PRINCE OF SONGKLA UNIVERSITY
FACULTY OF MEDICINE

Interhospital conference

Case 3

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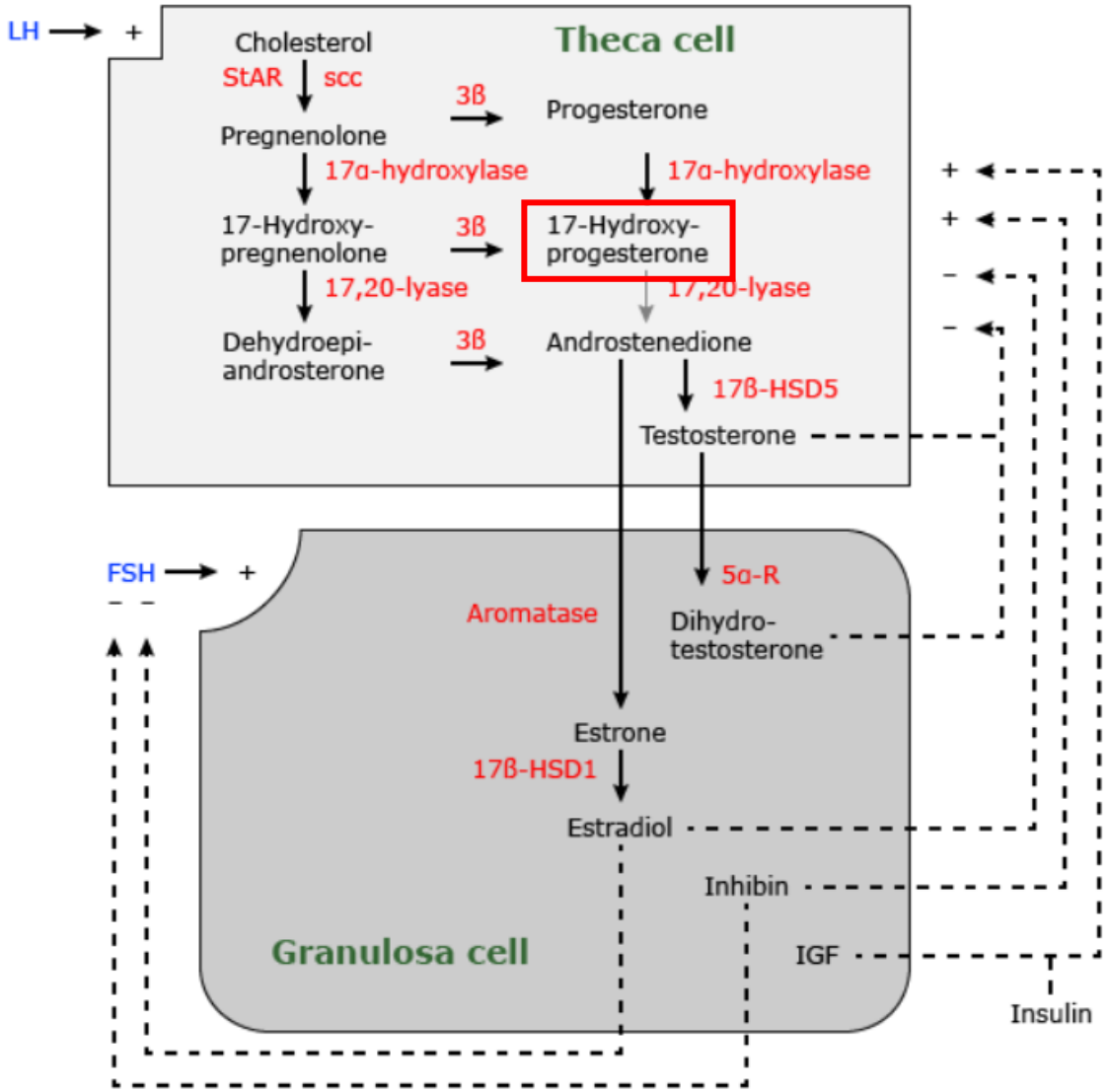
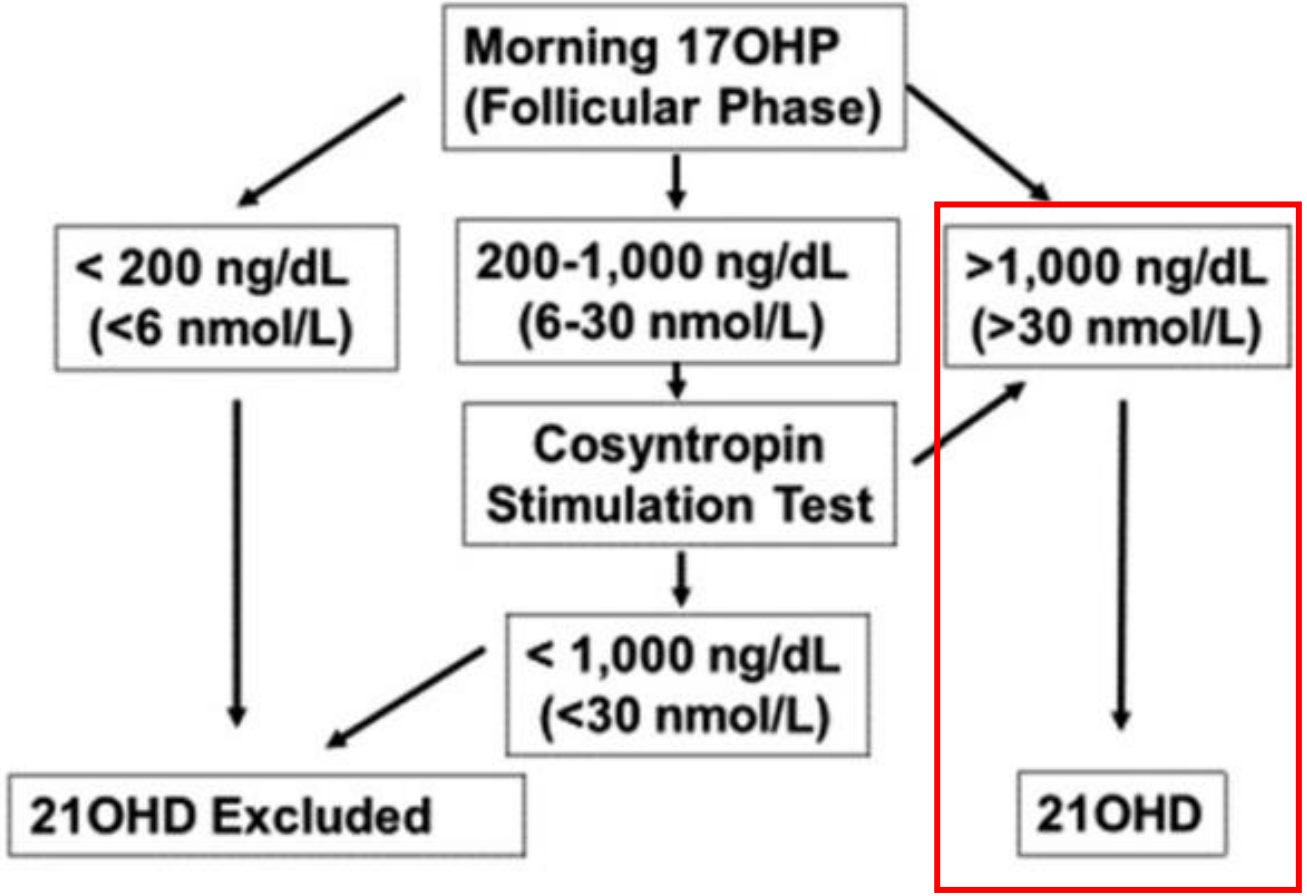
Ovarian steroid biosynthetic pathways





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Clinical presentation	Non-classic CAH	Ovarian tumor
Menstrual cycle disorders	Yes	Yes
Hirsutism	Yes	Yes
Acne	Yes	Yes
Alopecia	Yes	Yes
Clitoromegaly	Yes	Yes
Infertility	Yes	Yes
Age of onset	Adrenarche	Any
Progression	Gradual	Rapid

Case Report

Hyperandrogenism, Elevated 17-Hydroxyprogesterone and Its Urinary Metabolites in a Young Woman with Ovarian Steroid Cell Tumour, Not Otherwise Specified: Case Report and Review of the Literature

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Table 1: Pre- and postoperative laboratory results.

Tests (serum/plasma)	Concentration (before operation)	Concentration (2 weeks after operation)	Concentration (4 weeks after operation)	Reference interval
Luteinizing hormone (IU/L)	10.0	4.5	—	2.4–12.6 (follicular phase) 14.0–95.6 (ovulation phase) 1.0–11.4 (luteal phase)
Follicle-stimulating hormone (IU/L)	5.0	4.0	—	3.5–12.5 (follicular phase) 4.7–21.5 (ovulation phase) 1.7–7.7 (luteal phase)
Estradiol (pmol/L)	161	302	—	98–571 (follicular phase) 177–1153 (ovulation phase) 122–1094 (luteal phase)
Progesterone (nmol/L)	—	18.7	—	0.6–4.7 (follicular phase) 2.4–9.4 (ovulation phase) 5.3–86 (luteal phase)
Testosterone (nmol/L)	10.6	0.7	0.5	<1.7
Prolactin (mIU/L)	See footnote[a]	See footnote[a]	—	<496
Androstenedione (nmol/L)	28.2	4.2	3.0	1.1–6.5
DHEA-S[b] (μmol/L)	6.0	6.7	5.9	1.0–11.7
17-OHP (nmol/L)	52	4.3	0.7	0.6–4.0 (follicular phase) 1.0–6.0 (luteal phase)
Cortisol (1 mg overnight dexamethasone suppression test) (nmol/L)	21	—	—	<50
Fasting glucose (mmol/L)	7.2[c]	6.0	6.5	≥7.0: diabetes mellitus
HbA1c (%)	6.9[c]	—	—	≥6.5%: diabetes mellitus
CA125 (kU/L)	17	—	—	<35
Alpha-fetoprotein (μg/L)	8	—	—	<9
Human chorionic gonadotropin (IU/L)	<1	—	—	<1 (premenopausal non pregnant)

[a]Macroprolactin present, value within reference limits after PEG precipitation. [b]DHEAS: dehydroepiandrosterone-sulphate. [c]Results obtained at diagnosis, before initiation of anti-diabetic medication.

Table 3: A summary of steroid cell tumors, not otherwise specified (SCT–NOS) with 17-OHP concentration reported (n = 21, including the current case). Please refer to Supplementary Table S1 for a detailed summary of all cases.

		n	Reference
Age (years)[b]	23 (3–68)[a]	21	[9, 11, 22–27, 29–35, 44–48]
Extraovarian (%)	14 (3/21)	21	
Tumor size (cm)[c]	4.9 (1–12)[a]	20	[9, 11, 22, 24–27, 29–35, 44–48]
Evidence of malignancy (%)	5 (1/21)	21	
Serum testosterone concentration (nmol/L)[d]	12 (1.2–37)[a]	21	[9, 11, 22–27, 29–35, 44–48]
Elevated testosterone concentration (%)	100 (21/21)	21	
Serum DHEA-S concentration (μmol/L)[d]	2.9 (0.6–19.7)[a]	14	[9, 11, 23, 24, 26, 27, 29–32, 44, 45, 48]
Elevated DHEA-S concentration (%)	28 (5/18)	18	[9, 11, 23, 24, 26, 27, 29–34, 44–48]
Serum androstenedione concentration (nmol/L)[d]	35 (6.3–78)[a]	15	[9, 23–27, 29, 31–35, 44, 45]
Elevated androstenedione concentration (%)	88 (14/16)	16	[9, 23–27, 29, 31–35, 44, 45, 47]
Serum 17-OHP concentration (nmol/L)[d]	48 (1.8–312)[a]	16	[9, 11, 23–27, 29–32, 35, 44, 45, 48]
Elevated 17-OHP concentration (%)	81 (17/21)	21	[9, 11, 22–27, 29–35, 44–48]
Positive 17-OHP response after 1–24 ACTH stimulation (%)[e]	20 (2/10)	10	[11, 25, 27, 29–31, 33–35]
Hormonal co-secretion (hormones other than androgens) (%)	33 (7/21)		
Cortisol	10 (2/21)	21	[9, 11, 22–27, 29–35, 44–48]
Estradiol	14 (3/21)		
Estradiol and cortisol	10 (2/21)		

[a]Results expressed as median (range). [b]Age at diagnosis. [c]Largest dimension. If tumor size was not reported, the size of the affected ovary was used. [d]For results reported as "larger than X units", the value of X was used. For cases in which the diagnosis was delayed, the values at the age of diagnosis was used. Cases with no values reported (stated as "normal"), suspected errors in reporting units or unknown conversion factor to SI units were excluded. [e]Defined as positive if commented to be increased from baseline by authors and peak 17-OHP >30 nmol/L. Otherwise, it was arbitrarily defined an increase in 17-OHP, at either 30 min or 60 min after ACTH stimulation, by equal to or more than 100% of the basal value, and a peak value >30 nmol/L.

Thank you