

Huge adrenal mass

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Abstract

Background and Aim: Benign myelolipomas are rare tumors of the adrenal gland. Their incidence has been reported to be less than 0.4%, with a few growing to the size of giant bilateral adrenal masses. The development of these tumors has been associated with congenital adrenal hyperplasia (CAH) as in this case and with other adrenal hormonal diseases.

Methods: A 38-year-old patient with a history of ambiguous genitalia at birth, raised as female and later virilized and transitioned to male, presented with abdominal distension and bilateral adrenal masses. Imaging revealed large adrenal masses (the left being larger) and the presence of female internal reproductive organs. Lab tests showed elevated androgens, ACTH, and 17-OHP, confirming classic simple-virilizing congenital adrenal hyperplasia (CAH). The patient underwent a left adrenalectomy, which confirmed a myelolipoma, and is now on hormone replacement and will be monitored for the growth of the right adrenal mass. Genetic testing revealed a 46,XX karyotype, despite the male phenotype.

Results: Myelolipomas are rare, benign tumors composed of fat and blood-forming tissue, most commonly found in the right adrenal gland. While typically non-functional, they can sometimes be linked to endocrine disorders like Cushing syndrome. "Giant" myelolipomas are defined as those over 10cm in diameter and their cause is unclear but may be related to elevated ACTH levels. Management of these tumors is case-specific, with small, asymptomatic tumors monitored and larger or symptomatic ones often surgically removed. Excess ACTH is thought to play a role in the tumor's growth as it is associated with other conditions.

Conclusion: Persons with CAH may be at increased risk of developing adrenal myelolipomas, particularly if their CAH is poorly controlled. How and whether chronic exposure of the adrenal glands to high corticotropin levels increases the risk of developing myelolipomas remains a matter of speculation. The coincidence of myelolipoma and congenital disorder with subsequent overproduction of the adrenocorticotropin hormone and androgens, might be explained by the incipient of myelolipoma through chronic hormonal stimulation of the adrenal gland cortex. However, the etiopathogenesis of myelolipoma is still unclear.

Keywords: Benign myelolipomas; Adrenal gland; Giant; Adrenal hormones