Adrenocortical Carcinoma
Where's the delay?

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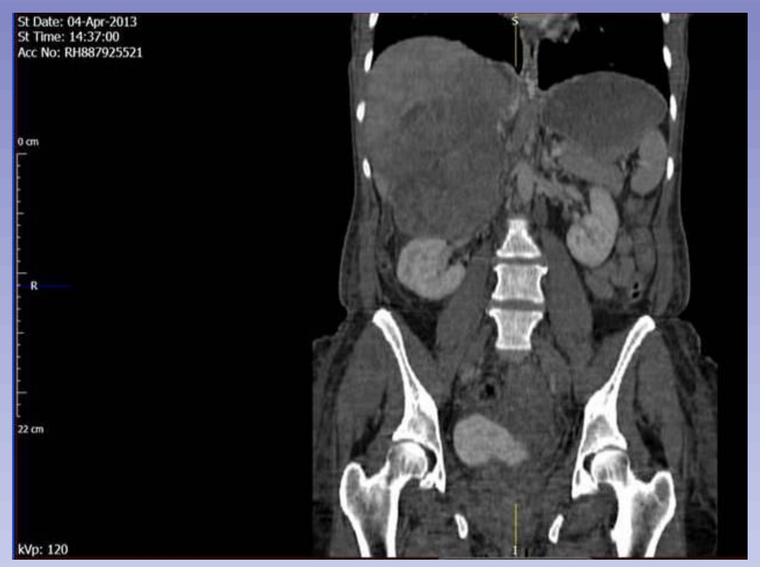
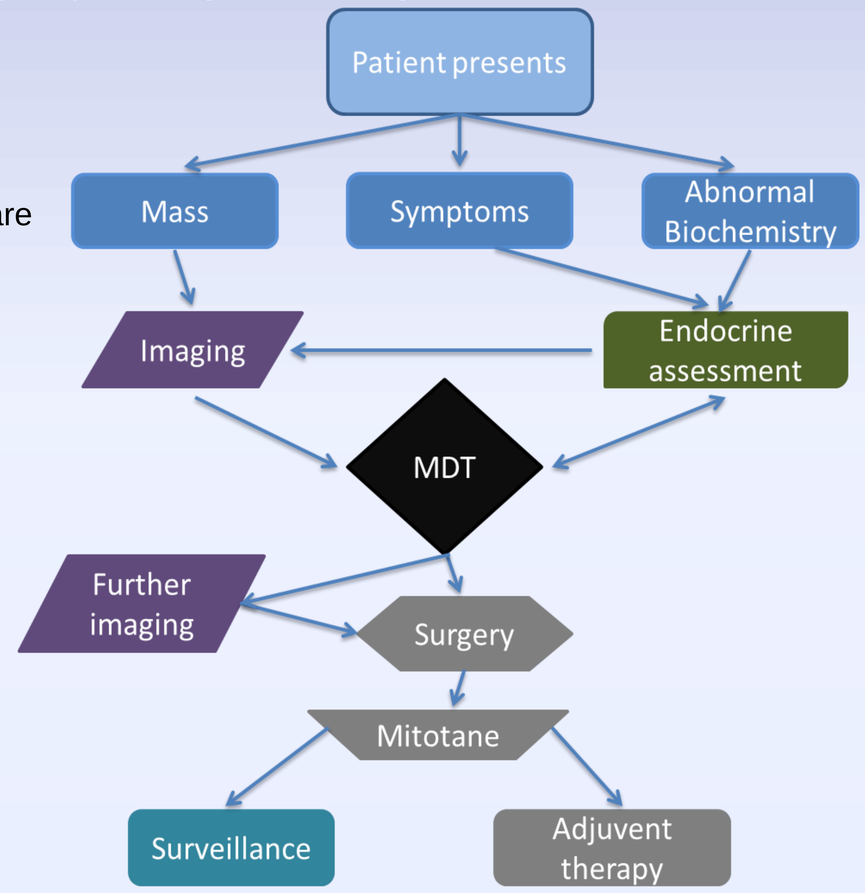


Introduction

• Adrenocortical carcinomas are rare (incidence 1-2 per million), present late and have a poor prognosis (1).
• Appropriate management of adrenal carcinomas involves collaboration within a multidisciplinary team (MDT) involving endocrinologist, surgeon, oncologist, pathologists, nurse specialists and radiologists (2)
• Rarity of the condition often necessitates multi-centre collaboration

Objective

•Five cases of adrenal carcinoma are described to demonstrate how multi centre care has opportunities to avoid delay (at primary, secondary and tertiary levels) and to optimise the pathway of care



Findings

5 patients (age 38 – 76yrs) presented between September 2011 and April 2013

Presentation:
• 3 patients presented with mass symptoms, 2 with secretory symptoms
• All patients had symptoms / primary care attendances for 3-6 Months prior to a diagnosis being made (one patient making her own diagnosis - this was the longest delay)
• All tumours presented at stage III-IV on imaging

MDT:
•Time to 1st MDT was <1 week to 2 months
(2 pts operated on elsewhere only had documented MDT discussion post operatively – discussion may have lead to earlier Mitotane and closer follow up)
•2 patients were discussed at cancer centres:
time to decision (not operable) at MDT was 10 days in one patient (an anguished wait as an inpatient!)

Surgery:
•Time to surgery from first presentation was 1 to 7 months. 1 case had a 3 month delay to enable combined approach and to lower cortisol (which was only modestly elevated).
•The surgery was performed by 4 different surgical centres (1 referred to another centre due to invasion of IVC (requiring combined approach), 2 presented post operatively from other centres, 1 operated on by adrenal surgeon)

Mitotane:
•All patients received Mitotane, however it was poorly tolerated at therapeutic levels (and some delay in initiating it).
•2 patient achieved therapeutic Mitotane levels
•4 patients offered chemotherapy (according to FIRM-ACT) : 2 declined (1 age 76yrs, 1 age 39yrs); 2 were too unwell to proceed

Outcome:
•All 4 patients referred to palliative care team have deceased (time to death: 1-17M)

Age	First Presentation	Biochemistry	Imaging	MDT decision	Surgery	Mitotane	Post-op Hydrocort	Other Rx	Outcome
64♀	Jan 2013 Secondary care (incidental)	Secretory A4 28.3nmol/l (0.8-11.9), ODST 9am Cortisol 147nmol/l	8.5cm R adrenal tumor (Stage 3)	22/05/13 DELAY	05/06/13 (Open) Adrenal surgeon	Y Therapeutic	Y	None	In remission + Mitotane Rx
38♀	Nov 2012 Secondary care (mass symptoms)	Presumed Secretory Cortisol secreting post op, 30min Cortisol 265nmol/l, high levels later DELAY	20cm L adrenal tumor, cardiac & lung metastasis (Stage 4)	15/01/13 (postoperatively)	13/12/12 (Open) Urologist	Y DELAY	Y	Palliative Cardiac RTX	RIP 04/02/13
39♂	Feb 2012 Secondary care (secretory symptoms)	Secretory 24 hr Ur Cortisol 2308, 3495, 3868 nmol/l (10-147)	8cm R adrenal tumor, IVC & lung metastasis (Stage 4)	17/02/12	24/05/12 (Open) Cancer centre (joint approach) DELAY	Y Therapeutic	Y	Ketoconazole Metyrapone	RIP 07/05/13
76♀	Sep 2011 Private (secretory symptoms) DELAY	Secretory 24 hr Ur Cort 426, 173, 299 nmol/l (10-147)	13cm L adrenal tumor & lung metastasis (Stage 4)	N/A	19/04/12 (Open) Urologist DELAY	Y	Y	Ketoconazole Metyrapone	RIP 18/02/13
54♀	April 2013 Secondary care (mass symptoms) DELAY	Secretory 24 hr Ur Cortisol 229, 172, 220umol/l (10-147), A4 63.2nmol/l(0.8-11.9), DHEAS 27.1umol/l (0.5-5.56)	11cm R adrenal tumor, cardiac,lung & liver metastasis (Stage 4)	11/04/13 DELAY	Inoperable	Y	Y	Metyrapone	RIP 26/04/13

Observations and comments

•Patients present late with adrenal cancer. The longest delay was making the diagnosis due to the rarity and complexity of symptoms not being recognised. Accessibility of endocrinology to give immediate GP advice and ongoing education may minimise this.
•Other delays included biochemical work-up, discussion through MDT, surgical treatment & post-op mitotane initiation.
•All patients had care involving more than one centre, from specialist centres to district general hospitals and delays in networking were relevant. Patients offered choice of care across centres all chose local care for follow up.
•Logistics of rapid diagnosis, investigation and treatment across primary, secondary and tertiary centres may improve prognosis but would definitely improve patient experience of care.

References;
1.Bruno Allolio. Clinical Review: Adrenocortical Carcinoma: Clinical update. Journal of Clinical Endocrinology and Metabolism. June 2006, 91(6):2027-37
2.Irina Veytsman. Management of Endocrine manifestations and use of Mitotane as a chemotherapeutic agent for Adrenocortical Carcinoma. Journal of Clinical Ooncology. September 2009, 27(27): 4619-29

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