Annals of Surgical Case Reports

Case Report
Published: 29 Apr, 2021

Long-Term Survival after *En-Bloc* Excision of Adrenocortical Cancer and Inferior Vena Cava

Radu Mihai[1], Zahir Soonawalla[1], Andrew Weaver[2] and Mario Petrou[3]*

[1]*Department of Surgery, Oxford University Hospital NHS Foundation Trust, Oxford, UK*

[2]*Department of Oncology, Oxford University Hospital NHS Foundation Trust, Oxford, UK*

[3]*Department of Cardiothoracic Surgery, Oxford University Hospital NHS Foundation Trust, Oxford, UK*

Abstract

Adrenocortical Carcinoma (ACC) is a very rare tumour with an estimated incidence of 1-2/ million population/year and an overall 5-years survival of 40%. Surgical resection remains the only treatment that can significantly improve survival of such patients. This case report illustrates the need for multidisciplinary collaboration in the care of patients with locally advanced ACC.

Keywords: Adrenocortical carcinoma; Adrenal surgery; Tumour; CT scan

Introduction

Adrenocortical Carcinoma (ACC) is a very rare tumour with an estimated incidence of 1-2/ million population/year and an overall 5-years survival of 40% [1]. Surgical resection remains the only treatment that can significantly improve survival of such patients. Due to the rarity of the disease, many patients with ACC are operated by surgeons with low volume practice in adrenal surgery [2]. Recently the European Society of Endocrine Surgeons summarized the published evidence for the expected volume-outcome correlation and recommended that surgery for ACC should be restricted to centers performing at least 12 adrenal operations per year [3]. A mechanism to reorganize surgical practice in different countries towards achieving this aim is yet to be established. In this context, the current service delivery for adrenal surgery in UK is in urgent need for centralization as the majority of adrenalectomies are done by surgeons doing a median of 1 case per year [4] and operations for ACC are done in centers where there was only one or none such case operated in the previous 12 months.

This case report illustrates the need for multidisciplinary collaboration in the care of patients with locally advanced ACC [5,6].

Case Presentation

A 28-years old man presented to his local hospital with sudden onset of severe right upper quadrant pain following few weeks of nonspecific abdominal discomfort, loss of appetite, regular vomiting and two stones eight loss. An abdominal ultrasound described a 157 mm × 112 mm mass at the upper pole of kidney and CT with contrast described a right-sided heterogeneous adrenal mass. At the time of his referral in December 2011 he denied any symptoms suggestive of pheochromocytoma or Cushing syndrome.

Investigations

Biochemically the tumour was non-functional [urine noradrenaline 564 nmol/24 h (N 120-590), adrenaline <60 nmol/24 h (N 0-190), dopamine 2717 nmol/24 h (N 650-3270), normal overnight dexamethasone suppression test].

CT scan (Dec 2011) described an adrenal mass, difficult to separate from the right crus of the diaphragm and segment VII of the liver. The right renal vein appeared to be likely thrombosed; the IVC was not well opacified (Figure 1A).

Treatment

In January 2012 right open adrenalectomy was attempted. Colonic hepatic flexure was densely adherent to tumor and could not be mobilized. The third part of duodenum appeared directly involved by tumour which also invaded the posterior wall of IVC over 6 cm to 10 cm. After involvement of a liver surgeon, a vascular surgeon and a cardiac surgeon it was deemed safer to

OPEN ACCESS

Correspondence:
Radu Mihai, Department of Endocrine Surgery, Churchill Cancer Centre, Oxford University Hospitals NHS Foundation Trust, Oxford, OX3 7LE, UK, Tel: 0044 1865 223555; E-mail: Radu.Mihai@ouh.nhs.uk

Received Date: *25 Mar 2020*
Accepted Date: *23 Apr 2021*
Published Date: *29 Apr 2021*

Citation:
Mihai R, Soonawalla Z, Weaver A, Petrou M. Long-Term Survival after En-Bloc Excision of Adrenocortical Cancer and Inferior Vena Cava. Ann Surg Case Rep. 2021; 4(2): 1046.

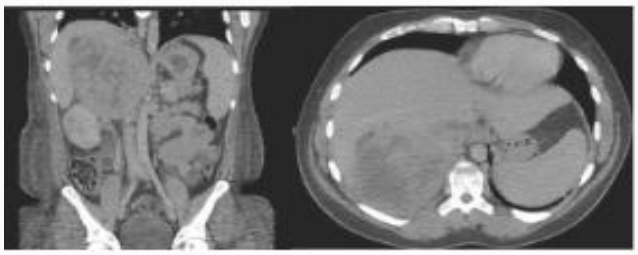

Figure 1A: Cross sectional imaging demonstrating change in appearance of large right-sided adrenocortical cancer during chemotherapy with mitotane, CT appearance at the time of initial presentation.

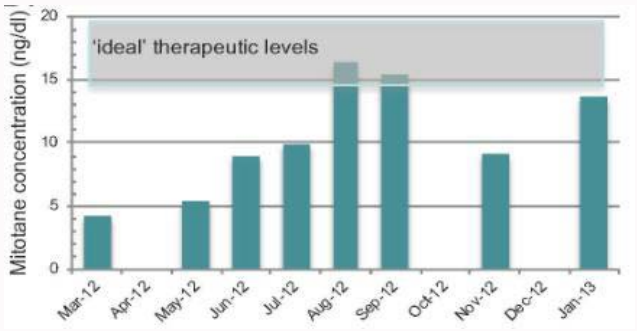

Figure 1B: Cross sectional imaging demonstrating change in appearance of large right-sided adrenocortical cancer during chemotherapy with mitotane. Mitotane blood levels during 'neoadjuvant' chemotherapy.

abandon the procedure.

A CT angiogram (Feb 2012) showed multiple arterial sources and therefore embolization was deemed to be unlikely to be effective. Mitotane chemotherapy was started in May 2012, starting with 3 gm/day and increasing up to 6 gm/day, but reaching a blood concentration of over 14 ng/dl only on one occasion over 6 months period (Figure 1B). A repeat CT scan in 2012 showed that the tumour had reduced in maximum dimensions (Figure 1C).

In April 2013 redo surgery was performed by a surgical team consisting of endocrine-, liver- and cardiac surgeon. The adrenal was mobilized en-block with the right kidney. The Infrarenal IVC and left renal veins were dissected before cardiopulmonary bypass was setup by performing sternotomy and cannulation in the proximal aorta, right atrium and IVC distal to right renal vein. A segment of IVC was resected in continuity with tumor and IVC reconstructed with a composite graft (homograft+bovine pericardium). Left renal vein was reimplanted in the reconstructed IVC.

The histological report described a large specimen (170 mm × 110 mm × 75 mm) containing a 80 mm, ACC with extensive vascular invasion, excised with a minimum margin of 1 mm, portal lymph node negative, proliferation index MIB1 up to 5% (T2N0M0, ENSAT stage III).

Outcome and follow-up

Postoperatively patient continued Mitotane chemotherapy for a further 2 years. As he had a nonfunctional ACC it remained impossible to use biochemical assessment to monitor disease recurrence. Hypoadrenalism during Mitotane chemotherapy was managed with high doses of hydrocortisone supplements (40 mg/day). This was stopped some 3 years after completing chemotherapy and at the present time the patient requires no steroid replacement.

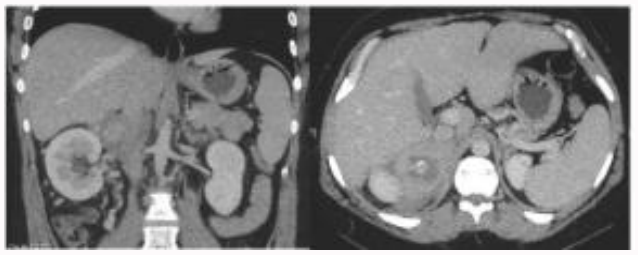

Figure 1C: Cross sectional imaging demonstrating change in appearance of large right-sided adrenocortical cancer during chemotherapy with mitotane, CT appearance after completion of 9 months of mitotane chemotherapy before the second attempt to resect the ACC.

Functional imaging with F18-FDG-PET was done on three occasions during his follow-up and showed only physiological uptake with no suspicion of recurrence in the adrenal bed or distant disease. His most recent cross-sectional imaging was normal (March 2020). Currently he is well and will continue radiological follow-up every 2 years.

Discussion

This case report illustrates the favorable outcome obtained in the treatment of a young patient who had a multivisceral radical resection under cardiopulmonary bypass after presenting with locally-advanced non-metastatic non-functional Adrenocortical Carcinoma (ACC).

When encountering a patient with locally-advanced ACC it remains very difficult to decide on the balance between the morbidity of the (possible) surgical treatment and its oncological benefits. In the absence of previous experience in treating similar patients it remains likely that clinicians might label patients as 'not suitable for surgical intervention' when their disease require an operation that is outside the surgeon's comfort zone and experience.

The common association of extensive local invasion or vascular involvement is likely to lead to a diagnosis of unresectable tumors and therefore some of these patients might not be offered surgical treatment. In a multicentre European study of 38 patients with ACC with IVC invasion complete resection was achieved in the majority of patients (20/38) with a perioperative 30-day mortality of 13%. The 25 patients had survived a median 5 months (range 2 to 51 months) and 13 patients were alive at median 16 months after the operation. The relatively low perioperative mortality and the long disease-free survival achieved by some patients should encourage surgeons with adequate experience to offer surgical treatment to patients presenting with advanced disease [7].

The support that cardiac surgeons can provide during treatment of ACC with IVC invasion has recently been summarized by our team [8]. This case provides clinical details of one such case, bringing evidence in favor of centralization of ACC care in centers where multidisciplinary surgical and oncological expertise can be provided.

The case also confirms the benefits of using neoadjuvant mitotane in patients with apparently unresectable tumors. This idea follows the concept of 'borderline resectable tumors' proposed by the MD Anderson clinicians when referring to patients who may have tumour or patient characteristics at presentation that argue against immediate surgery because of an unacceptable risk of morbidity/mortality, incomplete resection, or recurrence. Thirteen such patients treated with neoadjuvant mitotane therapy had satisfactory postoperative

outcomes, suggesting that response to Mitotane selects a subgroup of ACC patients with advanced disease that still benefit from radical surgery [6].

References

1. Tierney JF, Chivukula SV, Poirier J, Pappas SG, Schadde E, Hertl M, et al. National treatment practice for adrenocortical carcinoma: Have they changed and have we made any progress? J Clin Endocrinol Metab. 2019;104(12):5948-56.
2. Park HS, Roman SA, Sosa JA. Outcomes from 3144 adrenalectomies in the United States: Which matters more, surgeon volume or specialty? Arch Surg. 2009;144(11):1060-7.
3. Mihai R, Donatini G, Vidal O, Brunaud L. Volume-outcome correlation in adrenal surgery-an ESES consensus statement. Langenbecks Arch Surg. 2019;404(7):795-806.
4. Palazzo F, Dickinson A, Phillips B, Sahdev A, Bliss R, Rasheed A, et al. Adrenal surgery in England: better outcomes in high-volume practices. Clin Endocrinol. 2016;85(1):17-20.
5. British Association of Endocrine and Thyroid Surgeons (BAETS). Fifth National Audit Report. 2017.
6. Bednarski BK, Habra MA, Phan A, Milton DR, Wood C, Vauthey N, et al. Borderline resectable adrenal cortical carcinoma: A potential role for preoperative chemotherapy. World J Surg. 2014;38(6):1318-27.
7. Mihai R, Iacobone M, Makay O, Moreno P, Frilling A, Kraimps JL, et al. Outcome of operation in patients with adrenocortical cancer invading the inferior vena cava--a European Society of Endocrine Surgeons (ESES) survey. Langenbecks Arch Surg. 2012;397(2):225-31.
8. Greco R, Tsappa I, Mihai R, Petrou M. Surgical management of adrenal tumors extending into the right atrium. Gland Surg. 2019;8(Suppl 1):S53-9.