

ACTH-INDEPENDENT MASSIVE BILATERAL MACRONODULAR ADRENAL HYPERPLASIA

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Introduction

The estimated annual incidence of Cushing's disease ranges from 2 to 4 per million. Bilateral macronodular adrenal hyperplasia (BMAH) accounts for <1% of Cushing's syndrome causes.

Case report

We present a case of 58-year-old man who had been suffering typical clinical signs of hypercortisolism for 5 years. He presented hyperglycemia, arterial hypertension, central obesity, thin skin, hemorrhagic diathesis, edemas, weakness and emotional lability. Endocrine evaluation of adrenocortical axis proved hypercortisolism of adrenal origin with increased 24-hour urinary free cortisol (UFC) (792 mcg/24h [4.3-176]), a low doses dexamethasone test with not suppression (21.6 mcg/dL [<1.8]), a loss of cortisol circadian rhythm (16.1 and 20.6 mcg/dL [<7.5]) and low ACTH concentrations (<5 pg/mL [3.7-19.4]). Magnetic resonance imaging of the adrenal gland demonstrated bilateral adrenal masses (the right and the left adrenal measured were 3.1x5x3 and 5x7x3 cm respectively). A NP-59 iodocholesterol scintigraphy showed an increased uptake of both adrenal glands.

With the diagnosis of Cushing's syndrome ACTH-independent caused by bilateral adrenal masses surgery was made. Bilateral adrenalectomy was performed and histopathology revealed massive BMAH with multiple nodules ranging in size from several mm to 3 cm. The weight of right and left adrenal gland was 85 and 105 grams respectively.

After that, hypercortisolism was solved and patient's phenotype, diabetes and hypertension disappeared.

TABLE

	Before Surgery	After Surgery
24-hour urinary free cortisol [4.3-176 mcg/24h]	792	18 mcg/24h
Low doses dexamethasone test [<1.8 mcg/dL]	21.6	
Serum cortisol 9h [3.7-19.4]	16.1	0
Serum cortisol 21 h [<7.5 mcg/dL]	20.6	
Plasma ACTH [3.7-19.4 pg/mL]	<5	7.36
Glucose [70-110 mg/dL]	141	80
HbA1c [4-6%]	7.1	6.7

IMAGE

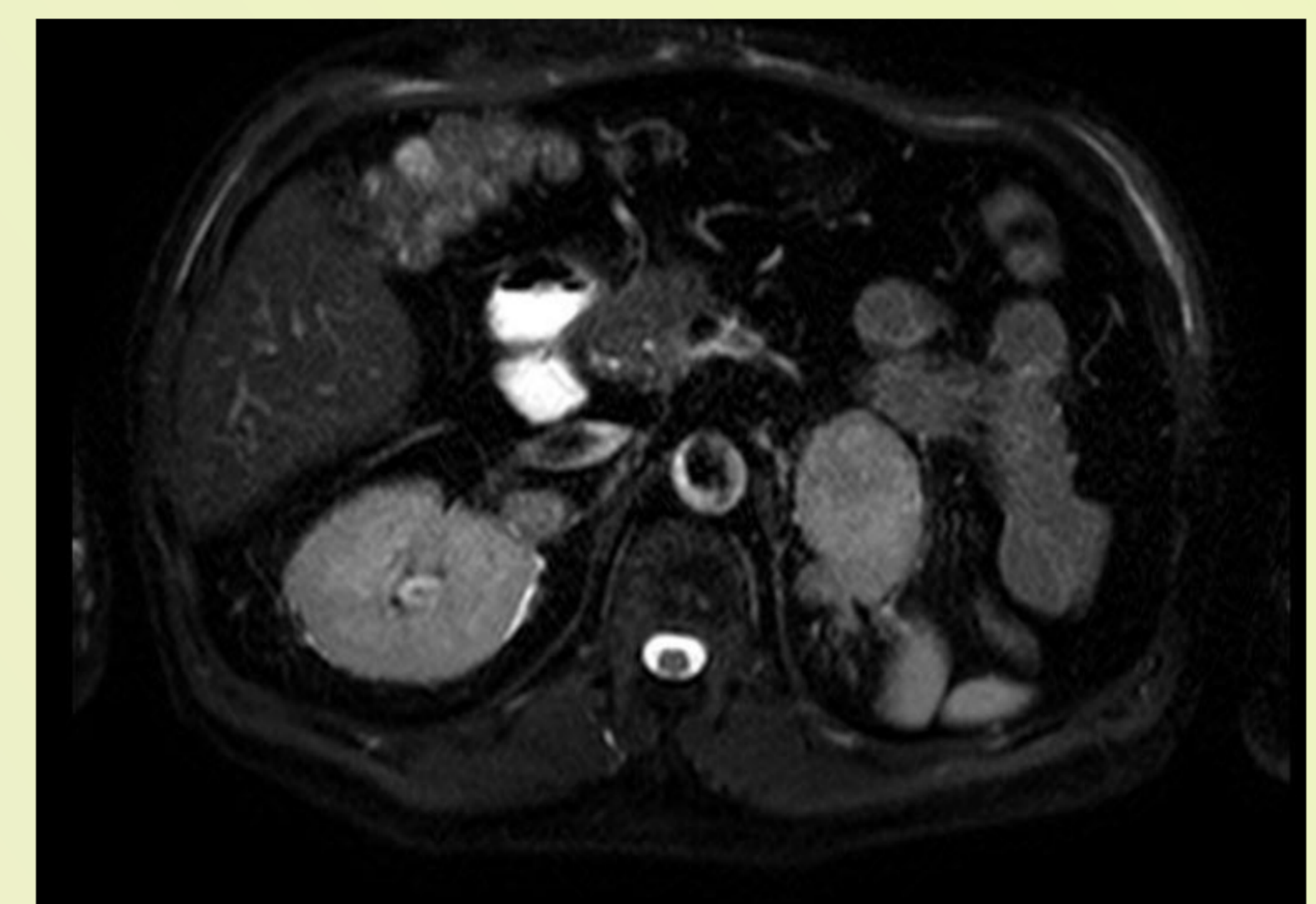


Table. Laboratory Tests.

Image. Magnetic resonance imaging suprarrenal showing bilateral adrenal masses.

Conclusions

BMAH is rare cause of Cushing's syndrome. It more often presents as an incidental radiological finding with underlying subclinical hypercortisolism. It is usually detected in the fifth or sixth decade of life. Bilateral adrenalectomy remains the main treatment for BMAH but unilateral adrenalectomy has been proposed in selected cases.

References

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