Polyorchidism in pediatric patients: a case report and a literature review

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ABSTRACT

Polyorchidism or testicular duplication is defined as the histologically confirmed presence of more than two testes.

This is the case of a full left testicular duplication in an 11-year-old patient presenting with occasional pain in the left hemiscrotum. At physical exploration, a palpable scrotal mass was detected. An ultrasonography was performed, which revealed the presence of testicular duplicity. Diagnostic suspicion was confirmed by means of a nuclear MRI and a histopathological study following excision.

Polyorchidism is a rare abnormality requiring a high degree of suspicion in the presence of an extratesticular mass. Most authors advocate conservative treatment and follow-up in the absence of pain, cryptorchidism, tumors, and unclear diagnosis.

KEY WORDS: Polyorchidism; Testicular duplication; Supernumerary testis; Childhood.

POLIORQUIDISMO EN LA EDAD PEDIÁTRICA: A PROPÓSITO DE UN CASO Y REVISIÓN DE LA LITERATURA

RESUMEN

El poliorquidismo o duplicación testicular se define como la presencia de más de dos testículos confirmados mediante histología.

Se presenta un caso de duplicación testicular izquierda completa en un niño de 11 años de edad que consultó por dolor ocasional en el hemiescroto izquierdo. En su exploración física destacaba una masa escrotal palpable. Se practicó ecografía que reveló la presencia de una duplicidad testicular. La sospecha diagnóstica fue corroborada mediante resonancia magnética nuclear y el estudio histopatológico tras la exéresis, confirmó el diagnóstico.

El poliorquidismo es una anomalía excepcional que require un alto índice de sospecha ante la presencia de una masa extratesticular. La mayoría de autores abogan por un tratamiento conservador y seguimiento si no causa dolor o si no existen dudas diagnósticas, criptorquidia o excepcionalmente, tumores.

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PALABRAS CLAVE: Poliorquia; Duplicación testicular; Testículo supernumerario; Infancia.

INTRODUCTION

Polyorchidism, testicular duplication, or supernumerary testis is defined as the histologically confirmed presence of two or more testes, either inside or outside the scrotum. It is an extremely rare congenital abnormality. Up to date, no specific chromosomal abnormalities have been found to be involved in it[1].

It is typically asymptomatic, with clinical diagnosis being achieved as a result of inguinal pain or the presence of an inguinoscrotal mass during childhood or adulthood. More than half of the cases are diagnosed at 15-25 years of age[2].

Differential diagnosis primarily with spermatocele, hydrocele, varicocele, epididymal cyst, aberrant epididymis, and testicular tumor should be carried out.

Ultrasonography and nuclear MRI (NMRI) are the diagnostic tools of choice. In simple testicular duplicity cases, diagnosis may be achieved by means of ultrasonography, but in case of doubt or in the presence of cryptorchidism or tumor mass, NMRI is much more sensitive[3].

Definitive confirmation of polyorchidism or testicular duplicity is achieved by means of a histopathological study.

CLINICAL CASE

This is the case of an 11-year-old male patient with history of bilateral inguinal hernia surgical repair in the first years of life. He presented with occasional discomfort in the left hemiscrotum, which he had been suffering from for months. At physical exploration, a left paratesticular mass was palpated.

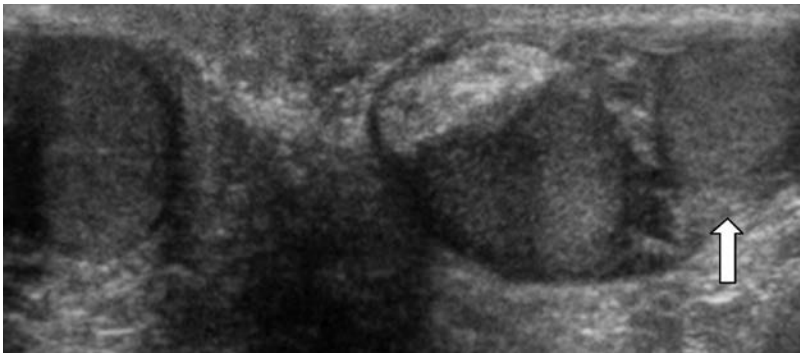

Figure 1. Ultrasound image. At the level of the left scrotum, a 10x8 mm diameter image is visualized. It is smaller than the left testis, but it has the same echogenicity, with an independent but similar vascularization.

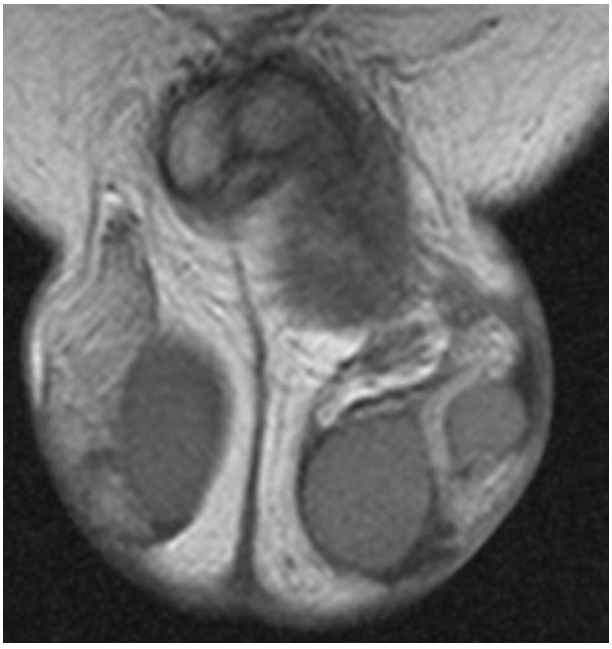

Figure 2. NMR image. In the left scrotal sac, an approximately 12x12 mm extratesticular mass can be seen lateral to the left testis. This could be a testicular duplication.

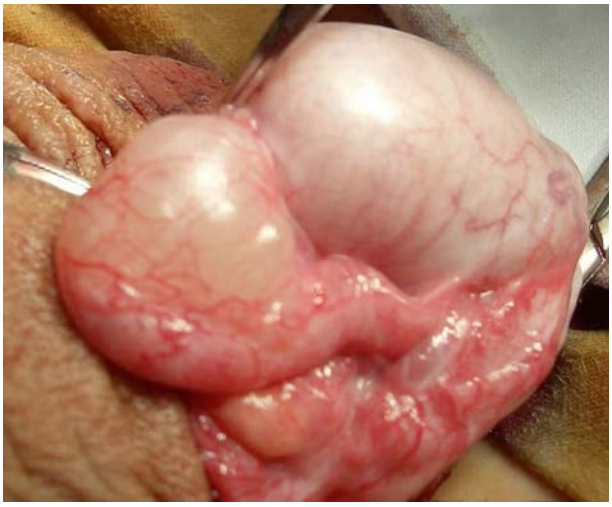

Figure 3. Surgical picture. Two testes are found in the left hemiscrotum, with completely independent vascularization, epididymis, and sperm cords.

A Doppler ultrasound examination of the scrotum was performed (Fig. 1), which showed an approximately 10x8 mm circular image at the external, inferior pole of the scrotum, with the same echogenicity as the testis, and an adequate and similar vascularization. Given the suspicion of testicular duplicity and considering how rare this entity is, decision was made to conduct a NMRI for diagnostic confirmation.

The NMRI (Fig. 2) demonstrated a normal left testis in terms of size and morphology considering the patient's age, and another adjacent 12x12 mm vascularized mass lateral to the testis, with very similar characteristics.

Blood tests and tumor marker studies were normal.

Given the suspicion of testicular duplicity – in spite of it being rare – and the patient's discomfort, upper left hemiscrototomy was decided upon (Fig. 3). The accessory testis was removed. It was macroscopically normal, smaller than the left testis, and had its own vascularization, epididymis, and ductus deferens – completely independent from the left testis structures. The postoperative period was uneventful.

The histopathological study (Fig. 4) allowed diagnosis of polyorchidism or full testicular duplicity to be confirmed. It demonstrated encapsulated testicular parenchyma with absence of spermatogenesis and presence of Sertoli cells only.

Today, the patient is 18, and he has been discharged from our department. He has had no symptoms whatsoever since he underwent surgery (7-year follow-up).

DISCUSSION

Polyorchidism or testicular duplicity is an extremely rare medical entity, with about 160 cases published in the literature[4].

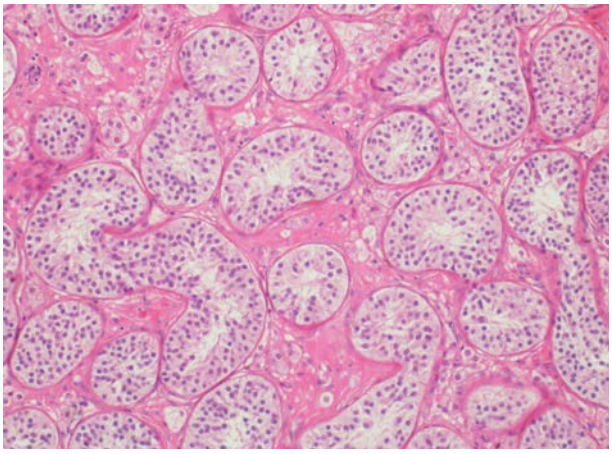

Figure 4. Histological finding. Seminiferous tubules of a normal architecture and size made up of Sertoli cells only, with no evidence of spermatogenesis. The intertubular space has a normal level of Leydig cells.

The first demonstration of polyorchidism was a post-mortem finding by Blasius in 1670. In 1880, Ahfeld[5] also described it at an autopsy. But the first case histologically confirmed during surgery was reported by Lane in 1895[6].

The embryological explanation of polyorchidism remains unknown.

The embryological development of the testis starts at pregnancy week 6, when the primitive testis is developed in the medial part of the primitive genital ridge, within the mesonephros. Polyorchidism could occur due to a longitudinal or transversal division of the genital ridge – the latter being the most widely accepted hypothesis –, or as a result of the degeneration of some parts of the mesonephric components[8].

Polyorchidism may be either unilateral or bilateral, and it can range from partial to full duplication, with completely independent testes and epididymides. Leung[7], Hancock[8] and Wolf's[9] classification, which is based on the anatomy of the testes and the adjacent structures, is the most widely accepted today.

It divides polyorchidism into 4 types:

- Type 1. The supernumerary testis has no epididymis or ductus deferens, without connection with the other testis.
- Type 2. The two testes share the same epididymis and ductus deferens.
- Type 3. The supernumerary testis has its own epididymis and shares the ductus deferens.
- Type 4. Full testicular duplication, with the testis having its own epididymis and ductus deferens.

Types 2 and 3 are the most frequent anatomical forms of polyorchidism, accounting for approximately 90% of the cases. The patient in this article had a type IV polyorchidism – the least frequent type – according to Leung's classification.

Supernumerary testes are typically functional reproductively speaking[10], except for type 1 testes, which have no epididymis or ductus deferens. Therefore, even though there are some cases associated with infertility[21], this is to be considered in polyorchidism patients undergoing bilateral vasectomy, since otherwise the latter may fail.

Polyorchidism is more frequent on the left side, and it can be associated with other urogenital abnormalities. In approximately half of the cases, testicular duplicity is associated with undescended testis or cryptorchidism[11]. In up to 30% of the cases, it is associated with indirect inguinal hernia[12,23]. The remaining cases are associated with torsion, hydrocele, epididymitis, varicocele, and even malign degeneration, the latter being frequent in poor testicular descent cases. Indeed, supernumerary testis can be associated with teratoma[13,14], anaplastic seminoma[15,16], choriocarcinoma[17], and rhabdomyosarcoma[18]. The incidence of torsion in these testes is approximately 13%[19].

Differential diagnosis should include other causes of extratesticular or paratesticular masses (hydrocele, epididymal cyst, tumors, etc.)[25].

Given how rare it is, it requires a high degree of suspicion. Surgical management has been an issue of discussion in various publications in the literature[22]. Today, in the absence of pain, unclear imaging diagnosis requiring biopsy, associated pathology – such as cryptorchidism –, and tumors[20], conservative management with a strict ultrasound and NMRI follow-up[24] is the most widely used approach.

If diagnosis is achieved through biopsy, and the supernumerary testis has a normal position within the scrotum, the characteristics of the spermatic cord should be studied, and orchidopexy should be carried out to minimize the risk of torsion or ascent to the inguinal canal[22]. However, if it is a cryptorchid testis, two options are available – orchidopexy and subsequent monitoring, or excision to prevent malignization risks.

Anyhow, in our view, polyorchidism treatment and follow-up should be a consensus decision that should take into account the patient's will and/or that of their parents in the case of underage patients.

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