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HEMACHROMATOSIS ASSOCIATED WITH BILATERAL LARGE ADRENAL MYELOLIPOMAS –CASE REPORT AND REVIEW OF LITERATURE

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BACKGROUND: Myelolipoma may present with hematopoietic, fat and bone components. They are rare cortical, nonfunctioning, benign neoplasms, generally unilateral, and usually discovered by accident.

METHODS: Here we present a case of hemachromatosis due to β-thalassemia. He developed bilateral large adrenal myelolipomas.

RESULTS: A 44-year-old male with acute hyperglycemia and a history of β-thalassemia with frequent RBC transfusions developed hemachromatosis. He was incidentally found to have bilateral adrenal tumors and underwent left adrenalectomy due to rupture of the myelolipoma during an episode of a truma. Laboratory findings includ the following: MRI of adrenal glands: right 9 cm, left 12 cm 8 years ago; testosterone 23 ng/dL (241-800); FSH 0.3 uIU/mL (1.4-1.8); LH 0.7 uIU/mL (1.5-9.3); metanephrine 107 ug/day (52-341); cortisol 8 a.m. 8.17, 4 p.m. 9.42; AC/PC C-peptide 1.84/1.15 ng/mL; ferritin > 2000 ng/mL.

DISCUSSION: Most of the affected patients are men in their fourth to sixth decade of life. A giant adrenal myelolipoma was discovered in a 51-year old man who presented with a huge abdominal mass and abdominal pain. The resected tumor weighed 6,000 g and could represent the largest such tumor ever documented in the literature. Myelolipomas are rare cortical, nonfunctioning, benign neoplasms but can be associated with Cushing's type of endocrine disorders, Conn's syndrome, Addison's disease, medullary hyperplasia and congenital adrenal hyperplasia. The etiology is unclear but most favor metaplasia of the reticuloendothelial cells of blood capillaries. Asymptomatic small tumors (< 4 cm) may be monitored with watchful waiting. Symptomatic tumors (> 4 cm) should be extirpated because of the risk of spontaneous rupture with retroperitoneal bleeding. Bilateral adrenalectomy for big tumors should be followed by medical replacement with hydrocortisone.

Keywords: B-thalassemia, hemachromatosis, adrenal myelolipoma,