CASE REPORT

TRES-ticles: A Rare Case of a Discontinous Type of Splenogonadal Fusion Presenting as Polyorchidism

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Polyorchidism is a very rare congenital anomaly, which is defined as having more than 2 testicles in a human male. To date, there are only a number of cases reported worldwide and most of them are published as case reports. Most cases of polyorchidism are associated with other male genitourinary anomalies such as varicoceles, hydroceles, inguinal hernias, and testicular malignancies. Even much more rarer are accessory spleens located intrascrotally, with only a handful of case reports in the last 80 years. Reported here is a case of a 33-year old male presenting with symptoms of intermittent scrotal pain for 2 years and extratesticular mass on ultrasound. Surgical management for this patient was done, removing one of the accessory testicles. Histopathology showed as an accessory spleen. Since this is a rare occurrence, the authors aimed to present splenunculus as a differential for patients with polyorchidism.

Key words: Polyorchidism, splenunculi, splenogonadal fusion

Introduction

An splenunculus, or accessory spleen, refers to the normal splenic tissue ectopic to its normal location, separate from the rest of the organ. As an organ of both hematopoietic and immunologic function, it forms from multiple smaller components as a condensation of the mesodermal mesenchyme, which lands normally on the left side of the dorsal mesogastrium. Failure of this separation can lead to one or more nodules being ectopic. Having the same embryologic origin with the testis, a splenunculus can result from an abnormal connection of ectopic splenic tissue, with other mesodermal structures, particularly with the gonadal tissue, termed as splenogonadal fusion, and present as polyorchidism, or supernumerary testicles. Surgical excision to facilitate histopathologic diagnosis is usually done in most cases, with most, if not all patients, report minimal to no complications and issues regarding sexual function and fertility, with reports of aesthetic satisfaction. This report aimed to present a rare case of an adult male with an accessory spleen presenting as polyorchidism, its work up as well as its surgical management.

The Case

Patient is a 33-year old male with a chief complaint of 2 years history of intermittent left scrotal pain. There were no associated fever, dysuria, lower urinary tract symptoms, nor purulent penile discharge noted. Patient also denied any history of trauma. The patient ultrasound revealed a supernumerary testis, with consideration of testicular torsion. However, the

physician opted for conservative management with intravenous antibiotics and pain control, which offered improvement of the symptoms. Further workup was requested, however, he was lost to follow-up. Two weeks prior to consult, the patient noted recurrence of left scrotal pain and swelling. Patient was admitted with spontaneous resolution of the swelling and pain. Patient opted discharge against medical advice due to financial constraints, and was advised outpatient urology consult hence referral to this institution.

On physical examination, the genitalia was grossly male, with 2 palpable testes in the left hemiscrotum, and 1 palpable testis in the right hemiscrotum. All testes were approximately 3 cm in their widest individual diameter (Figure 1). There was no enlargement of the inguinoscrotal area on Valsalva. The bilateral inguinal areas were unremarkable.

Pertinent findings on the pre-operative ultrasound revealed an accessory testis measuring 2.9cm x 2.9cm x 2.4cm in the left hemiscrotum with increased vascularity, with bilateral reactive hydrocele (Figure 2.1).. Pre-operative MRI of the lower abdomen revealed a fat-containing hernia in the right inguinal region extending inferiorly to the scrotal sac, displacing the right testicle. Native testicles are normal in size and intensity, however, an accessory testis measuring 2.4cm x 2.6cm, was identified in the upper hemiscrotal sac on the left side such testis exhibited a comparatively more rounded and hypo-intense image, also exhibiting mild homogeneous enhancement, with few thin septa noted within, with a separate structure which may represent the epididymis. Bilateral hydrocele was also appreciated. The inguinal lymph nodes were unremarkable. (Figure 2.2). Blood work-ups, urinalysis, and seminalysis were all normal. During this time, the patient was assessed as a case of polyorchidism and was prepared for scrotal exploration.

Intraoperatively, an accessory left testis was noted. It had knotted blood supply, was viable, and normal-sized but no vas deferens. The main left testis was viable, normal-sized with normal-looking spermatic cord.The right testis was intact, normal-sized and viable (Figure 3). A bilateral hydrocele, and an indirect inguinal hernia on the right were noted. Orchiectomy of the left accessory testis with bilateral orchidopexy and right herniorrhaphy were performed. Patient's post-operative course was unremarkable as he was discharged 2 days post-op.

The histopathology report for the left accessory testis revealed the absence of a vas deferens and epididymis (Figure 4.1). On cut section, the specimen revealed areas of extensive hemorrhage (Figure 4.2). Microscopically, it revealed areas containing lymphoid follicles and littoral cells lining sinusoids with discontinuous walls in a background of hemorrhage, similar to a normal histology of a spleen (Figure 4.3). It was signed out as congestion and hemorrhage of accessory spleen.

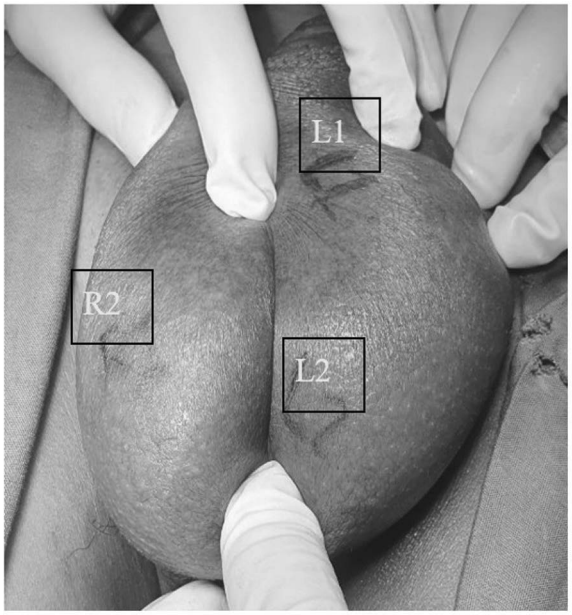

Figure 1. Patient's left testicles, labeled L1 and L2, and patient's right testicle, Labeled R1.

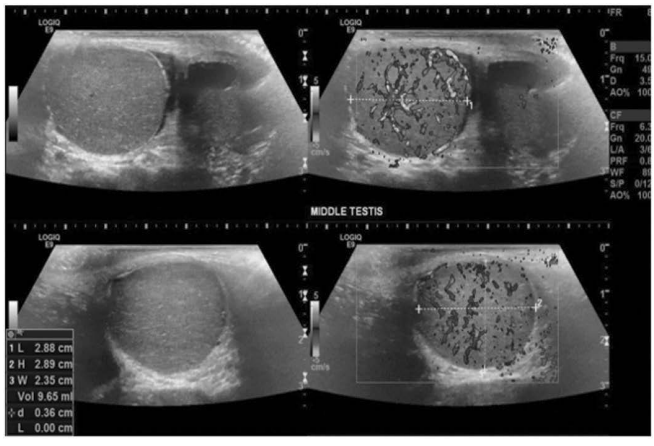

Figure 2.1. Pre-operative ultrasound showing left accessory testis .

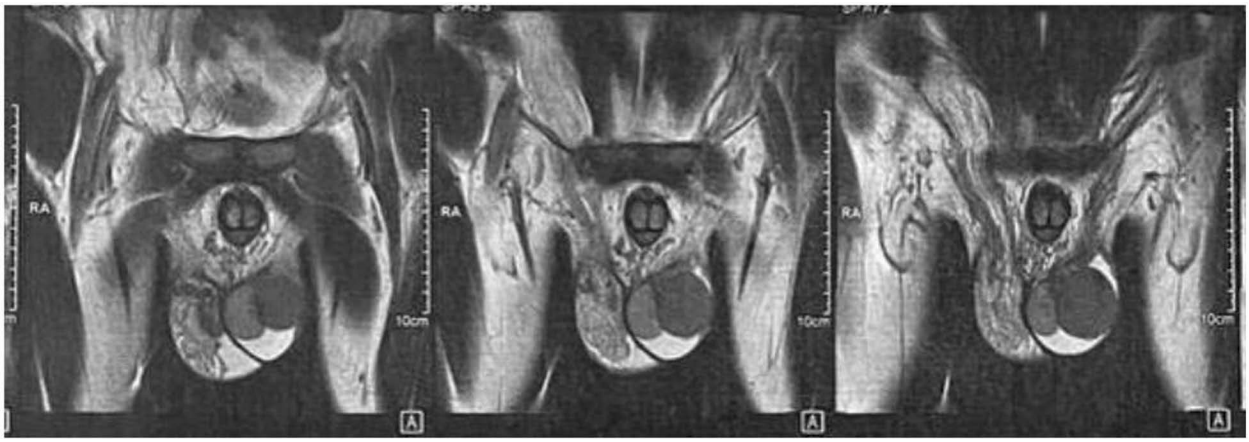

Figure 2.2. Pre-operative MRI showed the accessory testis in the upper hemiscrotal sac with mild homogenous enhancement.

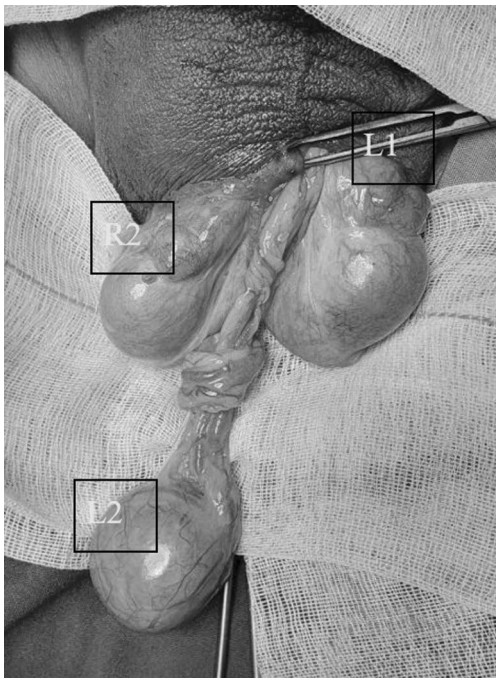

Figure 3. Left main testis (L1); Accessory testis (L2); Right main testis (R1)

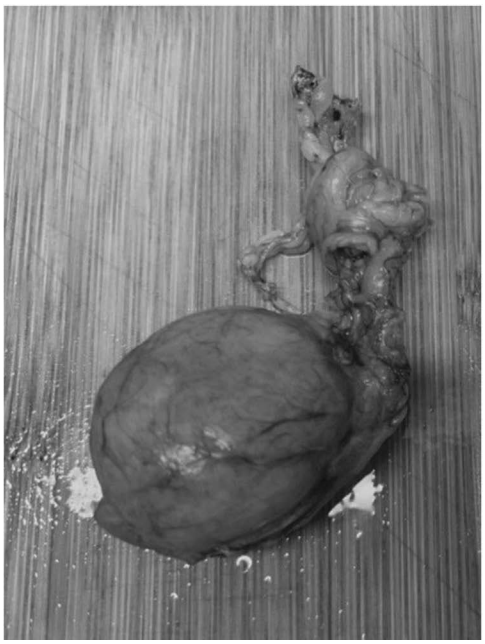

Figure 4.1 Left Accessory testis with absent epididymis and vas deferens.

Figure 4.2. Left Accessory testis, left (cut section) showing areas of extensive hemorrhage

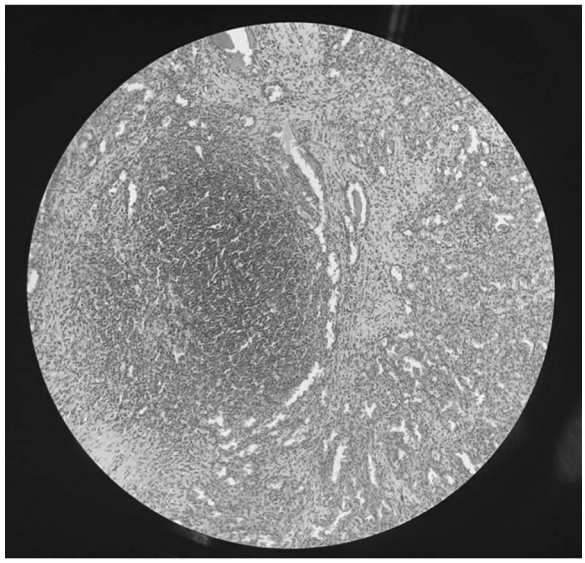

Figure 4.3. Histopathology findings showed areas with lymphoid follicles and littorial cell lining sinusoids.

Currently, the patient is doing well, asymptomatic, no limitations of activities of daily living, sexual function, and is back to work.

Discussion

Polyorchidism is a clinical entity where a person is assessed to have more than 2 testis, with triorchidism being the most common variant. It is a rare congenital anomaly of the genitourinary tract first described by Ahfeld, et al. in 1880. There are only less than 200 cases reported in the medical literature, with no age-specificity among the case reports, over the last 90 years. Normally, the testis begins to form around 6th week age of gestation from the genital ridge, which is a small protruberance on the dorsal coelemic wall. A number of theories have been postulated for the pathogenesis of polyorchidism, most of them are still poorly described due to lack of available data, but anomalous appropriation of cells differentiating into gonads, duplication or transverse versus longitudinal division of the urogenital ridge, or incomplete degeneration of a portion of the mesonephros and development of peritoneal bands to name a few.

As previously mentioned, triorchidism is the most common type, but cases of double polyorchidism have been reported (2 on each scrotum). The most common anatomical variant would be the left hemiscrotum having a supernumerary testicle, as seen in the present case.

The Leung classifications for supernumerary testicles are as follows: Type I lacks epididymis and vas deferens, thereby eliminating all reproductive potential; type II refers to a supernumerary testis which shares the epididymis and vas deferens of the other testis; type III, a testis that has its own epididymis but shares the vas deferens with the other testes and type IV, where there is a complete duplication of the testis, epididymis, and vas deferens. The patient was classified as type I, however, no appreciated spermatic cord was found, and intraoperatively, only supplying blood vessels were found, which raised the suspicion for another clinical entity. Differential diagnoses for polyorchidism include, but are not limited to, encysted hydrocele, spermatocele, a hematocele, or a varicocele, or a testicular or a paratesticular neoplasm. Even more rarely, a diagnosis of splenogonadal fusion or an accessory spleen in the gonad can be made, which was seen in the present case.[2]

Splenogonadal fusion (SGF) is another rare, benign, congenital anomaly where the spleen is abnormally attached to a gonad, which was first described by Bostroem, et al. in 1883. The pathophysiology of SGF begins during the 5th week of embryological development, where the splenic anlage develops in the left dorsal mesogastrium. This splenic progenitor comes into close contact with the left urogenital fold during the rotation of the embryonic gut. The urogenital fold contains the gonadal mesoderm, which matures into the human gonadal structures. The spleen-gonadal interaction continues until gonadal descent at 8th weeks age of gestation. Failure of separation of these primitive tissues result in SGF. There are two types: the continuous and the discontinuous type. The continuous type has a direct anatomical connection between the ectopic and normal spleen, connected by a cord which may be totally splenic in origin, beaded with splenic tissue nodules, or a totally fibrous band. The discontinuous type involves gonadal fusion with an accessory spleen or ectopic splenic tissue. The patient in the present

case has a discontinuous type of splenogonadal fusion. Knowing the embryologic physiology of splenogenesis and gonadal differentiation as far as anatomy is concerned, together with the histopathologic findings consistent with an accessory spleen, the patient can be diagnosed as having splenogonadal fusion.[1,3]

Conclusion

Polyorchidism is considered a rare clinical condition by itself, moreover, the accessory testis being a splenogonadal fusion makes it a rarer clinical entity. With over 200 published cases of polyorchidism noted, the possibility of it being a splenogonadal fusion makes it a differential in managing these types of patients. With this. appropriate awareness should be promoted not only for the urologist as well as the pediatricians who handles these cases initially.

References

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