

Gynecomastia Secondary to Choriocarcinoma In A Man With Persistent Mullerian Duct

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Persistent mullerian duct is a rare syndrome secondary to anti-mullerian hormone deficiency. Here we describe the case of a phenotypically fertile male who presented with bilateral tender gynecomastia. Other than a left undescendent testicle, which was palpable in the inguinal canal, he had normal external genitalia. Physical examination revealed an abdominal mass, palpable in the supra pubic area. The ectopic testicle showed endometrial tissue. Abdominal mass proved to be choriocarcinoma. Later he developed distant metastasis to the lung and paraortic area

Key Words: Persistent mullerian duct, Choriocarcinoma, Gynecomastia

Introduction

Persistent Mullerian duct syndrome (PMDS) is a rare form of internal male pseudohermaphroditism caused by failure of the fetal testis to produce Mullerian inhibiting substance or failure of the tissues to respond to this hormone. Patients usually present in childhood with an inguinal hernia or cryptorchidism. As with undescended testes, these gonads are at an increased risk of malignant transformation. Gynecomastia¹⁻³ is a rare presentation of this syndrome.^{4,5} We describe

a case of phenotypically fertile male who presented with bilateral gynecomastia.

Case Report

Mr. H.A, a 30-year old farmer, was admitted because of bilateral tender gynecomastia and decreased libido in the last 2 months. He has two children. He had an undescendent left testicle, palpable in the left inguinal canal (12 X 30 mm). His right testicle was normal. He also had a palpable firm mass below the umbilicus. Sonographic studies revealed normal right testicle (35X 16 mm), maldecendent left testicle (30 X 12 mm), pelvic mass of 70 X 120 mm (lobulated and mixed echo with central necrosis and a pressure effect on the bladder). An abdominal CT scan confirmed these findings. Laboratory study results showed testosterone of 11.4 n mol/l (N: 10-35), LH 12.5 (N: 1.5-18), FSH 0.03 (N: up to 20) mIU/ml, Estradiol 240 pg/ml (N: <90), β HCG was 58 and 195 mIU/ml on 2 occasions (N: 0-5); α -Feto protein (AFP) was 1.3 (N: <4), FBS 75 mg/dl, WBC 13100, Hb 13.2, and ESR 21. Chest radiography showed 2 ill-defined nodes in the right and left lungs. In the first surgical operation, the ectopic testicle was removed. This showed a testis of 1.5- \times 2 \times 3.5 centimeter with a 3cm long spermatic

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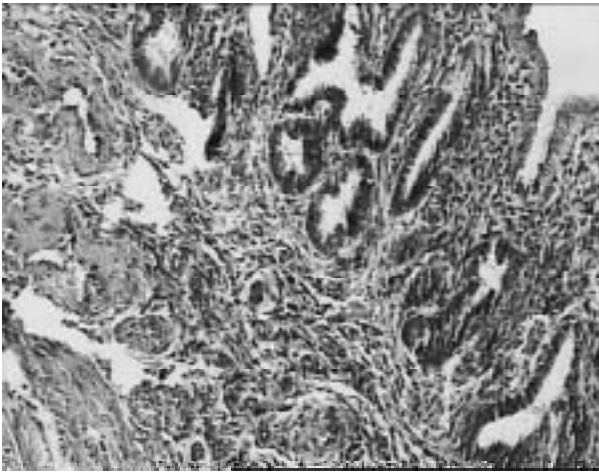


Fig 1. Endometrial stroma and glands in ectopic testicle, X 40.

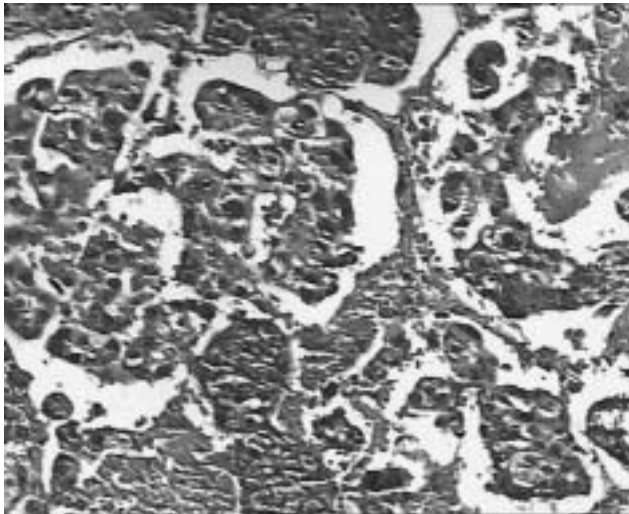


Fig 2. Biopsy of abdominal mass showing choriocarcinoma cells with necrosis, X 400.

cord. Histology revealed fibro muscular tissue with edema, dilated vessels and hemorrhage consistent with endometrial tissue (Fig. 1). After 10 days he underwent a second surgery and biopsy of the abdominal mass. 6-biopsy specimens confirmed hemorrhagic and necrotic mass with cytotrophoblasts and syncytiotrophoblasts with hyperchromatic nuclei and atypia, all charac-

teristic of choriocarcinoma (Fig. 2). Following surgery he underwent chemotherapy. He was under control for 8 months he was admitted again for lung metastasis and paraaortic lymphadenopathy. A second course of chemotherapy was planned; we do not have further follow up of this patient.

Discussion

Gynecomastia is the most common manifestation of estrogen excess in men.¹⁻³ Tenderness may be present in a third of patients. Enlargement is usually central and symmetric, although occasionally it is eccentric. Idiopathic and drug-induced gynecomastia is usually unilateral; however, in pubertal and hormonal cases, the changes are often bilateral.⁴ Pathologic gynecomastia and decreased libido may be due to testosterone deficiency, increased estrogen production, or increased conversion of androgens to estrogens. The pathological conditions associated with gynecomastia include congenital anorchia, Klinefelter syndrome, testicular feminization, hermaphroditism, adrenal carcinoma, liver disorders, and malnutrition and choriocarcinoma.^{1,4,5} Our patient had persistent mullerian duct syndrome which is secondary to anti-mullerian hormone deficiency.⁶ This patient was phenotypically male, fertile, and presented with gynecomastia; he was found to have an ectopic testis that contained uterine tissue. He also had an abdominal mass, found to be choriocarcinoma originating from the ectopic testicle. In this patient, gynecomastia is secondary to increased β -HCG and estrogen that is possibly due to conversion from testosterone.^{5,7}

In patients with unrecognised intra-abdominal testis, nonseminomatous germ-cell tumors are less common than pure seminomas.^{8,9} 20 cases of testicular neoplasia with PMDS have been reported; . About 10% of testicular tumours arise from an undescended testis,⁹ and the relative risk of testicular cancer in cryptorchidism is 7.5%.¹⁰ The location of the undescended testis also affects the risk

of developing a tumor, in that the higher the position of the undescended testis, the greater the risk of developing a malignant tumor.¹¹ The overall incidence of malignant transformation in the gonads of patients with PMDS

is 18%, which is similar to the rate in abdominal testes in otherwise healthy males.¹² We suggest that any case of gynecomastia be evaluated for secondary causes and congenital malformations.

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