure 1 CT image showing a recurrent large left suprarenal tumour.

revealed ACC with tumour-free surgical margin and the left kidney was unremarkable.

Currently, she presented to our department with the same complaint of generalized abdominal pain. Based on her history, we requested new multiphasic CT scan of abdomen and pelvis together with hormonal workup.

Multiphasic CT scan revealed a 6 cm ovoid mass in the mesentery (Fig. 2) to the left of inferior mesenteric vein with small indentation of related jejunal loop, as well as mesenteric supply and portal drainage suggestive of gastro-intestinal stromal tumour (GIST). Hormonal workup revealed normal ACTH level (<5 pg/ml), elevated serum cortisol level at 9 pm (20.3 µg/dl) while normal level at 9 am (19.7 µg/dl) and elevated urinary cortisol level (715 µg/24 h urine of 1700 ml). All other hormonal workup was normal.

Ultrasound-guided core biopsy was obtained from the described mass and submitted for histopathological examination which described a metastatic oncocyctic carcinoma. Further immune-histochemical staining to exclude carcinoid tumour

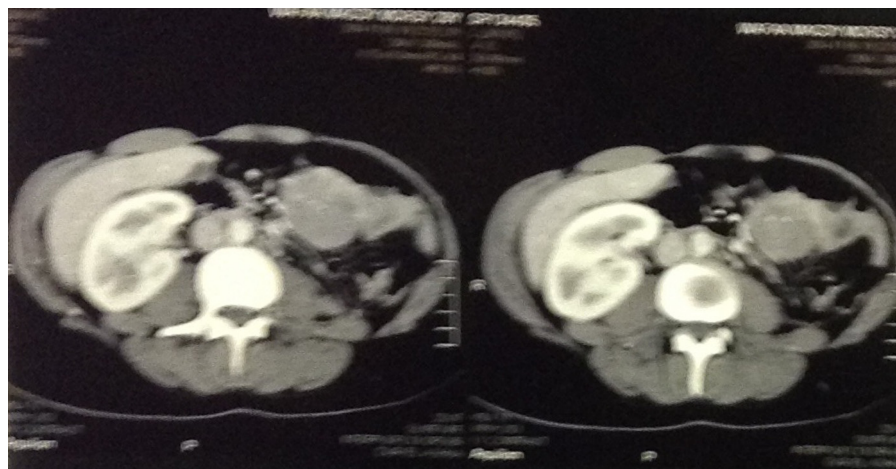


Figure 2 CT image showing a recurrent tumour within the mesentery.

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