A Rare Occurrence of Adrenal Leiomyosarcoma

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Clinical case

- A 61-year-old Caucasian female was being investigated under the gastroenterology team for ***chronic abdominal pain.*** She reported pain and nausea triggered by sweets.
- CT scan of the abdomen and pelvis did not show any pathology other than an ***incidental 2.3cm right adrenal nodule***. She was then referred to our endocrine team.
- She underwent CT and MRI of the adrenals to further characterise the lesion. These were reported as ***'indeterminate'*** but likely a ***benign adrenal incidentaloma***.
- Further testing confirmed a non-secretory tumour (24 hour urine metanephrines [3 samples]: negative; normal aldosterone/renin ratio, ODST: 71 nmol/L, normal LDDST)

Follow-up

- A follow up CT adrenal scan at 6 months showed an ***increase in the size*** of the adrenal nodule to ***3cm***.
- It was of ***heterogeneous density*** with delayed washout of contrast with a ***low-enhancing centre***.
- The increase in size of the tumour within 6 months and the characteristics seen on repeat CT adrenal scan were ***suspicious of malignancy*** and she was referred urgently to the tertiary centre for a surgical opinion.

Surgical findings

- As the pre-operative investigations suggested an ***adrenal tumour***, she was booked for a ***retroperitoneoscopic/laparoscopic right adrenalectomy.***
- However at surgery the tumour was even larger and found to be ***invading the IVC*** and clearly ***malignant***.
- A planned laparoscopic procedure was changed to an open procedure and a ***grade 2 right peri-adrenal leiomyosarcoma*** was resected with resection of the lateral wall of the IVC.
- Interestingly, the attached adrenal gland was ***normal***.
- Her sweet intolerance resolved after surgery.
- She is currently followed up in a dedicated leiomyosarcoma centre.

- ***Macroscopic description:*** Tumour measured 64mm by 40mm by 38mm. Ki67 proliferation fraction: up to 90%. Immunohistochemistry: tumour cells positive for SMA, desmin, MNF116 and h-caldesmon. Negative for S100p, Melan A, Inhibin, Calretinin and synaptophysin.

Results

Figure1: CT adrenal with contrast Feb 2015:
2.3cm right adrenal nodule. Pre-contrast: 48HU; early contrast 48-53HU; delayed contrast 81HU

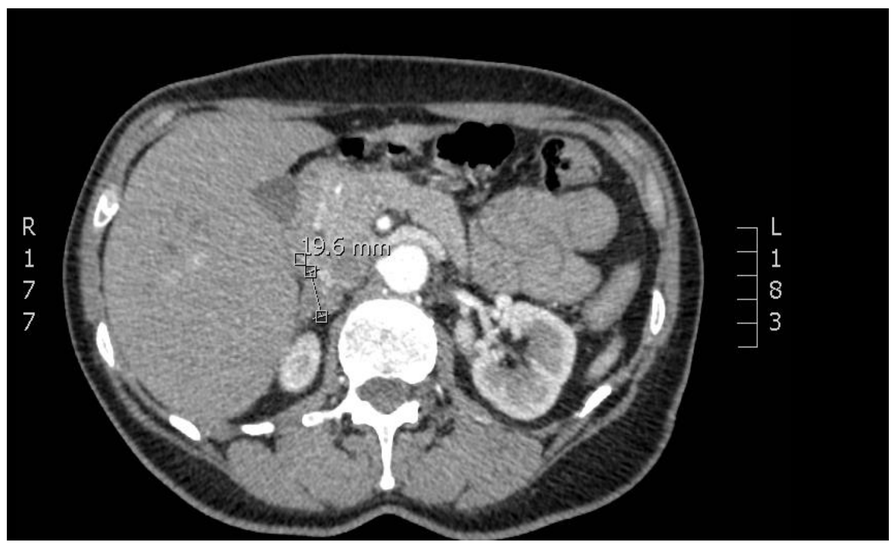


Figure2: **CT adrenal with contrast July 2015:**
3cm by 3cm right adrenal nodule.
Pre-contrast: 37HU; early contrast 50HU; delayed contrast 60HU

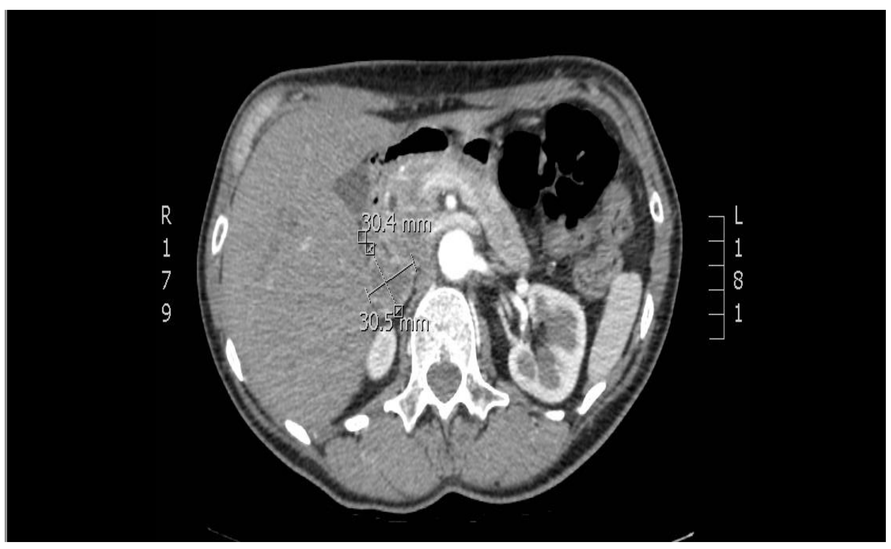
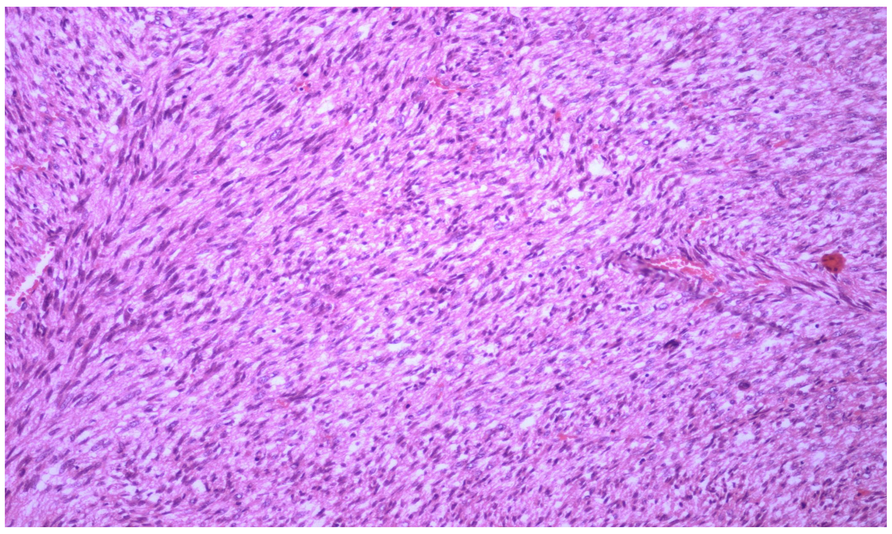


Figure 3: Histology
This H & E image is of the tumour at a higher x 10 magnification showing spindle cells exhibiting features of malignancy including mitotic activity and nuclear pleomorphism



CONCLUSION

- Mesenchymal tumours like leiomyosarcoma are associated with non-islet cell tumour-induced hypoglycaemia caused by the unregulated production of IGF-II and extensive glucose metabolism.
- This case highlights the importance of appropriate radiological assessment in adrenal incidentalomas by experienced adrenal radiologists.
- Monitoring of patients with suspicious looking lesions even if not meeting the initial criteria for surgery should be rigorous and discussed in a dedicated multi-disciplinary team.

References:

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