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 secretary 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
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64 | Jan 2013 | Secretory | 8.5cm R adrenal tumor (Stage 3) | 12/05/13 | Y | None | In remission | 6 Months prior to a diagnosis being made
| | | | DELAY | | | | | | 
| 38 | Nov 2012 | Secretory | 20cm L adrenal tumor, cardiac & lung metastasis (Stage 4) | 15/01/13 | Y | Palliative | Cardiac RTX | RIP | 04/02/13
| | | | DELAY | | | | | | 
| 39 | Feb 2012 | Secretory | 8cm R adrenal tumor, IVC & lung metastasis (Stage 4) | 17/02/12 | Y | Ketoconazole | Metyrapone | RIP | 07/05/13
| | | | DELAY | | | | | | 
| 76 | Sep 2011 | Secretory | 13cm L adrenal tumor & lung metastasis (Stage 4) | N/A | Y | Ketoconazole | Metyrapone | RIP | 18/02/13
| | | | DELAY | | | | | | 
| 54 | April 2013 | Secretory | 11cm R adrenal tumor, cardiac,lung & liver metastasis (Stage 4) | 11/04/13 | Inoperable | Y | Metyrapone | RIP | 26/04/13
| | | | DELAY | | | | | | 

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: