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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.

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

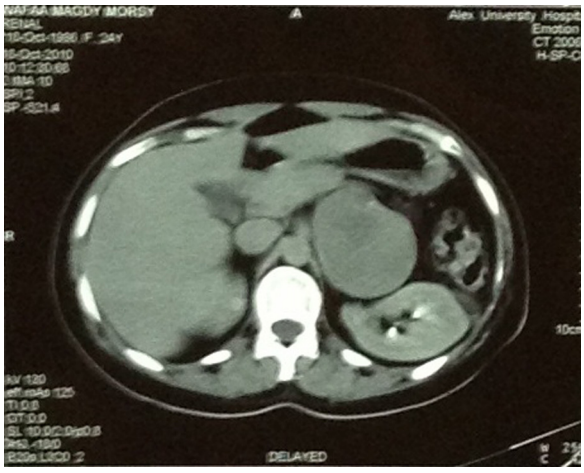

Figure 1 CT image showing a recurrent large left suprarenal tumour.

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

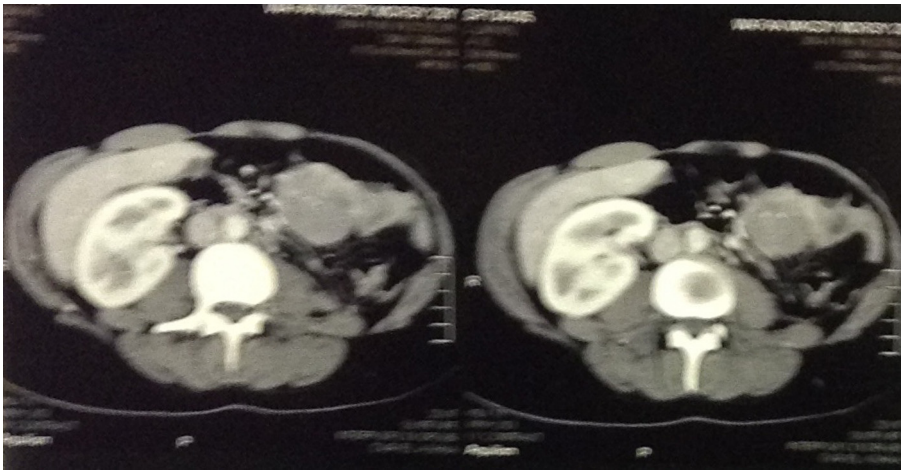

Figure 2 CT image showing a recurrent tumour within the mesentery.

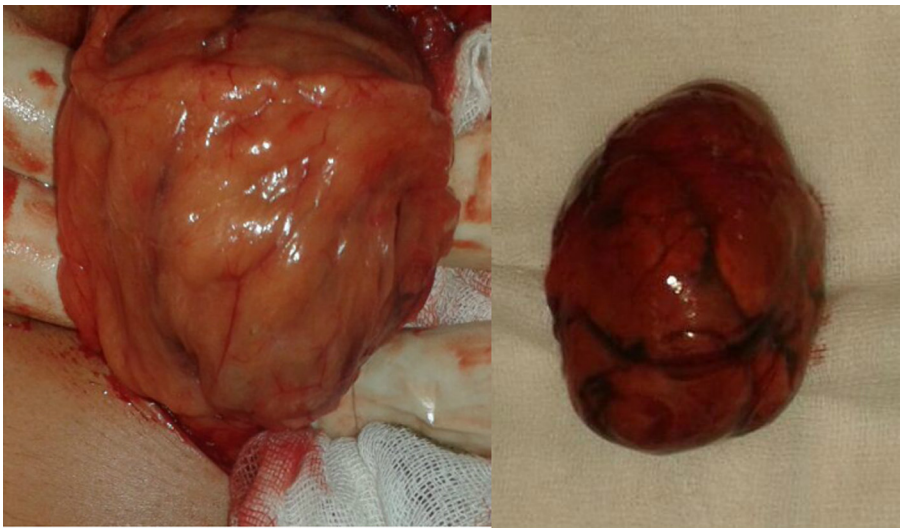

Figure 3 The insert shows the resected recurrent mass clearly identified intraoperatively, within the mesentery.

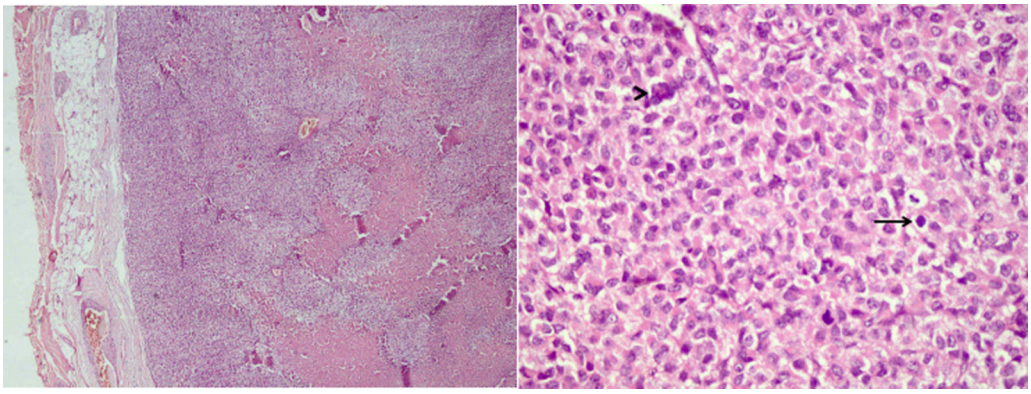

Figure 4 On scanning view, the tumour architecture was predominantly solid. Necrosis occupied a significant area of the tumour and had a geographical pattern (H&E, 40×). On high power examination, the vast majority of the cells are polygonal with abundant eosinophilic cytoplasm, a moderate degree of nuclear pleomorphism, prominent nucleoli, and vesicular chromatin pattern. Atypical mitotic figures were also present (arrow) as well as occasional bizarre nuclei (arrow head). (Left) Low power image and (right) high power image.

revealed negative for chromogranin with Ki67 labels about 10% of the nuclei.

Surgical exploration was done through a small midline laparotomy and the mass was easily identified within the mesentery (Fig. 3). Total excision was done. Post-operative course was smooth and the intraperitoneal drain was removed after 1 day.

Histopathological examination of the excised mass described a 6.5 cm × 6 cm × 3.5 cm grossly encapsulated mass with microscopic features of a tumour composed of solid sheets of eosinophilic cells with vesicular moderately pleomorphic nuclei and wide areas of necrosis. Final diagnosis was metastatic ACC (Fig. 4).

Discussion

We report a rare case of late metastatic ACC to the mesentery about 10 years following left adrenalectomy. Although the radiological features of the mass together with the biopsy results were strongly suggestive of GIST, the final pathology of the excised mass was metastatic ACC. We used Ki67 staining that was positive for 10% of nuclei. It triggered us to proceed for exploration for its high association with malignancy and shorter disease specific survival [11].

Our case presented with a tumour recurrence in an abnormal location, 10 years following initial radical surgery. Apart from abdominal discomfort, she had a great performance status with no major symptoms or signs of advanced malignancy. In a retrospective analysis from the German ACC registry, out of 154 patients presented with recurrence following initial radical resection, 101 underwent re-surgery. The best predictors of prolonged survival after first recurrence were time to first recurrence over 12 months and radical resection [12].

Local recurrence and/or metastases are common after initial radical resection for adrenal tumour, reaching up to 50% of cases [13,7]. These cases have a low 5-year survival rate of 0–6% [13,7]. Hermsen et al. [14] described 6 cases of late recurrence with excellent long survival. Our case presented with local recurrence 5 years following

initial radical resection, then 5 years later with a single metastasis within the mesentery, associated with great performance status. Our case is currently followed for 6 months following the third surgery, with no clinical and radiological evidence of recurrence.

Conclusion

Adrenocortical carcinoma may present with late metastasis in an abnormal location including the mesentery. Any mass suspected to be gastrointestinal stromal tumour, in the presence of past history of ACC should be investigated thoroughly by CT, core biopsy and immunohistochemical staining.

Authors' contribution

Ahmad Beltagy: data collection, literature review and writing the manuscript.

Ahmed Kotb: literature review, writing the manuscript and surgical management for the case.

Mohamed Shaaban: radiological diagnosis and analysis for the case.

Mona Abd-Elhadi and **Mervat Hamza**: pathological study of the tumour.

Ahmed Elabbady, **Mohamed Adel Atta**, **Mohamed Abdelrahman**: analysis of the surgical case and mentoring and revision of the manuscript.

Ethical committee and patient consent

Ethical committee approval and patient consent were obtained.

Conflict of interest

None declared.

Source of funding

None.

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