Management of supernumerary testis: a rare case of polyorchidism in a 2-year-old boy

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Polyorchidism is a rare congenital abnormality described as the presence of three or more testicles. We report the case of a 2-year-old boy with intermittent left-sided scrotal swelling and an ipsilateral undescended testis. During the operation, a left cryptorchid testis was found at the external ring, with further dissection revealing a second vas deferens attached to an additional intrascrotal testis. The cryptorchid testis was pexied in the left hemiscrotum with the second descended testis left in place. A review of the literature reveals over 150 cases of supernumeracy testis. Triorchidism is the most common form with the third testis typically located within the scrotal sac. Supernumeracy testis is frequently associated with undescended testis, testicular torsion, inguinal hernia, and hydrocele. Management is controversial with some advocating for orchidectomy due to concerns of malignancy. The family was educated about the risk for malignancy and the importance of regular follow-up and testicular self-examination starting at puberty. *Ann Pediatr Surg* 13:225–227 © 2017 Annals of Pediatric Surgery.

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Introduction

Polyorchidism is a rare anatomical and congenital malformation defined as the presence of three or more testicles. Thus far, there are over 150 cases reported in the literature, though this number likely underestimates the prevalence given most remain clinically undetected. Triorchidism is the most common variant of polyorchidism [1]. The majority of the cases are discovered incidentally in teenagers during the evaluation and/or surgical management of other inguinoscrotal pathologies, including undescended testes (40%), inguinal hernias (30%), hydrocele/spermatocele (9%), testicular torsion (15%), and testicular neoplasms (6%) [2]. Most polyorchid testes are found within the scrotal sac (68%) but they may also be found within the inguinal canal (23%) or intra-abdominally (9%) [1]. Polyorchidism is associated with a small, but not insignificant risk, for malignant transformation. As a consequence, surgeons must assess the testis' viability and anatomical configuration to decide whether it will contribute to hormonal function and fertility and weigh this benefit against the risk for testicular cancer. Taken together, the management of polyorchidism remains controversial with some advocating definitive orchidectomy, whereas others endorse a close follow-up and self-examination to maximize future fertility. Polyorchidism is rarely encountered in children less than 5 years of age, thus making it worthy of reporting.

Case presentation

An otherwise healthy, 2-year-old boy presented with a 4-month history of a left-sided intermittent, scrotal swelling. His parents reported no concerns of associated pain with the swelling and noted that it often had a "blue tinge" to it. Examination revealed a normal penis with a normal testis in the right hemiscrotum. The left testis was palpated at the level of the external ring and felt to be smaller in size than did the right testis. The left testis could not be brought down to the base of the scrotum. There was no associated swelling at the time. Interestingly, the patient had a history of painless right-sided scrotal swelling consistent with an infantile hydrocele, which resolved spontaneously.

Investigations

A clinical diagnosis of a suspected left-sided hernia with an ipsilateral undescended testis was made and discussed with the parents. No further investigations were conducted.

Differential diagnosis

The differential diagnosis of scrotal masses in this age group includes hernia, hydrocele, spermatocele, epididymal cysts, fibrous pseudotumor epidermoid cyst, adenomatoid tumor, and papillary cystadenoma.

Treatment

An open inguinal approach for the repair of the presumed left-sided hernia and undescended testicle was conducted. At groin exploration, a normal right testicle was again palpated in the right hemiscrotum. The spermatic cord was isolated allowing identification and high ligation of the processus vaginalis. Ongoing dissection of the left inguinal region revealed a left cryptorchid testis at the level of the external ring. Further dissection revealed a smaller additional intrascrotal testis with its own epididymis that shared a common vas deferens with the larger cryptorchid testicle. Both left testes appeared grossly normal, although smaller than the contralateral testis (Figs 1 and 2). A decision was made to preserve both testes to optimize future fertility. The larger cryptorchid testis was pexied in the left hemiscrotum with the second descended testis left in its native position. After the case, the right testis remained palpable in the right hemiscrotum; in the left hemiscrotum, the larger of the two testes was easily palpable, whereas the smaller testis was palpated with some difficulty.

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Outcome and follow-up

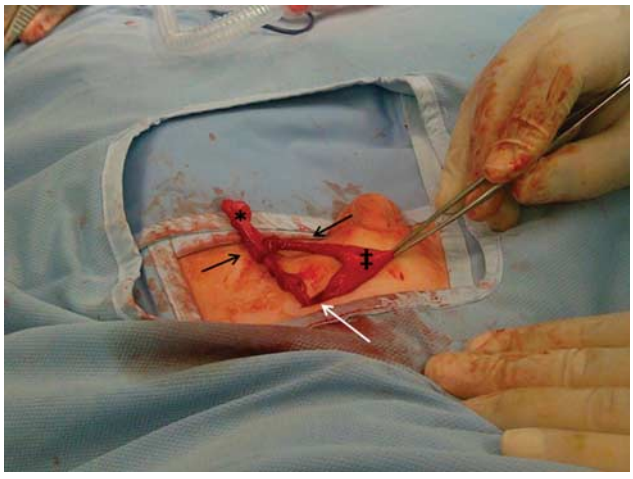
One month after surgery the patient was seen in follow-up clinic and doing well. On examination one of two equally sized testicles were palpable in each hemiscrotum, in addition to the left supernumerary testis that was palpated with some difficulty. The family was further educated regarding the risk for malignancy in cryptorchid and polyorchid testes and the importance of continuous follow-up after puberty.

Discussion

Polyorchidism is a rare congenital anomaly that is often incidentally detected during the evaluation or surgical management for other associated inguinoscrotal pathologies including hernia, hydrocele, undescended testis, epidermoid cyst or testicular torsion. There have been over 150 cases of polyorchidism reported in the literature, with the average age at diagnosis in the late teenage years [1]. Triorchidism has a left-sided predominance with the extra testis generally being smaller than the normal contralateral and ipsilateral testicles, which is concurrent with our patient's presentation. Various permutations of the location of the polyorchid testis are reported in the literature, but by far the most common topology is within the scrotum [1]. The mechanisms of polyorchidism is unknown. Primordial testes begin to form during the sixth week of embryological life from the primitive genital ridge. Some suggest that incomplete degeneration of the mesonephros or aberrant division or duplication of the genital ridge before 8 weeks gestation account for testicular duplications [1].

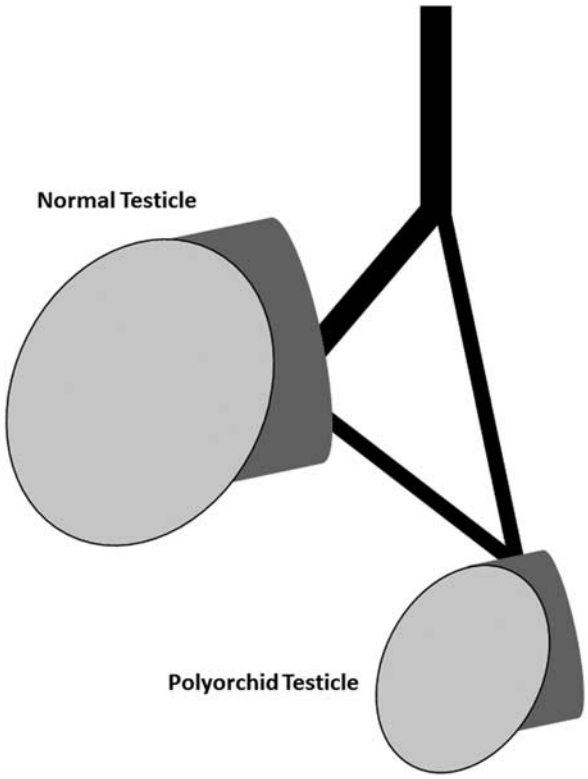
Testicular duplications have been classified anatomically by the connections between the two testes. Bergholz *et al.* [3] has described a comprehensive taxonomy of polyorchidism where type A is drained by a vas deferent and type B is not. On the basis of this classification system our patient was deemed to be an A2; the small distal testicle's vas deferens, and vascular pedicle were a continuation of the vas and vascular pedicle of the proximal testicles, and it had its own epididymis (Figs 1 and 2).

Fig. 1



Intraoperative photograph showing two testicles on the left side. The smaller polyorchid testis (*) and normal testis (‡) with their respective epididymis and vasculature (black arrows) and share a common vas deferen (white arrow).

Fig. 2



Schematic representation of left-sided polyorchid testes discovered intraoperatively.

The management of polyorchidism is still highly debated in the literature. In the past, supernumerary testicles were routinely removed because of the potential for testicular torsion and to eliminate the risk for malignancy. However, there has been a shift in this paradigm with some now advocating for testicular preservation to maximize hormonal function, spermatogenesis, and fertility [1,4]. Patients are monitored for potential complications through follow-up visits and education. One of the most important considerations when deciding the fate of the accessory testis is determining whether it has the potential to contribute to fertility. In the absence of an epididymis and/or vas deferens, the supernumerary testicle does not have an outflow tract to transport sperm and hence will never contribute to reproduction. Thus, removal in this situation is appropriate.

The risk for testicular malignancy in polyorchidism is hard to define given its rarity. Testicular cancer in polyorchidism is reported to have an incidence of 6%, which is 142-fold higher in testicular cancer patients compared with the general population [1]. But as in the case of cryptorchism, education and self-examination are appropriate alternatives to orchiectomy, especially if the testes may support spermatogenesis and hormone production. Cryptorchidism with intra-abdominal location of the supernumerary testis has the most significant risk factor for malignancy [1]. These factors should be considered when deciding the fate of the accessory testicle. The child in our case presented with intermittent left inguinal swelling concerning for hernia and a cryptorchid testes. The diagnosis of a left-sided intrascrotal triorchid testes was made intraoperatively. We advocate for the

preservation of the accessory testicle and performing an orchiopexy when the testis appears grossly normal with its own epididymis and vas deferens. Ensuring the testis is intrascrotal facilitates self-examination to monitor for malignancy once the child reaches puberty. In our patient, the smaller supernumerary testis was located within the left hemiscrotum and appeared grossly normal and was drained by its own vas deferens; given the low risk for malignancy and its potential to contribute to reproduction we left it *in situ* and counseled the parents on the remote risk for malignancy. Furthermore, we recommended careful follow-up with annual physical examinations in addition to self-testicular examinations once the child reaches puberty. Serological markers, ultrasonography, and/or MRI imaging can also be utilized for surveillance if additional risk factors for testicular cancer exist [1,5].

Learning points

(1) Polyorchidism is a rare congenital anomaly where three or more testicles are present.
(2) Polyorchidism carries an increased risk for malignancy and requires patient education and self-examination.
(3) Polyorchid testes may contribute to spermatogenesis and future fertility if it has adequate drainage of a deferent duct.

Conflicts of interest

There are no conflicts of interest.

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