An unusual case of supernumerary testes.

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October 17, 2022

Abstract

Supernumerary testes is a rare genetic anomaly characterized by 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 classifications of polyorchidism or previous cases and discuss our management.

Declarations:

Funding: N/A

Acknowledgments:

We disclose no conflict of interest, any sources of support or external funding.

Written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy.

Key clinical message:

When facing cases of supernumerary testes, physicians should move away from considering surgical excision and biopsy as exclusive or first line management and instead we place emphasise on conservative management if imaging shows no abnormalities.

Abstract:

Supernumerary testes is a rare genetic anomaly characterized by 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 classifications of polyorchidism or previous cases and discuss our management.

Keywords: Polyorchidism, Scrotum, Testes, Ultrasound, Paediatric surgery, Urology
**Background:**

Polyorchidism or supernumerary testicle is an uncommon congenital anomaly characterised 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. Further, we reviewed the literature for polyorchidism and current management strategies for it.

**Case presentation:**

A 3-year-old boy was brought by his parents with the concern of 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. Further, 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, position and in terms of consistency. No varicocele or lymphadenopathy was detected on either side.

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 X 3.48 cm. (Figure 1)

His alpha fetoprotein level was normal and MRI abdomen and pelvis did not reveal any other additional abnormality. During the surgery we found a left indirect inguinal hernia along with two testes in the left scrotum. The superior testis was larger in size (0.90 X0.94 X 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 X3.48 cm) and had an epididymis along with a vas deferens. (Figure 2). The patient underwent left inguinal hernia repair along with orchidopexy for both testes in the left scrotum. A biopsy was deemed to be unnecessary due to the small nature of the supernumerary testes, the lack of any gross abnormalities of the tissue and normal imaging. The plan was discussed with parents before surgery regarding close future follow up for the early detection of any associated complications.

**Discussion:**

Polyorchidism/Polyorchism 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). The most common location of the extra testis is inside the scrotum, superior or inferior to the ipsilateral testicle (3). In our present case, the supernumerary testis was found below the left testis, in the scrotum.

Usually, the patients present with an asymptomatic mass detected incidentally. However, these supernumerary testes are sometimes brought to clinical attention due to associated complications such as testicular torsion or malignancy (4). First line imaging should be ultrasound, followed by magnetic resonance imaging, as clinical examination alone is not sufficient for the diagnosis (5). On ultrasound the 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).

Singer et al (7) classified polyorchidism into four distinct types. Type A: The supernumerary testicle doesn’t have an epididymis and vas deferens. Type B: An epididymis and vas deferens is shared by supernumerary testicle with another testicle. Type C: The supernumerary testicle has its own epididymis but vas deferens is shared with a regular testicle. Type D: The supernumerary testicle has complete duplication of testicles, with its own epididymis and vas deferens. Our patient had a type D supernumerary testicle; however, the larger testis was devoid of epididymis and vas deferens, a feature which is unusual compared to other cases reported so far.
Bergholz and Wenke performed a meta-analysis of the 140 cases of histologically proven polyorchidism (8) and found triorchidism was the most common type. Most of the supernumerary testicles (64%) were drained by a vas deferens. There was a predominance of the left side with 64.5% of the patients being affected. Most cases were incidentally detected during surgery performed to treat other complaints like testicular torsion, inguinal hernia, scrotal pain or undescended testicle. These features are mimic our index patient. Further, in the same meta-analysis, 17 years was the median age at diagnosis which was much greater compared to our patient (3 years).

Complications related with polyorchidism are maldescent (40%), torsion (15%), inguinal hernias (30%), hydrocoele (9%) and malignancy (6%) (9). Most malignancies reported are of the testicular origin and in rare instances rhabdomyosarcoma of the cremasteric muscle (9), and most of the malignancies were detected in the undescended supernumerary testicle (8).

Management of polyorchidism has been vastly discussed and debated. Some authors suggest for close follow-up whereas, others recommend excision of the supernumerary testicle considering its malignant potential (9). Recently, with advancement in monitoring through MRI and ultrasound imaging, a more conservative approach can be followed (8).

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 (11). On the other hand, surgical exploration allows biopsy along with fixation of the testes to avoid torsion. Further, it can also provide information regarding if the testis has an outflow tract and if testis can contribute to the spermatogenesis (11).

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 (11). Non-scrotal location of supernumerary testis is considered as the most important risk factor for malignancy (8). Considering the risk of malignancy was deemed to be very low in our case due to scrotal placement of the extra testis and normal imaging, we decided to preserve it with close follow-up with the help of high-resolution ultrasound for early detection of any potential complication.

**Conclusion:**

Since supernumerary testes is an extremely uncommon condition there is no clear consensus in the management. In this case report, we emphasize the success of ultrasonography in the diagnosis of extra testes and the need for a scrotal examination in paediatric patients presenting with signs of inguinal hernias. We establish that when considering management options, the risk of malignancy should be the main consideration before engaging with surgical excision and that biopsy is not essential if imaging shows no abnormalities.

**References:**


Figure legends:

**Figure 1:** An ultrasound examination of the scrotum revealed another small homogeneous tissue in the left scrotal sac measuring 0.67 X 3.48 cm near the left testis.

**Figure 2:** During the surgery we found 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 however had normal blood supply. Inferior testis was smaller in size and had epididymis along with vas deferens.