Polyorchidism
A Case Report and Review of Literature

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Keywords: testis, urogenital abnormalities, ultrasonography, magnetic resonance imaging

Urol J (Tehran). 2006;3:180-3.
www.uj.unrc.ir

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Received April 2006
Accepted June 2006

INTRODUCTION

Polyorchidism is an extremely rare congenital anomaly of the urogenital system and refers to the presence of more than two testicles. About 100 cases of this disease have been reported in the English literature, so far.[1] Its most common presentation is triorchidism.[2] The left side is predominantly affected. Approximately 50% of the cases are detected between 15 and 25 years of age.[3] The majority of patients are asymptomatic or present with painless inguinal or scrotal masses, undescended testis, and rarely, torsion of the supernumerary testis.[4] We report triorchidism in a 24-year-old man.

CASE REPORT

A 24-year-old man presented with a dull pain in the left hemiscrotum. He did not have any previous history of urogenital complaints. Physical examination of the left scrotum revealed 2 ovoid, nontender, soft, mobile lumps which were homogeneous in consistency. The right testis was normal in palpation. No lymphadenopathy was detected. Laboratory tests including the serum levels of α-fetoprotein, β-human chorionic gonadotropin, follicle-stimulating hormone, luteinizing hormone, and testosterone were within the reference range. Semen analysis showed no abnormality.

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Ultrasonography revealed the presence of 2 testes within the left hemiscrotum with complete septation, a similar echotexture, and a vascular flow pattern similar to the vascular flow of the normal right testis (Figure 1). A mild hydrocele was also noted on the right side. The right testis measured approximately 4 × 3 × 2 cm, and the 2 soft-tissue structures of the left side measured 2.5 × 2 × 2 cm and 2 × 2 × 1.5 cm. There was no focal abnormal echogenicity suggesting malignancy. Scrotal MRI confirmed 2 soft-tissue structures in the left hemiscrotum with normal signal intensity at T1w and T2w images. Both testes had a tunica albuginea with low-signal intensity (Figure 2).

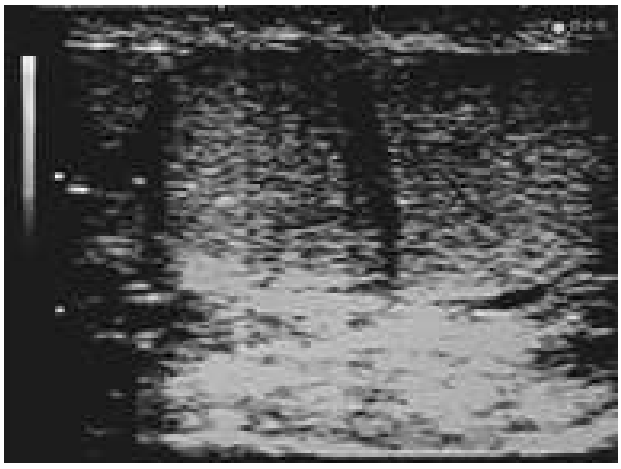

Figure 1. Ultrasonography of the left scrotum showed 2 soft-tissue structures with complete septation and similar echotexture to the normal right testis in the transverse section.

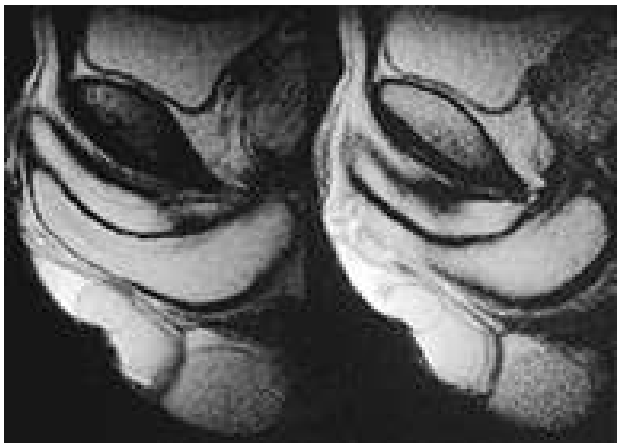

Figure 2. Coronal plane magnetic resonance imaging demonstrated 2 testes in the left hemiscrotum.

The findings were compatible with the diagnosis of testicular duplication. The patient was followed up conservatively.

DISCUSSION

Polyorchidism is a rare congenital anomaly and 100 cases have been reported in the literature, to date.[2] Testicular duplication may be due to the duplication of the genital ridge or longitudinal or transverse division of it before the 8th week of the gestation period.[3,5] A functional classification is reported by Thum based on the embryonic development as follows: type 1, the supernumerary testis does not have epididymis and vas deferens; type 2, the supernumerary and the regular testes are linked to each other with a common epididymis and they both have a common vas deferens (incomplete division); and type 3, the supernumerary testis has its own epididymis but its vas deferens is shared with the regular testis (complete division).[6] Our case falls into the type 3 category of this classification with complete left testicular duplication.

The most common anomalies associated with polyorchidism are inguinal hernia (30%), maldescended testis (15% to 30%), testicular torsion (13%), hydrocele (9%), varicocele (< 1%), hypospadias (< 1%), anomalous urogenital union (< 1%), and malignancy (< 1%).[5]

To date, a few cases of polyorchidism associated with malignancy have been reported in the literature, but there is no evidence supporting the idea of a relation between these two entities.[7] Assessment of the true malignancy potential of polyorchidism is difficult due to its low incidence and frequent association with other risk factors, such as cryptorchidism.[8] The appropriate management of polyorchidism remains unclear. Careful evaluation is necessary in every suspected testicular or scrotal finding to avoid misdiagnosis. However, if there is not any coexistent disorder, testicular tumor markers are negative for malignancy, and tumors can be ruled out by ultrasonography or MRI, surgical exploration or biopsy is not necessary. Consequently, these patients can be followed up conservatively.[7]

Splenogonadal fusion is a rare anomaly due to abnormal connection of the splenic tissue to the gonad or the mesonephric structures during embryonic development. Typically, the malformation manifests as a testicular mass such as polyorchidism. Ultrasonography demonstrates an ovoid structure attached to the testicle which has a homogeneous echogenicity similar to the echogenicity of the testicle. Thus, the differentiation of splenogonadal fusion and polyorchisism cannot be made solely on the basis of ultrasonographic findings. Some authors have described the use of technetium Tc 99m sulfur colloid scintigraphy to identify the areas of ectopic splenic activity, and thereby, adding information to the imaging diagnosis.[9] Magnetic resonance imaging also helps distinguish between the testicular and extratesticular pathologic processes and determine the solid and cystic lesions.[10]

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EDITORIAL COMMENT

We read, with great interest, the article by Dr Kharrazi and colleagues. Polyorchidism is an unusual abnormality of the genital tract in which supernumerary testicles are present, usually within the scrotum. It can be located in the scrotum, inguinal region, or in the abdomen. This anomaly is usually associated with inguinal hernia, testicular torsion, hydrocele, cryptorchidism, and testicular cancer.

The exact embryological explanation of this anomaly is not fully understood in spite of several theories proposed to explain the different anatomical types.[1] Three main theories have been proposed. The first suggests initial longitudinal and the second suggests transverse division of the genital ridge, either through some local accident or by development of the peritoneal bands. The result of longitudinal division may be a complete duplication of the vas, epididymis, and testis, as it is present on the both sides in this case. The more common form of polyorchidism is associated with transverse division in the genital ridge resulting in the duplication of the testis with a single epididymis, vas deferens, and tunica albuginea.[1,2] Another hypothesis of 2-fold primordial glands on either side is supported by 8 previously reported cases,[3] where there are 2 testes with 2 separate epididymides and their vas deferens.

Polyorchidism is a very rare congenital disorder with fewer than 100 cases reported in medical literature.[3] Only one case has been reported from our country.[3] The most common form is triorchidism or tritestes in which 3 testes are present. The condition is usually asymptomatic, but can increase the risk of testicular cancer.

Leung described the anatomical variations on the possible embryological basis. Polyorchidism occurs in several forms[4]:

Type 1: the supernumerary testis lacks an epididymis and vas deferens and has no connection to the other testes.

Type 2: the supernumerary testis shares the epididymis and the vas deferens of the other testes.

Type 3: the supernumerary testis has its own epididymis and shares a vas deferens.

Type 4: complete duplication of the testis, epididymis, and vas deferens is seen.

Type 2 is the most common form of polyorchidism, and types 2 and 3 together account for more than 90% of the cases. Except in type 1, the supernumerary testis is usually reproductively functional. The supernumerary testis is most often found in the left scrotal sac. Similarly, Singer and associates suggested an anatomical as well as functional classification of polyorchidism.[5]

Type 1: supernumerary testes attached to the draining epididymis and vas deferens with reproductive potential (Leung, type 2, 3, and 4).

Type 2: testes with lack of such an attachment without having any reproductive potential (Leung, type 1). Each of these 2 types are again subdivided into 2 groups (A and B) depending on their location in the scrotum (orthotopic) or outside the scrotal sac (ectopic), respectively. Combining this classification with knowledge of potential complications, they proposed a management strategy.

Polyorchidism is generally diagnosed via an ultrasonographic examination of the testes. However, the advent of ultrahigh-frequency probes with vastly expanded dynamic ranges makes monographic evaluation a safe, inexpensive, and highly accurate modality to accomplish the diagnosis. Color flow and Power Doppler can help accurately diagnose abnormalities of the vascular supply in most cases; ie, torsion. But other scrotal pathologies must be considered as a differential diagnosis of this entity. In all of the reported cases (except for few with ultrasonographic diagnosis but not surgical confirmation) the final diagnoses were confirmed by surgical exploration. Polyorchidism was suspected by ultrasonography, but follow-up ultrasonographic studies showed a decrease in the echogenicity of the scrotal structure. Surgical exploration revealed the testis and epididymis to be completely separated, with no duplicated testis.[6]

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