

Case of the Day

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Case courtesy of Drs. Zener R., Durant E., and
Mujoomdar A.

Clinical History

- 49F with refractory ectopic Cushing's Syndrome status post distal pancreatectomy, splenectomy, left nephrectomy and left adrenalectomy for a grade-3 ACTH producing pancreatic neuroendocrine tumor (pNET) involving the spleen, left kidney, left adrenal gland

Clinical History

- Refractory ectopic Cushing syndrome with labile hypertension secondary to significantly elevated AM cortisol of 1976 mmol/L (reference range: 119-618 mmol/L)
- Required aggressive intravenous anti-hypertensive management
- Complicated post-operative course precluded the patient from right adrenalectomy
 - Retroperitoneal hematoma with transfusion-dependent progressive anemia
 - Acute kidney injury
 - Developing sepsis

Clinical History

- PMHx: Pulmonary embolus
- Medications: Labetalol, Octreotide, Pip-Tazo, Vancomycin, Insulin
- Allergies: None



Figure 1. Post-operative contrast enhanced axial CT image demonstrates a hyperplastic right adrenal gland (4.3 x 1.6 x 2.0 cm), hypervascular liver metastases, and retroperitoneal hematoma

Proposed Procedure

- Since the patient was not a surgical candidate for right adrenalectomy, percutaneous microwave ablation of the right adrenal gland was planned
- Pre-procedural endocrinology and anesthesia consultations
 - Intra-procedural beta-blockade and arterial blood pressure monitoring for hypertensive crisis
 - Performed with patient under general anesthesia

Procedure Details: Percutaneous Adrenalectomy with Microwave Ablation

- CT-guided, posterior intercostal approach
- Two 17G 2 cm microwave probes advanced into the hyperplastic right adrenal gland
 - Needles at least 15 mm apart to prevent thermal injury
 - Several needle positions performed
- 4 cycles of MWA performed to allow for overlapping ablation zones



Figure 2. CT-guided placement of microwave ablation probes into the right adrenal gland

Procedure Details: Percutaneous Adrenalectomy with Microwave Ablation

- 1st cycle of MWA
 - 45 watts x 10 minutes
- Needles withdrawn 1.5 cm
- 2nd cycle of MWA
 - 45 watts x 10 minutes
- Needles repositioned into cranial part of gland
- 2 additional MWA cycles with same parameters

Procedure Details: Percutaneous Adrenalectomy with Microwave Ablation

- Post-ablation care
 - Monitoring of ACTH and AM cortisol
 - Initiation of glucocorticoid and mineralocorticoid supplementation
 - Ongoing therapy relating to liver metastases:
 - Octreotide
 - Chemoembolization of liver metastases

Decreasing morning cortisol post-percutaneous adrenalectomy using microwave ablation

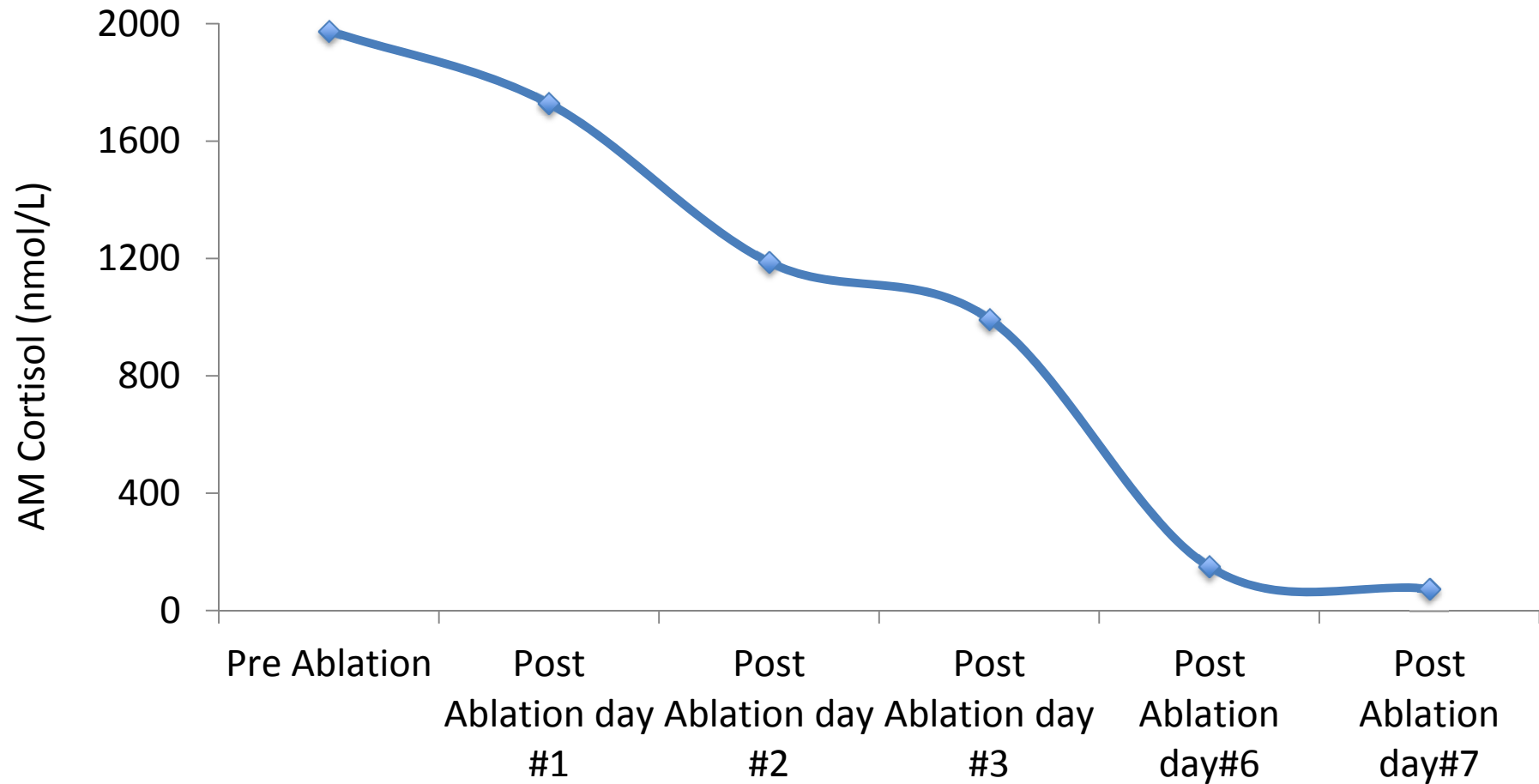




Figure 3. Contrast enhanced axial CT image post right adrenal ablation

Clinical Background: ACTH-producing Pancreatic Neuroendocrine Tumors

- Pancreatic neuroendocrine tumors (pNET)
 - Rare tumors; less than 5% pancreatic malignancies
 - Account for 8-18% of Cushing syndrome secondary to ectopic ACTH secretion ^{1, 2, 3, 4, 5}
 - 95% of ACTH-producing pNET are malignant
- Elevated ACTH results in excess adrenal cortisol secretion interfering with the normal H-P-A axis, leading to hypercortisolism
- Clinical features of hypercortisolism
 - Labile hypertension, hypokalemia, weight gain, purple striae, diabetes, hirsutism, acne, hyperpigmentation, bruisability, proximal myopathy, psychiatric changes

Clinical Background: ACTH-producing Pancreatic Neuroendocrine Tumors

- Traditional pNET management
 - Primary tumor resection +/- bilateral adrenalectomy to control ectopic Cushing syndrome
 - +/- Adjuvant loco-regional therapy if metastases
 - +/- Adjuvant hormonal related medical therapy
 - Post-operative monitoring for refractory Cushing syndrome with serial serum cortisol measurement
- Successful treatment results in primary adrenal insufficiency
 - Patients require lifelong glucocorticoid and mineralocorticoid supplementation

Clinical Background: Managing Refractory Hypercortisolism

- How are refractory hypercortisolism cases definitively managed?
 - Surgical adrenalectomy
 - Invasive
 - Need to be a good surgical candidate

Clinical Background:

Managing Refractory Hypercortisolism

- Is there a role for percutaneous ablation in non-surgical candidates?
 - Increasingly used to treat primary and secondary adrenal tumors in patients⁶
 - With unresectable disease
 - Who are poor surgical candidates
 - Previously reported to treat Cushing syndrome secondary to adrenal adenomas⁷
 - No prior reports of percutaneous ablation for treating ectopic Cushing syndrome

Clinical Background: Percutaneous Ablation

- The benefits of microwave ablation compared to radiofrequency ablation
 - Faster
 - Large ablative volume
 - Less susceptible to heat sink effects
 - N.B. Rapid temperature increase can occur with adrenal ablation predisposing patients to hypertensive crisis
 - As such, it should be performed under general anesthesia with appropriate pre-procedure anesthesia and endocrinology consultations

Conclusions and Teaching Points

- This is the first report detailing percutaneous adrenalectomy using microwave ablation to successfully treat ectopic Cushing syndrome in ACTH-producing pNET
- Percutaneous MWA is a safe and minimally-invasive intervention that can be considered in the treatment of non-surgical candidates with ectopic Cushing syndrome
- Follow-up serum morning cortisol should be performed to assess its effectiveness
- Multi-disciplinary management with IR, surgery, endocrinology and anesthesia, with monitoring for and treatment of hypertensive crisis is important

References

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