A Rare Case of Congenital Adrenal Hyperplasia with Giant Adrenal Myelolipoma

Texell Longoria-Dubocq, MD; Roberto Torres-Aguiar, Karla Ruiz-Vega, MD;
Rafael De Ayala-Hillmann, MD; Reynold Lopez-Enriquez, MD

Adrenal incidentalomas are tumors located in the adrenal glands and found on imaging done for purposes not related to adrenal disease. In other cases adrenal mases can be radiologically found when an adrenal-hormone secreting tumor is suspected, such as a pheochromocytoma or Cushing's diseases. Adrenal incidentalomas may be classified as functional or non-functional based on whether they produce hormones, such as aldosterone, cortisol, and androgens, or catecholamines. Studies indicate that around 8% of adrenal incidentalomas are adrenal gland myelolipomas (AGMs). AGMs are non-malignant masses that can cause the compression of vital organs and vessels if said masses become large enough. In patients with congenital adrenal hyperplasia (CAH), adrenocorticotropic hormone (ACTH) levels tend to be elevated due to the lack of adrenal-hormone production. Patients with CAHs are treated with steroids that suppress ACTH levels and prevent adrenal gland hyperplasia. Around 10% of AGMs are found in untreated CAHs. Our patient was a 36-year-old male who was on steroids due to CAH and intermittent abdominal pain; a CT scan revealed a large left adrenal mass that was displacing organs towards the right. Pathological analysis revealed an AGM exceeding 30 x 23.6 x 16.7 cm. This AGM is one of the largest ever to be reported in the literature. [*P R Health Sci J 2020;39:226-228*]

Key words: Adrenalectomy, Adrenal Myelolipoma, Congenita Adrenal Hyperplasia

Most adrenal gland myelolipomas (AGMs) are found incidentally on imaging or at autopsy. AGMs are soft, round, multinodular masses composed of adipose tissue and hematopoietic cells. According to past autopsy studies, the incidence of AGM is 0.2% in the general population, and they—AGMs—make up 8% of all incidentalomas. Incidence increases with age; AGMs occur 3 times more often on the left side and have a 1:1 female-to-male ratio. Bilateral AGMs have a 12% incidence in patients with adrenal myelolipoma (1). A report including 420 cases of AGM reported the average age at diagnosis to be 51 years; 10% of the cases were associated with untreated congenital adrenal hyperplasia (CAH) and 7.5% were associated with adrenal hypersecretion of aldosterone or cortisol (2). The case presented in this study is the largest AGM ever reported in a patient with CAH who is being treated with steroid replacement therapy.

Diagnosis

For the most part, adrenal masses are discovered on imaging that is being done for reasons not associated with the adrenal gland. AGM is a benign disease that rarely becomes symptomatic. Due to the varying amounts of adipose tissue and myeloid cells, AGMs appear echogenic or hypoechoic on US images (3). In a series of 86 cases, the studied AGMs were composed of 50 to 90% fat, and 24% contained calcifications that were visible on a CT scan. A diagnosis can be made with a CT scan showing fat attenuation of -10 to -20 HU, or on an MRI demonstrating a composition of at least 50% fat (4). If the CT scan findings are non-diagnostic, technetium-99m sulfur colloid scintigraphy—a technique that enhances the erythropoietic elements present in AGM—can be used (5). On occasion, an AGM can cause symptoms due to the synthesis of hormones. Therefore, adrenal mass functionality needs to be ruled out by measuring metanephrines and hormones, which can change preoperative and perioperative management. To that end, urinary and serum metanephrine levels can be measured, a urinary free cortisol or dexamethasone suppression test performed, the plasma aldosterone/plasma renin activity ratio determined, or hormone testing undertaken (1).

A percutaneous biopsy of an adrenal mass is rarely needed, since it would not provide additional information that would change how the mass is to be managed (1). If the history and clinical signs suggest the presence of metastasis or an infectious process, a biopsy may be indicated. Contraindications

Department of General Surgery, School of Medicine, University of Puerto Rico Medical Sciences Campus, San Juan, Puerto Rico

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Address correspondence to: Texell Longoria-Dubocq, MD, University of Puerto Rico Medical Sciences Campus, PO Box 365067, San Juan, PR 00936-5067. Email: texell.longoria@upr.edu

for such a procedure are coagulopathy and suspicions of pheochromocytoma, because of the potential risk of hemorrhage and hypertensive crisis, respectively (6).

Management

The most frequent indications of the necessity for the surgical resection of an AGM are abdominal pain, flank pain, the presence of a tumor having a diameter greater than 4 to 6 cm, that tumor's having an atypical radiographic appearance, and IVC compression. Large tumors should also be surgically removed when the patient is presenting with symptoms of organ compression, hemorrhage, or continuous growth. Each case should be managed individually, based on symptomatology (1).

For masses not requiring immediate surgical intervention, continued surveillance should be a clinical decision based on the imaging characteristics and size of the mass. Surveillance is not recommended for lesions that are less than 4 cm in size and that have clearly benign features on imaging (7). To evaluate whether a mass is growing, however, a follow-up CT scan either at 3 to 6 months (for a radiologically suspicious lesion) or 12 months (for a mass with benign characteristics) is recommended (8). Tumors larger than 4 cm should be considered for surgical resection due to the increased risk of adrenal carcinoma associated with such tumors, which risk has been identified in retrospective studies. Imaging characteristics should also be used to determine the need for surgical intervention (7). If an adrenal mass meets the above indications, a resection can be performed through an open or laparoscopic approach, depending on the size of the adrenal mass and the surgeon's preference (9).

Case

A 36-year-old obese male with a medical history of CAH, obstructive sleep apnea, depression, and epilepsy presented at our clinic with intermittent abdominal pain and difficulty breathing. His CAH was being adequately managed with oral hydrocortisone, 15 mg, twice daily; he was properly adherent to therapy and endocrinological follow-up was being performed regularly. A CT scan demonstrated a well-encapsulated left retroperitoneal soft-tissue mass measuring 30 x 23.6 x 16.7 cm (Figure 1). The mass was displacing the pancreas and spleen, anteriorly, as well as compressing the left hemidiaphragm. No compression of vascular structure was seen. The left adrenal gland could not be identified. According to the laboratory results, the mass was non-functional; a percutaneous biopsy indicated that it was an AGM.

We decided to manage the AGM surgically, due to the patient's symptoms. An open approach via a midline laparotomy was taken. The mass was dissected free from the surrounding structures. Figure 2 shows the adrenal mass on an instrument table. The patient was discharged home on post-operative day 7 and seen at an outpatient clinic 1 week later. The surgical wound healed adequately, with no complications. A pathology report indicated that the AGM measured 31 x 23 x 9 cm.

Conclusion

Metaplasia of reticuloendothelial cells in the capillaries of the adrenal gland is theorized to be the origin of myelolipomas. Stimuli such as infection, stress, necrosis, and prolonged exposure to adrenocorticotropic hormone (ACTH) may contribute to the formation of myelolipomas (9). CAH increases the risk of AGM in patients who are not treated with steroid replacement, since elevated ACTH levels stimulate adrenal gland growth (10). This case represents the second largest AGM reported in the literature, the largest being 31 x 24.5 x 11.5 cm (11). Other cases have demonstrated the bilateral enlargement of AGMs in patients not adequately treated with steroids (2). Our case was unique since even after the adequate management of his CAH (including his proper adherence to therapy), the patient developed the aforementioned large AGM measuring 31 x 23 x 9 cm. The proper treatment of CAH prevents the growth of the adrenal gland by suppressing ACTH levels, but not in this particular patient. Our conclusion is that factors other than ACTH stimulation play an important role in the development of these large tumors.

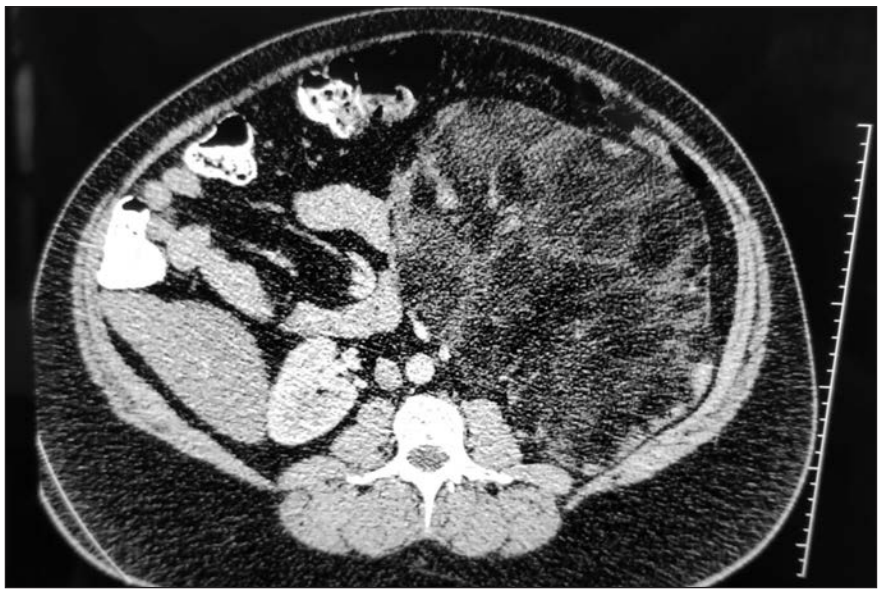

Figure 1. Pre-operative abdominal CT scan demonstrating a left adrenal gland mass measuring 30 x 23.6 x 16.7 cm.

Resumen

La mayoría de los tumores en las glándulas adrenales son no funcionales y benignos. Hoy día muchos de ellos son descubiertos por estudios radiológicos por síntomas

no relacionados a enfermedades de las glándulas adrenales. Este diagnóstico se conoce como "incidentaloma" como es descrito en la literatura médica en inglés. Los "incidentalomas" pueden ser funcionales o no funcionales dependiendo si secretan hormonas o catecolaminas. Mielolipomas adrenales ocupan un 8% de la prevalencia de tumores adrenales. Los mielolipomas son tumores benignos que crecen cuando son estimulados por la hormona adrenocorticotropica, o ACTH, por sus siglas en ingles. Pacientes que padecen de hiperplasia adrenal congénita tienen altos niveles de ACTH debido a la falta de producción de hormonas adrenales. Estos pacientes se tratan con medicamentos que remplazan estas hormonas. Al ser tratados medicamente, los niveles de ACTH se normalizan y evitan el crecimiento de las glándulas adrenales o tumores adrenales. Alrededor de 10% de los pacientes con hiperplasia adrenal congénita no tratadas desarrollan mielolipomas. Aquí presentaremos un paciente de 36 años con un mielolipoma adrenal que medía 30 x 23.6 x 16.7 cm. El paciente padecía de hiperplasia adrenal congénita cual estaba adecuadamente tratada con esteroides. Presentamos una de las masas adrenales más grande descrita en la literatura.

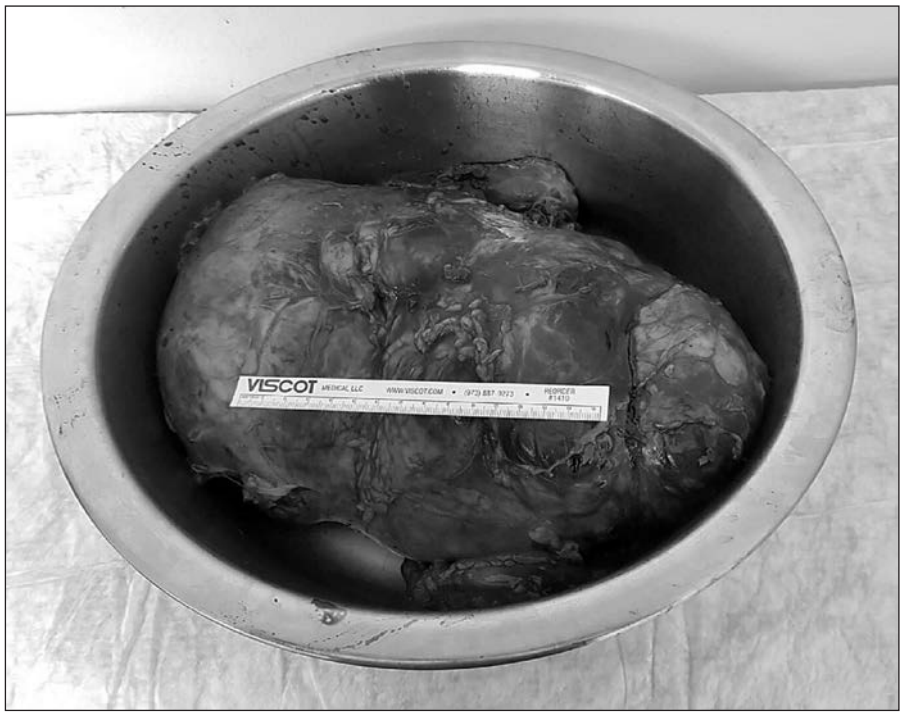

Figure 2. Left adrenal gland mass, after surgical removal, measuring 31 x 23 x 9 cm.

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