

CASE REPORTS

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Management of a very rare case of polyorchidism: a case report

Wissam Jamal Al Tamr^{1†}, Kareem Omran^{2*†} , Rajebhosale Prashan³ and Agrawal Pooja³

Abstract

Background Polyorchidism is a rare genetic anomaly characterized by the patient having more than two testicles. In the literature, only around two hundred cases have been reported. We present an unusual new case which does not conform to the traditional classifications of polyorchidism or previously reported cases. We discuss our diagnostic techniques as well as management decisions for this case and aim to raise awareness about the management choices available.

Case report A 3-year-old male presented to the clinic by his parents due to concern of a painless swelling in the left inguinal region. Physical examination revealed features of a left indirect inguinal hernia. Furthermore, scrotal examination revealed a firm, non-compressible painless mass in the left scrotum which was inferior to the left testis. Ultrasoundography suggested the mass as an extra testis, and MRI was followed which showed no signs of malignancy. Due to the reassuring nature of these findings, a conservative approach was taken, with the patient referred for regular follow-up.

Conclusion When facing cases of polyorchidism, physicians should move away from considering surgical excision and biopsy as exclusive first-line management. Instead, we place emphasize and raise awareness about the option of conservative management if imaging shows no abnormalities. Ultimately, the aim of this paper is to raise awareness among the paediatric surgeon community that while excision may be a valid option, it is not the only treatment.

Keywords Polyorchidism, Scrotum, Testes, Ultrasound, Paediatric surgery, Urology

Background

Polyorchidism or supernumerary testicle is an uncommon congenital anomaly characterized by more than two testicles. In the literature, only around two hundred cases have been reported so far [1]. Hereby, we report a case of a 3-year-old boy who presented with a left inguinal hernia and was coincidentally found to have an additional

mass near the left testis which on surgical exploration was revealed to be an additional testicle. We reviewed the literature for polyorchidism and discuss the current management strategies and methods for diagnosis.

Case presentation

A 3-year-old male was brought to the clinic by his parents with the concern of a painless swelling in the left inguinal region which was incidentally noticed by the father since the age of 3 months.

Physical examination revealed features of left indirect inguinal hernia. Furthermore, scrotal examination revealed a firm, non-compressible painless mass in the left scrotum which was inferior to the left testis. The right testis was normal in size and position and in terms of consistency. No varicocele or lymphadenopathy was detected on either side.

Wissam Jamal Al Tamr and Kareem Omran should be considered joint first authors.

*Correspondence:

Kareem Omran
ko378@cam.ac.uk

¹ NMC Royal (Al-Zahra) Hospital, Sharjah, United Arab Emirates

² Department of Public Health and Primary Care, University of Cambridge, Cambridge CB2 1TN, UK

³ Al Zahra Hospital, Sharjah, United Arab Emirates



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Fig. 1 An ultrasound examination of the scrotum revealed another small homogeneous tissue in the left scrotal sac measuring 0.67×3.48 cm near the left testis

An ultrasound examination of the scrotum revealed bilateral testicular microlithiasis with left inguino-scrotal fatty hernia. It was noticed that there was another small homogeneous tissue in the left scrotal sac measuring 0.67×3.48 cm (Fig. 1). An MRI of the abdomen and pelvis followed. This was to check for any signs of malignancy, but it revealed no additional abnormalities. An alpha fetoprotein level was taken, as it may be a marker of malignancy; however, this was also normal.

The patient was then taken for surgery to repair the left inguinal hernia. It was observed to be an indirect hernia,

with two testes in the left scrotum. The superior testis was larger in size ($0.90 \times 0.94 \times 1.19$ cm) with a volume of 0.52 ml. It did not have an epididymis and vas deferens; however, it had a normal blood supply. The inferior testis was smaller in size (0.67×3.48 cm) and had an epididymis along with a vas deferens (Fig. 2). The patient underwent left inguinal hernia repair along with orchidopexy for both testes in the left scrotum, to prevent future cases of torsion.

A biopsy was deemed to be unnecessary due to the small nature of the extra testis. Moreover, the lack of

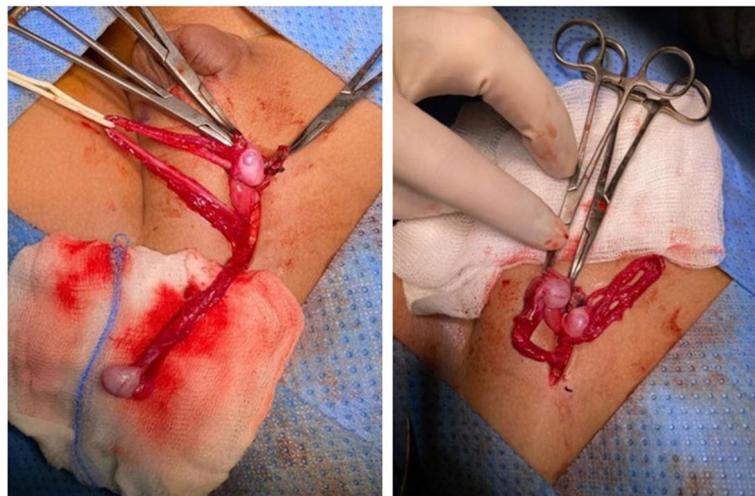


Fig. 2 During the surgery, we found a left indirect inguinal hernia along with two testes in the left scrotum. Superior testis was larger in size and was without epididymis or vas deferens but had a normal blood supply. The inferior testis was smaller in size and had an epididymis along with vas deferens

any gross abnormalities of the tissue and normal imaging made conservative management more assuring. The management plan was discussed with the parents prior to surgery, and it was decided that close follow-up in the future for the early detection of any associated complications was ideal.

Discussion

Polyorchidism/supernumerary testes is defined as the presence of more than two testes and is an extremely rare entity. Triorchidism, i.e. three testes, is one of the commonest forms of polyorchidism where the extra testis is usually found on the left side [2]. Furthermore, polyorchidism has been classified into four distinct types by Singer et al. [3], depending on the presence of an epididymis or a vas deferens. Although our index patient carries similarities to a “type D” (the supernumerary testicle has complete duplication of testicles), the lack of epididymis and vas deferens makes it a unique case to the literature.

Polyorchidism is typically asymptomatic, with presentation to healthcare being attributed to a mass being detected incidentally. However, sometimes, supernumerary testes are brought to clinical attention due to associated complications such as maldescent (40%), torsion (15%), inguinal hernias (30%), hydrocoele (9%), and malignancy (6%) [3]. The majority of documented malignancies are of the testicular origin and, in rare instances, rhabdomyosarcoma of the cremasteric muscle [4]. Most of these testicular malignancies were detected in the undescended extra testicle [5].

To investigate this genetic anomaly, first-line imaging is ultrasound [6]. Magnetic resonance imaging should then be followed for better visualization of signs of malignancy. Clinical examination alone is not sufficient for a diagnosis to be made. On ultrasound, supernumerary testes can be visualized as freely mobile, well-defined ovoid structure with an eco-structure like that of normal testes, and Doppler can be used to determine presence of blood flow [6].

Management of polyorchidism has been vastly discussed and debated. Some authors suggest for close follow-up, whereas others recommend excision of the extra testicle considering its malignant potential [4]. Recently, with advancement in monitoring through MRI and ultrasound imaging, a more conservative approach can be followed [5]. Conservative approaches involve the diagnosis and monitoring of the supernumerary testis with the help of high-resolution sonography and MRI. If there is no concomitant disorder and a testicular tumor is ruled out on imaging, surgical exploration with biopsy is not required [7]. On the other hand, surgical exploration allows biopsy along with

fixation of the testes to avoid torsion. Furthermore, it can also provide information regarding if the testis has an outflow tract and if the testis can contribute to the spermatogenesis [7]. Definitive indications of orchidectomy are malignant or dysplastic changes on biopsy, ultrasonography suggestive of malignancy, absent spermatogenesis, or situations where regular follow-up is not reliable [7]. Non-scrotal location of polyorchidism is considered as the most important risk factor for malignancy [5]. As such, we suggest that when planning for management of this condition, surgeons and paediatricians alike must not only consider the radiological findings but also the gross anatomy as well as societal barriers and availability of follow-up.

Conclusion

When facing cases of polyorchidism testes, physicians should move away from considering surgical excision and biopsy as exclusive first-line management. Instead, we place emphasize and raise awareness about the option of conservative management if imaging shows no abnormalities, and the patient can be readily followed up. Furthermore, we would like to emphasize the need for a scrotal examination in all paediatric patients presenting with signs of inguinal hernias, as this is a common manifestation for polyorchidism. With the varying nature of this anomaly, adequate and proper imaging using MRI is essential for screening for malignancy. However, the success of ultrasonography as both first-line imaging as well as a diagnostic tool for extra testes cannot be neglected.

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Authors' contributions

Dr. Wissam Jamal Al Tamr: senior surgeon who operated the case and supervised the write up. Kareem Omran: editing/write-up of manuscript and literature review. Dr. Rajebhosale Prashan: senior physician involved in the care of patient and write up of manuscript. Dr. Agrawal Pooja: senior physician involved in the care of patient and review of manuscript. The authors read and approved the final manuscript.

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Competing interests

The authors declare no competing interests.

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