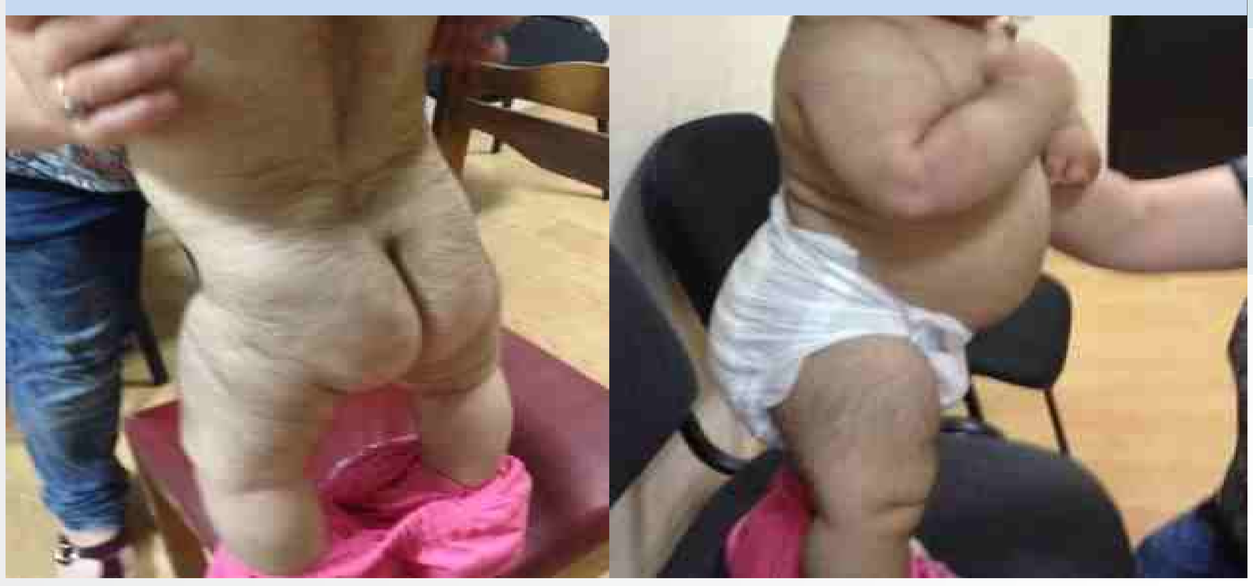
Rare Case of Androgen Producing Tumor in 14 Month Old Girl.

N.Kheladze[1], E Lundberg[2] , N. Totogashvili[1] , T. Tkeshelashvili[3]
[1]M. Iashvili Children's Central Hospital, Tbilisi, Georgia
[2] Institute of Clinical Science/Pediatrics, Umeå University, Umeå, Sweden
3 A. Tsereteli State University, Medical Faculty, Kutaisi, Georgia

Background: Adrenocortical tumors (ACT) rare in children. Mostly occurs in younger age before 4 years and predominance of girls.
ACT represents 1.3% of all carcinomas in paediatric age group and 0.2% of all pediatric neoplasms.
Symptomes of endocrine hyperfunction is present in 80-90 percent of cases.
Case report: 14 month old girl presented with signs of progressive hirsutism started first few months of life. She was referred to our clinic due to suspect of virilizing CAH.
Physical exams showed virilization in combination with signs of overproduction of other adrenal hormones: pronounced hirsutism (Ferriman Gallway score 25), clitoromegaly, hoarse voice, widespread acne on face. She has varus deformation of legs and a signs of pronounced rickets.
She has not received vitamin D supplement. She had accelerated growth.
Her weight was over 98th percentile and height corresponded to 75th percentile for age and sex.

Before surgery



Test	Result	Reference Range for Age
17-OHP	3.51 ng/ml	0.16 – 1.02
Testosterone	7.06 ng/ml	0.03-0.32
DHEAs	2.58 µg/ml	0.05-0.55
Cortisole	232 ng/ml	30-210
Blood Glucose	75 mg/dl	60-100
Insulin	16.51 MicU/ml	< 11

Advanced Bone age, more than 2 years compared to chronological was noted.
Abdominal ultrasound revealed enlarged liver and spleen. MRT showed tumor size 5.5/4/4.7 cm in left upper quadrant in retroperitonium

Complete resection of encapsulated tumor with no connection to adrenal s was performed.
No signs of local invasion or metastasis was found.
Tumor was assessed locally as well as by Institute of Pathology in Kiel, Germany

Pathology Report
"The tumor consists of medium to large tumor cells of varieble size. Nuclei are round to avoid, focally very pleomorphic and enlarged. Cytoplasm is broad and pale to strong eosinophilic. The tumor cells are grown in large sheets,focally with prominentrabecular pattern.There are small areas of necrosis and large haemorrhages. In the periphery there are rests of the adrenal gland. **9 mitosis in 50 HPF. Ki 67 stainig is 5 % in the average, focally up to 15 %** Immunohistochemically the cells express **inhibin, Melan A and synaptophysin. Negativity for S100 and chromogranin.**
<u>Conclusion:</u> It is a pleomorphic tumor with positivity for inhibin and Melan A adjacent to the adrenal gland which I would classify as an Adrenal cortical tumor.
Based on the information we have it is difficilte to decide whether it is an adenoma or already a carcinoma."

3 month after surgery

1.5 year after surgery

Postoperatively a brief episode of hypertension and polyuria occurred; therefore 24-hour blood pressure inpatient monitoring was conducted twice.
The ambulatory BP monitoring was recommended further.
Testosterone was normalized in a week after surgery.
Follow up visits every 3- 6 months, later showed no signs of metastasis, change in appearance with progressive decrease of virilisation and hirsutism to Ferriman Gallway score 9. Hormonal profile is normal, re-measured on multiple occasions. CT scan of chest and abdomen and bone scans were conducted after one year post-surgery, with no signs of metastasis. She is persistently overweight and still has pronounces varus deformity of the legs.

Nothing to disclose