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SUN-401 New Right Adrenal Nodule and Beckwith-Wiedemann Syndrome in a Girl with Left Adrenocortical Tumor Resection

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Abstract

Background: The adrenocortical tumors have been reported in patients with Beckwith-Wiedemann syndrome, but the optimal way of management is not well studied. We report a case of left adrenocortical adenoma status post left adrenalectomy who subsequently diagnosed with Beckwith-Wiedemann syndrome, thereafter found to have a growing right adrenal nodule. **Clinical Case:** A 9-year-old female who was initially seen in Endocrinology clinic when she was seven months old. At that time, she was referred for pubic hair that was preceded by acne around the age of 5 months. Screening labs showed elevated DHEA 137 mcg/dl and testosterone 161 ng/d. MRI revealed a 2.5 cm well defined rounded homogeneously enhancing soft tissue mass located at the anteroinferior to the lateral limb of the left adrenal gland. She had left adrenalectomy at seven months of age. Pathology of the mass revealed adrenocortical adenoma. She was referred to hematology and oncology. Very mild hemi-hypertrophy started to appear as patient got older, so further work-up revealed loss of methylation of DNA at the DMR2 which is consistent with Beckwith Wiedemann Syndrome. All symptoms had resolved, and she had multiple screening including Ultrasounds and AFP. All her images and AFP remained normal. Then at the age of 5 years, she started to have new pubic hair and acne. CT scan revealed right adrenal thickening and nodularity with a 5 mm nodule. It has been monitored with frequent MRI alternated with ultrasounds. The nodule has increased in size gradually over the years to 15.79 mm x 8.72 mm. PET scan showed focal increased FDG activity within the 1.1 cm x 0.7 cm right adrenal nodule. Bone age was normal. After consultations of multiple national centers, there was no consistency about how to manage adrenal tumors in patient with BWS. Therefore parents were given choices of total right adrenalectomy vs partial adrenalectomy vs observations. Parent chose watching waiting, so decision was made to follow up with MRI every three months. **Conclusions:** Although patients with BWS get screened with abdominal U/S frequently for adrenal tumors as the part of the tumor surveillance, there is no consensus on management options if such tumors are detected. This case raises the need for uniform guidelines to outline the optimal approach of management of



adrenocortical tumor in patient with BWS.

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