



Dysgerminoma Can Present as Isosexual Sexual Precocity: Beta -hCG Should Be Part of the Work-Up

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BACKGROUND INFORMATION

Ovarian cancer in the pediatric age group is rare: so rare that according to North American registries from 1992-1997, out of 1.6 million females with a cancer diagnosis, only 67,746 were ovarian cancers, and of those, only 302 were in the birth to 14-year-old group.¹ Germ cell tumors are the most common type of ovarian tumor found in children from birth to 19 years of age, and dysgerminomas are the most common malignant ovarian germ cell tumors. They are bilateral in 10-20% of cases. Tumor markers that may show an elevation include LDH and CA-125. Dysgerminomas typically exhibit rapid growth, and as a result, the patient generally complains of increasing abdominal girth and/or a palpable abdominal mass.¹ The survival rate for all stages of dysgerminomas is 85% (95% for stage IA).¹ In the case below, signs of precocious puberty led to the early diagnosis of a dysgerminoma. It is rare that the initial presentation of a dysgerminoma is isosexual sexual precocity; there have been only a few cases reported in the literature over the last ten years.

CASE PRESENTATION

A seven-year-old female presents with isosexual sexual precocity. The challenges of the diagnosis in this patient are delineated.

A seven-year-old female was referred to a reproductive endocrinologist. Her mother reported precocious sexual development. Physical examination revealed scant axillary hair; acanthosis nigricans on the back of the neck, the axillary region, and the inguinal region; and mild hirsutism on the back. The patient showed no neurologic deficits. Tanner stage III breasts and pubic hair were present.

The patient underwent an extensive work-up (including multiple ultrasound examinations and a CT scan of the pelvis) that revealed no obvious etiology. As a result of a negative work-up, the presumptive diagnosis was premature activation of the hypothalamic-pituitary axis even though

the patient had a pre-pubertal response to a GnRH stimulation test. The patient's bone age was noted to be advanced (age 11), and the patient was therefore started on a GnRH analog to block the idiopathic activation of her pituitary gland. The patient underwent urine hCG testing before administration of the GnRH analog, and in the third month her pregnancy test was positive. This was confirmed with a serum beta-hCG. Radiographic evaluation for a mediastinal mass and a transrectal sonogram of the ovaries were performed at this time. The transrectal sonogram under anesthesia revealed a 1-2 cm mass on the left ovary. The patient underwent immediate laparoscopy and left salpingo-oophorectomy. Pathology revealed a dysgerminoma that stained positive for hCG.

DISCUSSION

Differential Diagnosis of Precocious Puberty²

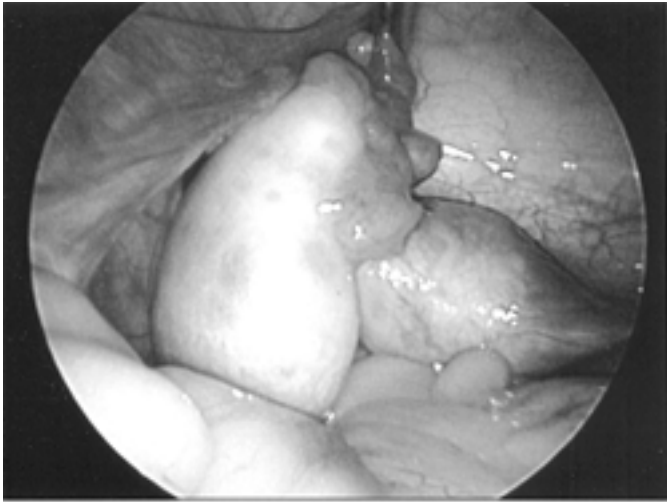
Findings	Idiopathic	CNS Tumor	McCune-Albright	Hypothyroid	Ovarian Tumor	Adrenal Tumor	Patient
Premature Thelarche	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Premature Adrenarche	Yes	Yes	Yes	Rarely	Yes	Yes	Yes
Neurologic Deficit	No	Yes	Yes	No	No	No	No
Bone age	Advanced	Advanced	Advanced	Advanced	Advanced	Advanced	Advanced
Abdominal /Pelvic Mass	Sometimes	No	No	No	Usually	No	Eventually Yes

Types of Germ Cell Tumor^{2,3,4}

	Immature Teratoma	Mature Teratoma	Embryonal Carcinoma	Endodermal Sinus tumor	Dysgerminoma
Bilateral	Malignant (2%-5%)	10 - 20%	Rare	Rare	5%-10%
Hormone Markers	N/A	N/A	AFP, Beta -hCG	AFP	LH and Sometimes Beta-hCG
Seen with Precocious Puberty	N/A	N/A	Sometimes	Occasionally	Occasionally

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Beta-hCG Should Be Part of the Work-up
(Continued)



From the differential diagnosis of precocious puberty, one can observe that the potential etiologies were narrowed by a thorough work-up that left only ovarian cancer and idiopathic causes as options. As no mass was detected on multiple radiological examinations, the initial treatment led to the correct diagnosis. In this case, the dysgerminoma produced beta-hCG, which was the cause of the patient's precocious puberty. If the beta-hCG had not been positive, then conservative management could have been maintained until the point at which the patient experienced abdominal discomfort secondary to growth of the mass.

In conclusion, there are several possible etiologies for sexual precocity. Among these is an ovarian tumor, and a germ cell tumor is the most common ovarian tumor in this age group. In general, beta-hCG is not a consistent part of the work-up. Beta-hCG is not routinely obtained for isosexual sexual precocity when no mass is present on radiologic examination. We propose that this test should be considered in any patient presenting with isosexual sexual precocity even in the absence of a radiologically detectable mass.

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