Polyorchidism. Atypical Presentations of Supernumerary Testicle Can Lead to Misdiagnosis

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Abstract

Polyorchidism is a rare urogenital curiosity defined as the presence of a supernumerary testis. We, hereby, report a 20-month old male who presented with atrophic testis in the left hemiscrotum after previous orchiopexy due to cryptorchidism and the presence of an inguinal mass at the deep inguinal ring resulted to be an additional testicle. We review the literature and propose its management.

Introduction

Polyorchidism is a rare genital abnormality of the male genitalia which refers to present an extra testicle; the commonest type of polyorchidism is triorchidism, or tritestes, where 3 testes are present. Until now about two hundred cases have been reported in the literature [1]. The majority of polyorchidism cases are determined coincidentally while researching for the causes of other symptoms. It is associated with testicular malignancy, cryptorchidism, inguinal hernia, varicocele and torsion [2,3]. We describe one case of triorchidism and cryptorchidism in a 20-month old male while performing a surgical exploration of the left inguinoscrotal region due to inguinal mass that appear during a routine pediatric ultrasound examination. We review the literature and propose its management.

Keywords: Polyorchidism; Triorchidism; Cryptorchidism; Supernumerary Testicle; A Congenital Testicular Anomaly

Case

A 20-month old boy was referred to us because of the atrophic testis in the left hemiscrotum after previous orchiopexy due to cryptorchidism and the presence of an inguinal mass at the deep inguinal ring that had appeared during a routine pediatric ultrasound examination ten months after orchiopexy. Physical examination revealed atrophic left testis and normal right testis. There is no significant prior medical illness or history of trauma was given. Surgical exploration of the left inguinoscrotal region revealed the atrophic testis in the scrotum and one more distinct small testicle with common vessels but with absent vas deferens. The testis in the scrotum was excised because of its atrophic nature. Likewise, the cranial testis was excised due to the intraabdominal position just behind the inguinal ring, the smaller size and the absence of the vas deferens. Histological examination of the removed testis showed elements of atrophic testicular tissue and the histological examination of the surgical specimen showed an involuted testicle.

Discussion

Polyorchidism is a rare congenital anomaly defined by the presence of more than two histologically proven testes ipsilaterally. Triorchidism is the most common presentation. The earliest reported findings of this malformation came from autopsies in 1660 by Blassius [4]. With recent improvements in imaging studies, an increasing number of cases are diagnosed via US or MRI. About 200 cases of polyorchidism have been found in a recent review of the literature [5-7]. It is usually diagnosed as an incidental finding in children and young males during surgical procedures of associated pathology such as in our case. Several explanations have been reported for the embryogenesis of this anomaly, including anomalous appropriation of cells, longitudinal duplication or transverse division of the genital ridge, degeneration of parts of the mesonephric duct, local vascular accident or development of peritoneal bands [8-12]. The most common form of polyorchidism is associated with a transverse division in the genital ridge, resulting in duplication of the testis with a single epididymis and vas deferens. There are various classifications based on embryologic development, anatomical and...
functional arrangements of the testes and their outflow paths. A functional classification system is published by Leung based on embryologic development: type 1 supernumerary testis without epididymis and vas deference; type 2 supernumerary testis shared common epididymis and vas deference with ipsilateral testicle; type 3 supernumerary testis has its own epididymis but shares a common vas deferens with the ipsilateral testicle; type 4 there is complete duplication of testis, epididymis and vas [13]. However, according to our opinion, there is another classification suggested by Bergholz et al., based on anatomy that is more useful and more appropriate to use. A testis being drained by an outflow path (vas deferens) was coded as type A, whereas undrained testes without connection to a draining vas deferens were coded as type B. Type A testes were further divided into 4 subgroups. A1: Supernumerary testis has its own epididymis and vas deferens, A2: Supernumerary testis has its own epididymis but common vas deferens with neighboring testis, A3: Supernumerary testis has its common epididymis and vas deferens with neighboring testis, and A4: Supernumerary testis has its own vas deferens but common epididymis with neighboring testis. Type B testes were further divided into 2 subgroups. B1: Supernumerary testis has its own epididymis and B2: Supernumerary testis lacks an epididymis, thus consisting of testicular tissue only [14]. Our case falls into the type 1 category of functional classification and using the second classification, our case was type B2.

Among the reported cases the most common type of polyorchidism was A3 (22%), followed by A1 (17%), and A2 (14%). Undrained testes (type B) counted for 18% of all described cases, of which 10% were type B1 and 8% B2. [15]. In most reports as well as in our case there is preponderance for the left side, similar to the laterality in testicular torsion and regression [9,10]. About 75% of supernumerary testes are intrascrotal, with another 20% located in the inguinal canal and 5% in the retroperitoneal space [16,17]. The most common anomalies associated with polyorchidism are inguinal hernia (30%), maledescended testis (15% to 30%), testicular torsion (13%), hydrocele (9%), varicocele (< 1%), hypospadias (< 1%), anomalous urogenital union (< 1%), and malignancy (< 1%) [18,6,9-11].

It is a known fact that there is an increase in testicular malignancy in patients with cryptorchidism as compared to the normal male population. In addition cryptorchidism appears to be the most important risk factor for malignancy in patients with supernumerary testes [14]. The estimated increased risk of malignancy in supernumerary is about 6%, and the commonly encountered histological types include seminoma, teratoma, embryonal and choriocarcinoma [19,4,20-22]. Thus, patients with nontesticular supernumerary testes require appropriate counseling.

There are insufficient data in the literature on the ideal management of polyorchidism. Careful diagnosis is necessary in every suspicious scrotal/inguinal finding to avoid misdiagnosis. When performing surgical treatment, meticulous intra-abdominal and intrainguinal exploration must be undertaken. Some authors have stated that the diagnosis of polyorchidism can be made with sonography alone and that surgical exploration is unnecessary to confirm the presence of an accessory testis [23]. Others suggest an orchidectomy of the duplicated testis to decrease the risk of malignancy [24]. However, the association of polyorchidism with proved risk factors, such as cryptorchidism, makes it difficult to estimate its true malignant potential. The decision of the surgeon depends on several factors, such as the position of the supernumerary testis (ectopic, inguinal or abdominal), the reproductive potential, its size and volume (normal or atrophic), the age of the patient, and psychological aspects that are related to follow-up (ie, anxiety of the patient) [25]. This surgical procedure was routinely performed in the eighties.

Nowadays, there is a tendency to perform a biopsy and orchiopexy to prevent torsion as well as to preserve the endocrine function, but this type of management requires regular ultrasound checkups in order to control the development of a possible malignancy [6,9,10,26-28]. 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 [29].

The advantages of using sonography for the investigation of polyorchidism are that it provides high-resolution images that permit differentiation of a normal testis from a tumor mass with ease, it is flexible, convenient, requires only a few minutes of scanning time, and it involves no radiation exposure and is thus suitable for repeated use in scrotal examinations. MRI is the most sensitive diagnostic imaging technique in the classification of intrascrotal pathology but, it cannot be considered a routine examination in pediatrics because of the absolute need of deep sedation or general anesthesia [23,30].

We strongly believe that exploratory surgery should be performed in all symptomatic cases. The excision of supernumerary testes is considered necessary when: a) the testis is atrophic, dysplastic or has no reproductive capacity, b) there is a suspicion of malignancy by means of serological tests, histological studies, ultrasound or MRI scan c) placing of the testis in the scrotum is impossible, d) in cases where regular follow-ups are not reliable and e) parental request arises.
References


