Functional adrenocortical oncocytoma – a rare cause of progressive virilization and secondary amenorrhea

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INTRODUCTION

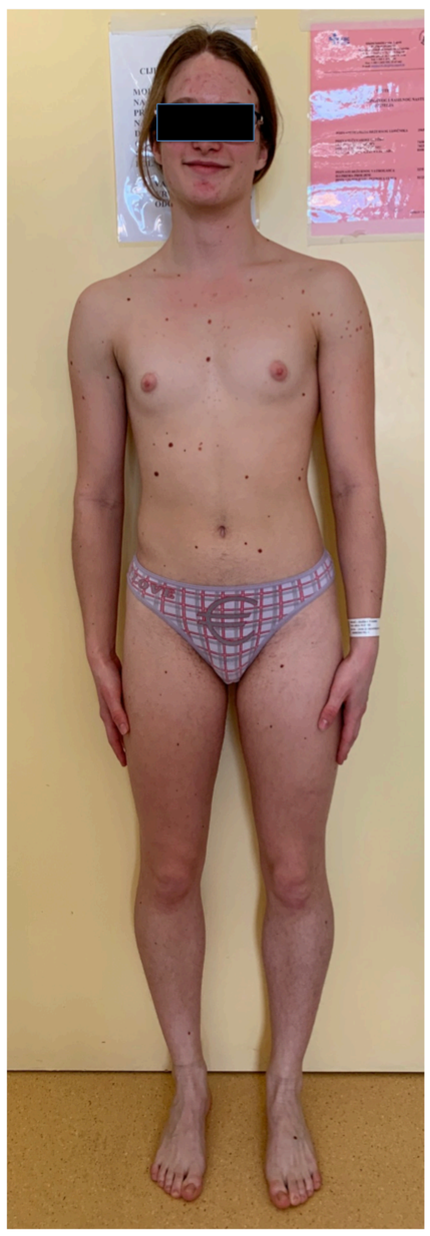
Oncocytomas are rare epithelial tumors that can be found in various tissues such as kidney, salivary and endocrine glands.

Adrenocortical oncocytomas (AON) are very rare tumors with around 160 patients described in the literature. Generally they are regarded as benign and mostly hormonally nonfunctional. When hormonally active, these tumors produce adrenal steroids resulting in various clinical presentations such as virilization, feminization, and Cushing or Conn syndrome.

Until now, only 8 pediatric patients with functional adrenocortical oncocytomas (FAON) have been described in literature.

Table 1. Laboratory findings pre-operation, 7-days and 4 months post-operation.

	pre-operation	7 days post-operation	4 months post-operation	normal range
LH	**0.1**		6.8	1.8-11 U/L
FSH	**0.3**		4.8	0.2-7.8 U/L
E2	92		416	47-517 pmol/L
DHEAS	**26.8**	3.4	3.8	2-10 umol/L
androstenedione	**21.8**	3.9	10.7	1-12 nmol/L
testosterone	**17**	0.7	1.4	0.4-1.7 nmol/L
17-OHP	5.9			0.3 – 6 nmol/L
ACTH	5.4		6.7	1.6-13.9 pmol/L
cortisol	594		482	138-690 nmol/L
aldosterone	286		314	90-830 pmol/L
PRA	0.8		1.2	0.2- 5.7 ug/L/h
beta-HCG	negative			
AFP	2.7			<7 ug/L



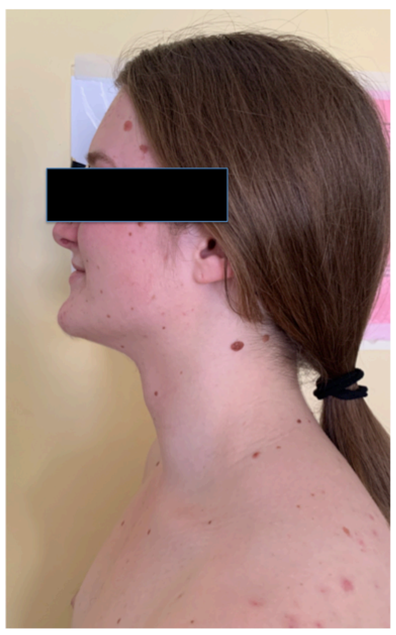


Figure 1. The girl at the age od 15.5 years. Note masculine apperance.

CASE REPORT

The patient is 15.5 year old girl referred for secondary amenorrhea lasting 8 months. The girl is a professional athlete. (Figure 1)

Slowly progressive virilization was reported for almost 2 years. At presentation she had deep voice, acne, hirsutism (Ferriman-Gallwey score 22), clitoromegaly and atrophic breasts. Blood pressure was normal. Laboratory findings are listed in Table 1.

Abdominal CT scan showed right adrenal gland mass measuring 4 cm in diameter. (Figure 2.)

Subsequently laparoscopic right adrenalectomy with lymphadenectomy was performed. Pathohistological diagnosis revealed ***oncocytic adrenocortical tumor*** with benign characteristics according to Wieneke criteria and Bisceglia classification. Adrenal androgen levels normalized completely after the surgery and the girl regained menstruation 1.5 month following tumor extirpation.

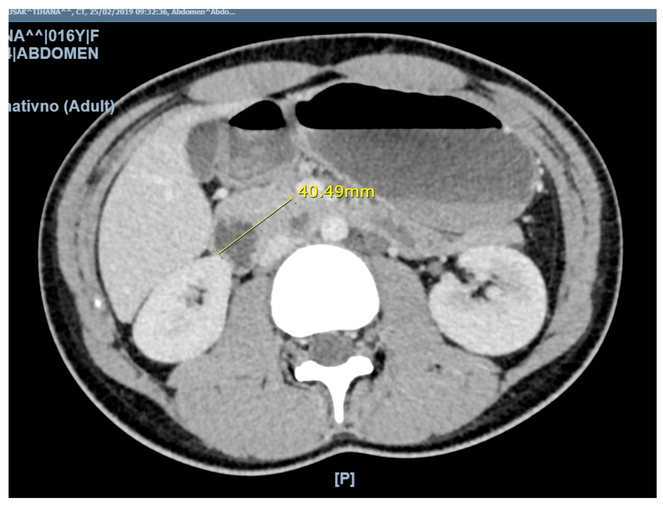


Figure 2. Abdominal CT scan showing tumor in right adrenal gland.

DISCUSSION/CONCLUSION

Functional adrenocortical oncocytomas are very ***rare tumors*** with yet unidentified pathogenesis and potential risk factors for their development. They are mostly discovered in adults, but can also be found in ***children***. There is no specific age distribution in children with FAON.

Unlike adults, all children with FAON presented with ***right sided*** adrenal mass and very strong ***female preponderance*** (8/9). Most of the patients with FAON (7/9), including our, presented with symptoms of ***androgen excess***. All but one were pathohistologically classified as ***benign*** at the time of diagnosis. Follow-up time ranged from 1-84 months in children with FAON. None of them had signs of disease recurrence at that time.

Due to extreme rarity of this tumor in children ***no clear evidence regarding its' true potential***, neither specific management guidelines exist. ***Long-term follow-up*** of these children should be recommended.