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 adrenal glands 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 blood glucose 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

Figure 1: CT adrenal with contrast Feb 2015: 2.3cm right adrenal nodule. Pre-contrast: 48HU; early contrast 48-53HU; delayed contrast 81HU

Figure 2: 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

v 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.

v This case highlights the importance of appropriate radiological assessment in adrenal incidentalomas by experienced adrenal radiologists.

v 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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