Polyorchidism: A Case Report and Review of the Literature

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Introduction

Polyorchidism is a rare congenital anomaly defined as the presence of supernumerary testes with the most common presentation with two homolateral and one contralateral testes. Since the first case reported in 1880; more than 100 cases were published in the literature [1]. Here we report a 37 year old case with left supernumerary testes presented with painless mass and had a medical history of infertility.

Case

A 37 year old man presented with a complaint of painless lump in his left scrotum. He had a medical history of infertility and after 10 years of marriage, they had twins with assisted reproductive methods and had no diagnosis for the left scrotal mass. On physical examination, a normal right and left testis and a left soft mass approximately the same size as the left testis and separated two vasa deferentia were palpated. On laboratory tests, two separate spermiogram showed oligoasthenoteratospermia and normal serum levels of alpha fetoprotein, beta human chorionic gonadotropin and lactate dehydrogenase were detected. Ultrasound (USG) examination demonstrated a normal left testis measuring 44 × 34 × 21 mm and supernumerary testis measuring 37 × 25 × 22 mm with identical echogenity. The right testis was normal measuring 44 × 34 × 21 mm. All three glands had separate epididymis. Magnetic Resonance Imaging (MRI) confirmed the presence of a supernumerary testis in the left hemiscrotum. All glands had identical signal characteristics on T2 weighted images [Figure 1a-b]. No therapeutic approach was performed, but sonographic follow-up (i.e. every 6 months) was suggested for the patient because of the risk of malignancy.

Discussion

Polyorchidism is a rare congenital urogenital anomaly causing supernumerary testicles. The etiology of polyorchidism is still unclear. At about the sixth week of embryological life the primordial testis begins to develop from the medial aspect of the primitive genital ridge, while the epididymis and vas deferens arise from the wolffian duct [2]. Several theories about the etiology of polyorchidism exists. Theories include, duplication or transverse vs longitudinal division of the urogenital ridge, incomplete degeneration of a portion of the mesonephros, anomalous appropriation of cells and development of Peritoneal foldings. The most accepted theory explains that anomalous division of the embryonal genital ridge was the cause with or without the mesonephros, before the 8th week of gestation either through local accident or development of peritoneal bands [3]. Depending upon the level of the division and the segmentation plane, the supernumerary testis may or may not remain in communication with the epididymis and vas deferens [4].

Leung classified polyorchidism on the basis of embryologic development [5]. In type I, the division separates a small part of the genital ridge which does not contact the mesonephric duct and as a result the supernumerary testis lacks an epididymis and vas deferens. In type II the division of the genital ridge occurs in the region where the primordial gonads are attached to the mesonephric ducts and the supernumerary testis has its own epididymis. In type III, the supernumerary testis has its own epididymis and shares the vas deferens with the regular testis in a parallel fashion. In this type of polyorchidism, there is an incomplete longitudinal division of the genital ridge and the proximal portion of the mesonephric duct. In type IV, which is the least common, complete longitudinal duplication of the genital ridge and mesonephric duct occurs, with resultant complete

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duplication of testes, epididymides and vas deferens. Type II is the most common type and with type III compromises 90% of the cases. Singer et al. [6], proposed a classification based upon anatomical and functional potential of the supernumerary testis separating the polyorchidism in type 1 and 2. Type 1 is an accessory testes attached to the draining epididymis and vas deferens with reproductive potential (Leung type II, III, IV). Type 2 is an accessory testes with no reproductive potential with lack of attachment to an epididymis and vas (Leung type I). Both types subdivide into two groups according to their location in the scrotum or being outside the scrotal sac.

Diagnosis of polyorchidism is mostly incidental. Painless groin or scrotal mass is the presenting symptom in 16% of the cases in the literature [7]. Though polyorchidism has no specific clinical presentation, should always be kept in mind when evaluating a scrotal or inguinal mass.

Systematic analysis of the literature on polyorchidism revealed data for 140 cases with histological confirmation [7]. The median age was 17 years and most patients were young adults with age range of 11 (25th percentile) to 25 (75th percentile) years. In most cases, a single and generally left-sided supernumerary testis was present as in our case [7,8]. It has been hypothesized that the left testis may be more prone to subdivision because of its greater size and different vascular topographic anatomy compared to the right testis [9]. The majority of supernumerary testes were found in the scrotal region (66%), followed by inguinal (23%) and abdominal (9%) positions [7]. This congenital anomaly typically causes no impairments, but it is randomly associated with inguinal hernia (24%) cryptorchidism (22%) and testicular torsion (15%) [7]. Reports also show an increased risk of testicular malignancy in the presence of polyorchidism [10,11]. Of these neoplasms 88.8% were malignant [7]. Wolf et al. [2], found approximately a 5% incidence of malignancy including seminoma, teratoma, and chorionicarcinoma. In a recent review of the literature Bergholz et al. [7], found 6.4% neoplasm of the supernumerary testes consisting 140 cases.

Infertility (20%) is a common finding in polyorchidism; 37% of these patient have tubular atrophy, sertoli cell pattern (without leydig cells) or lack spermatogenesis [12]. Although scrotal lump was our case's presenting symptom, he had a medical history of infertility and had children with assisted reproductive methods and still have abnormal spermogram.

The sonographic appearance of polyorchidism is a scrotal mass that has an echo pattern identical to that of the ipsilateral testicle [13]. Colour Doppler sonography shows flow characteristics similar to those of the ipsilateral testis. On MRI, a round or oval shaped structure showing typical signal characteristics of testicles, with homogeneous intermediate signal intensity on T1 weighted and high signal intensity on T2 weighted images was found [14]. With the advent of diagnostic procedures like ultrasound and MRI, conservative approaches for the management of polyorchidism were advocated [15]. In the literature, the current treatment of an uncomplicated polyorchidism is conservative with close sonographic observation in the absence of complicating conditions such as cryptorchidism, torsion or malignancy [16].

Conclusion

Polyorchidism is a rare congenital urogenital anomaly with unclear etiology. Diagnosis based on ultrasonographic findings and MRI may provide an additional information. The treatment of choice is conservative management with sonographic follow up for uncomplicated cases.

References

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