Young male with left adrenal mass

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Accepted 3 July 2016

DESCRIPTION

A 20-year-old male patient presented with a 5-month history of left hypochondriac pain and mass. He denied any associated constitutional symptoms. Clinically, he was overweight with no cushingoid or virilising features. Abdominal examination revealed a huge non-tender mass arising from the left hypochondrium. There was no hepatomegaly and the testicles were normal.

Routine blood investigations including tumour markers and plasma and urine hormonal assays were normal. CT scan (figure 1A) and MRI (figure 1B) revealed a large left retroperitoneal mass (15×12 cm) with small bilateral lung deposits. After excluding pheochromocytoma, US-guided

transcutaneous biopsy raised the possibility of non-functioning adrenocortical carcinoma (ACC). Laparotomy revealed locally advanced adrenal tumour (figure 2A) with segments V and VII hepatic deposits. Hence, a palliative resection was carried out (figures 2B) and the histopathology confirmed ACC (weight: 2.11 kg; size: 24×10×9 cm) with negative lymph nodes (figure 3). Immunohistochemistry expressed focal positivity for vimentin, inhibin and cytokeratin. He was later lost to follow-up and therefore presumed dead after 3 months.

ACC is a rare tumour which has poor prognosis and the 5-year survival rate is barely 20%.¹ Distant metastases at presentation occur in 50% of cases.¹

Figure 1 CT scan (panel 1A) and MRI (panel 1B) of the left adrenal mass showing the inhomogeneous appearance of the lesion with necrosis in both imaging modalities. Differential diagnoses include benign solid tumours (eg, oncocytoma of the kidney or adrenocortical adenoma) and malignant lesions (eg, hypernephroma or adrenocortical tumour). The low fat content differentiates this lesion from benign adrenocortical adenoma.

Figure 2 Photograph of the adrenal mass at laparotomy before surgical resection (panel A). Panel B showing the operative site after the resection of the mass with the preserved left renal vein and inferior vena cava.

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To cite: Meshikhes A-WN,
Abdel Gawad WM, Al-
Saeed JY. *BMJ Case Rep*
Published online: [please
include Day Month Year]
doi:10.1136/bcr-2016-
215669

Figure 3 Low-power magnification microphotograph of the tumour (panel A) featuring sheets and nests of malignant epithelial cell with eosinophilic cytoplasm and pleomorphic nuclei. Areas of tumour necrosis are also seen (H&E $\times 40$). Panel B is featuring sheets of tumour cells with prominent nuclear pleomorphism and several mitoses are present (H&E $\times 400$).

Preoperative transcutaneous biopsy—though performed in this case—may not be necessary and has a complication rate of 11%, but 70% diagnostic sensitivity.² Although the resectability rate is 50%,³ treatment is by aggressive surgical resection with or without lymph node dissection even in presence of metastases.^{1 3}

ACC exhibits poor response to chemotherapy, but mitotane remains the only drug that produces tumour and metastatic regression in incompletely-resected or non-resectable carcinomas.

Learning points

- ▶ Adrenocortical carcinoma is a rare tumour which is often large in size, locally advanced and metastatic at presentation.
- ▶ It is associated with poor prognosis; the 5-year survival rate barely exceeds 20%.
- ▶ Although the resectability rate is <50%, treatment is aggressive surgical resection with or without lymph node dissection even in presence of metastases.

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Acknowledgements The authors would like to acknowledge Dr Samir Amr, chairman of the pathology department, King Fahad Specialist Hospital, Dammam and Dr Fouad Al-Dayel, Consultant pathologist, King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia for providing the pathology microphotographs with the legends.

Contributors A-WNM was the primary physician who managed the case. He wrote the final draft. WMAG participated in the management of the case, searched the literature and wrote the initial draft. JYAI-S shared the medical treatment of the case and helped in the initial draft.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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