Diagnostic Dilemma in a Patient with Central Precocious Puberty: Ovarian Steroid Cell Tumor

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INTRODUCTION

Ovarian steroid cell tumors (OSCT) are rare sex cord-stromal tumors of the ovary and comprise <0.1% of all ovarian tumors.

CASE

We herein report a case of an OSCT in a 9-year-old girl patient who presented with central puberty preocious unresponsive to gonadotrophine relasing hormon analog (GnRH) therapy.

CASE

A 9 year old girl presented with breast development noticed 4 months ago and menarche 3 months ago.

Her height was 141,5 cm (+1,3 SDS), body weight was 35,4 kg (+0,9 SDS) and body mass index was 17,6 kg/m^2 (+0,5 SDS). Her thelarche was Tanner stage 3, and pubarche was Tanner stage 2. She had dysmorphic face with frontal bossing and flat nasal bridge. She also had hypertrichosis and acnes.

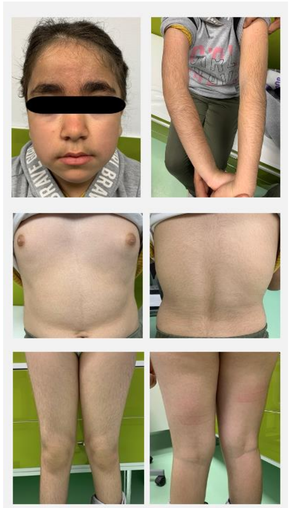
Laboratory results were as follows: follicle-stimulating hormone (FSH): 12,2 U/L, luteinizing hormone (LH): 8,1 U/L and estradiol: 93 ng/L. Bone age was 12 years.

On admission pelvic ultrasonographic (US) examination uterus was 56x20 mm, right ovary 34x16 mm, left ovary 35x16 mm, and a few millimetric follicle cysts were observed.

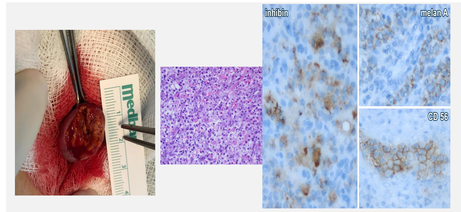
Cranial magnetic resonans imaging (MRI) was normal.

GnRH therapy (leuprolide acetate therapy 3,75 mg/28 days) was initiated due to centrally rapidly progressing puberty and early menarche. On her control examination 3 months later, growth rate was 6,7 cm/year, breast development Tanner stage 3, and hyperpigmentation of the nipples were detected. LH and estradiol were 4.4 U/L and 420 ng/dl (very high) respectively. She did not show any clinical improvement in the third month of treatment.

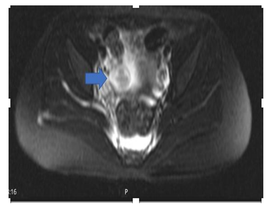
Her pelvic US repeated because of increase in estradiol; uterus was 65x36x24 mm, endometrium double foldable 10 mm, fundus/cervix ratio 1.5. Right ovary was 34x28x17 mm (8.5 ml) left ovary was 15x12x10 mm (0.9 ml). A solid mass 26x20x14 mm defined as homogeneous and hyperechogenic according to ovarian parenchyma, and hypervascular. The mass in the ovary was removed laparoscopically using the ovarian-sparing surgery technique.



	At Admission	Pre-op	Post-op
FSH	12,2 U/L		2,7 U/L
LH	8,1 U/L	4,4 U/L	1,13 U/L
E2	93 ng/dl	420 ng/dl	<20



The histopathological examination revealed benign OSCT-not otherwise specified (NOS) composed of cells positive for inhibin, Melan A, GATA3, CD10, CD31, CD56 and negative for estrogen receptor, cytokeratin, P53, desmin, S 100, and placental alkaline phosphatase.



A solid mass 26x20x14 mm defined as homogeneous and hyperechogenic according to ovarian parenchyma, and hypervascular.

CONCLUSIONS

In this case peripheral puberty due to OSCT was combined with central rapidly progressing puberty. Estrogen-secreting ovarian masses may not be detected by USG when they are still small in size. Radiological imaging may be repeated, especially in rapidly progressing puberty cases showing clinical and hormonal progression despite treatment.

REFERENCES

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