Adrenocortical carcinoma associated with giant bilateral myelolipomas in classic congenital adrenal hyperplasia

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ADRENOCORTICAL CARCINOMA ASSOCIATED WITH GIANT BILATERAL MYELOLIPOMAS IN CLASSIC CONGENITAL ADRENAL HYPERPLASIA

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Short title:
ACC IN GIANT MYELOLIPOMA AND CONGENITAL ADRENAL HYPERPLASIA

Conflict of interest: none declared

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32-year-old male patient, diagnosed with classic form of congenital adrenal hyperplasia (CAH) due to 21α-hydroxylase deficiency as a neonate was referred to the Department of Endocrinology in March 2018, due to poor disease control and rapidly enlarging bilateral adrenal masses (Fig.1A&B). A year and a half before the admission the adrenal biopsy was performed (in other hospital) which revealed benign tumor myelolipoma. However due to the mass effect and heterogenic appearance on imaging the left side tumor was surgically removed two years after its initial presentation. Macroscopic examination revealed the mass of 350 millimeters in the longest dimension and almost 2 kilograms of weight (Fig.1C). Microscopically, the tumor was composed mainly of “myelolipoma” tissue, with adrenocortical cell “islands”. However, when assessed using Weiss score, these foci were diagnosed not as adrenocortical adenoma, but adrenocortical carcinoma (ACC). From among 9 histologic criteria of malignancy, 6 were positive (with 3 necessary to diagnose ACC). These were: nuclear grade (Fuhrman’s III), atypical mitotic figures, 10% of clear cells, diffuse architecture, sinusoidal and capsular invasion (Fig.1D). Therefore, chemotherapy with mitotane was introduced due to large size of the tumor, previous biopsy and testicular adrenal rest tumors. The patient is finally adhering to rigorous hormonal treatment and monitoring regimen. He is currently waiting for right-sided adrenalectomy.

Myelolipomas are uncommon, benign and non-functioning adrenal masses, composed of mature adipose tissue and haemopoietic elements. They usually require surgical excision only when become large and symptomatic [1]. However, in very rare cases, collision tumors of two or more histologically distinct compounds are encountered, like myelolipoma co-existing with adrenocortical adenoma or carcinoma, for example [2]. Increased prevalence of adrenal tumors has been reported in poor-compliance CAH patients, due to chronic adrenocorticotropic hormone (ACTH) stimulation which acts as a growth and metaplasia-
inducing factor. However, less than 20 cases of giant bilateral myelolipomas in CAH patients have been reported to date and the role of ACTH or other hormones in their development is still uncertain [3].

The presented case highlights the need for proper and careful management of CAH patients. However, it also shows that the risk of malignancy exists even in tumors considered as benign. Adrenocortical carcinoma is a rare neoplasm with low survival rates [4]. Histopathological diagnosis based on multiparameter scoring systems is often challenging. In general, adrenal tumors should preferably be excised and examined as a whole, thus indications for biopsy are very limited and only to cases in which the diagnosis would change the course of treatment. In our patient the biopsy was completely misguided. What is more, the suspicion of ACC constitutes strong contraindication for biopsy due to the possibility of neoplasm cells spread in the needle conduit and so mitotane treatment was introduced as this kind of additional treatment may be associated with better outcome [4]. Mitotane has a direct and selective cytotoxic effect on adrenal cortex cells, thereby inducing permanent atrophy of any normal or malignant adrenal tissue that is left after the operation. However, also in patients with CAH that are not candidates for surgery, it may play a role in restoring fertility impaired by testicular adrenal rest tumors [5].

References:


Fig 1. A&B. Abdominal CT scan of giant myelolipomas. For the previous 2 years of poor hormonal control, rapid growth of adrenal masses from 22x13x20 millimeters on the right side and 73x29x49 millimeters on the left side to 67x48x27 (black arrow) and 198x191x120 millimeters (white arrow), accordingly, had been observed.

C. Polycyclic, solid, encapsulated left adrenal collision tumor of 350x110x90 millimeters and 1780 grams of weight.

D. Hematoxylin-eosin (H&E) staining histology of the tumor seen at 200-fold magnification in light microscopy. Black arrow A - adrenal cortical carcinoma. Black arrow B - myelolipoma