

Triorchidism: A case report and review of the literature

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Abstract

Polyorchidism or supernumerary testes is a very rare condition with only about 200 confirmed cases in the current literature. Most frequently, polyorchidism is found to be a triorchidism and intrascrotal in position, with primordial affection of the left side.

In theory, only histological confirmation can provide definite diagnosis, however, nowadays this condition can be diagnosed and differentiated from other extra- and paratesticular masses by ultrasound and subsequent MRI, being the characteristic ultrasound and MRI features of normal testicular tissue.

Further management depends on the position of the supernumerary testicle, i.e. scrotal or ectopic. Also the reproductive potential and possible concomitant symptoms and complications have to be taken into account in choosing for orchiectomy versus follow-up.

Keywords

polyorchidism; triorchidism; supernumerary testicle; MRI; ultrasound

Case Presentation

A 16-year old male in perfect condition presented himself at the urology department at our hospital after being transferred by his general practitioner for a painless swelling in the left hemiscrotum that had been persisting for a few weeks. This patient has had no significant problems in his clinical history.

Clinical investigation confirmed a small, mobile, smooth swelling at the level of the epididymis, superior to the left testicle. This testicle was somewhat smaller than the right one. The right testicle appeared normal on palpation. The results of the blood work came back negative, alfa-1 fetal protein was normal, as was Beta Human choriogonadotrofin and lactate dehydrogenase.

Ultrasound, as well as MRI (T1- and T2-weighted images in three orthogonal planes and fat saturated T1-weighted images after intravenously administered Gadolinium contrast), both showed the right testicle being somewhat bigger than the left (4.2 vs 3.5 cm in length). Superior to the left testicle an extra-testicular "mass" was found, measuring 1.8 cm in length. Both testicles and the "mass" were homogeneously isoechogenic relatively to each other on ultrasound; no intratesticular lesions were discovered, nor was any hypervascularisation (figure 1). One hyperechoic epididymis was uncovered adjacent to the native and supernumerary testicle in the left hemiscrotum.

MRI produced similar findings: comparable hyperintensity on T2 and hypointensity on T1, with homogenous contrast enhancement (figures 2-5). And one adjacent T1-isointense and T2-hypointense epididymis. The diagnosis of triorchidism with a scrotal supernumerary testicle which shared its epididymis with the ipsilateral testicle was made (figures 1 and 6).

A small cyst in the right epididymis was an additional finding.

No histological confirmation has been performed, no surgery has been planned; the patient will be followed up regularly by ultrasound.

Discussion

Polyorchidism or supernumerary testes is a very rare condition with only about 200 confirmed cases in the current literature [1]. Blasius had described the first case in 1670 and first histological confirmation was provided in 1895 by Lane [1,2]. Most frequently, polyorchidism is found to be a triorchidism with intrascrotal position of the testes (in 66% of patients) and primordial affection of the left side. Other frequent positions are inguinal and retroperitoneal [3].

Several theories have been circulating regarding the embryological origin of testicular duplication: amongst others a division and duplication of the gonadal ridge and degeneration of the mesonephric (Wolffian) duct (testicular tissue originates from the gonadal ridge and the epididymis and vas deferens from the mesonephric duct) [4,5].

Based on this transverse division or doubling of the gonadal ridge, Leung proposed a classification of polyorchidism into 4 categories (figure 7). Only a small part of the gonadal ridge divides in type A polyorchidism, without the mesonephric duct; so the supernumerary testicle lacks an epididymis and vas deferens. In type B, division takes place at the part of the gonadal ridge where the primordial gonads are connected to the mesonephric duct, therefore both testes share the same epididymis and vas deferens. If an incomplete division of the mesonephros, attached to the gonadal ridge, occurs, both testes and epididymis only share the vas deferens (Type C). In type D (the least common) complete division occurs and both testes are drained by their own epididymis and vas deferens [4].

Polyorchidism is an incidental finding in most cases, since only a few patients complain about symptoms, caused by concomitant disorders and complications. These complications most commonly involve maldescent and inguinal hernia, followed by testicular torsion. Also associated are hydrocele, varicocele and spermatocele [3].

A supernumerary testicle should be differentiated from other extratesticular masses (hydroceles, varicoceles, lipomas, tumors, etc.) and paratesticular masses (hernias, scrotal calculi, etc.) [6-8].

In theory, only histological confirmation can provide definite diagnosis, however nowadays, we can diagnose a supernumerary testicle with near certainty using both ultrasound and MRI (after intravenously administered Gadolinium contrast) [6,9].

With a low cost and readily available, scrotal ultrasound has a high sensitivity in detecting subclinical lesions in the testes and malignant features in a palpable lump. Normal testicular tissue has a homogeneous and mildly coarse echotexture. The tunica vaginalis/albuginea appears as an echogenic outline of the testicle and the rete testis contains of a hypoechoic region near its mediastinum. The

epididymis is isoechoic or mildly hyperechoic relative to the testicle: the head is a round or oblong structure located near the superior pole of the testicle, which extends down to the body and tail, and further into the vas deferens. On spectral Doppler, both the testicle and epididymis demonstrate a low-resistance arterial waveform. A similar ultrasound pattern and vascular flow as the normal ipsilateral testicle is found in the supernumerary testicle (figure 1) [7,9-11].

The supernumerary testicle also has the same MRI characteristics as a normal testicle: it demonstrates a homogenous iso- to hyperintensity on T2-weighted MR images (Figures 2 and 3) and hypo- to isointensity on T1-weighted images (figures 4-6). The testicle is encapsulated by the T1- and T2-hypointense tunica albuginea. The epididymis is isointense on T1-weighted images, relative to testicular tissue, but hypointense on T2-weighted images. After intravenous gadolinium-based contrast administration, the testicle and the epididymis enhance in a homogenous pattern (figures 5 and 6).[6-9]MRI is superior in detection and diagnosis of lesions of the testicle, and is very helpful if ultrasound diagnosis is not certain; but appears to have only a confirmative role in uncomplicated case. Furthermore, the cost of MRI should be kept in mind when suggesting it for follow-up [7,9].

Even so, there is a suspected increased risk for malignancy and a known increased risk for maldescent [3,12]. Therefore, there is some discussion about management of cryptorchidism: whether to follow up on regular intervals, or to perform an orchidectomy. Generally, management is guided by the classification of the supernumerary testicle and the presence (or absence) of concomitant symptoms or complications [3,5,10,13].

Singer proposes a classification for management based on anatomical variation and reproductive potential. Type 1 includes accessory testes which have a reproductive potential, meaning they are connected to an epididymis and vas deferens. A type 1a polyorchidismis located in the scrotum, type 1b in any ectopic position. Type 2 testes have no attachment to the testicular drainage system and are subsequently divided in a type a and type b, intrascrotal and ectopic respectively [13].

In their meta-analysis, Bergholz et al. also tried to define a management protocol, based on the classification of polyorchidism proposed in their case report published in 2007 (figure 6) [1,3]. They suggest a division into two types. Namely supernumerary testes with drainage by a vas deferens (type A) and without vas deferens drainage (type B), the latter by definition not contributing in spermatogenesis. Further subdivision is made according to the attached structures: type A1 has its own epididymis and vas deferens, type A2 with its own epididymis and a shared vas deferens, and the last type, A3, shares its epididymis and vas deferens. Type B is subdivided in type B1 with its own epididymis and type B2 without epididymis [1].

Following management protocol has been introduced by a few authors. Since ectopic testes have an increased risk of malignancy, orchiectomy should be performed. Scrotal testes that have no reproductive potential (no epididymis and vas deferens) should be biopsied, and will be removed in case of malignant or dysplastic change. Scrotal testes that have a reproductive potential should be removed in case of a positive ultrasound for suspected malignancy. They may also be removed if biopsy reveals absent spermatogenesis, and following the patient's wish esorin circumstances where follow-up is unlikely [3,5,13].

Uncomplicated scrotal polyorchidism with reproductive potential should be managed by follow-up with imaging and serum markers. An argument in favor is that sampling error or the focality of the disease can yield a negative biopsy result, so these cases can be easily placed on a follow-up regimen [10].

Conclusion

Polyorchidism is a very rare condition that can be diagnosed and differentiated from other extra- and paratesticular masses by ultrasound and subsequent MRI, given its characteristic ultrasound and MRI features.

Further management depends the position of the supernumerary testicle, being scrotal or ectopic. Together with that, the reproductive potential and possible concomitant symptoms and complications have to be taken into account in choosing for orchiectomy versus follow-up.

Figures

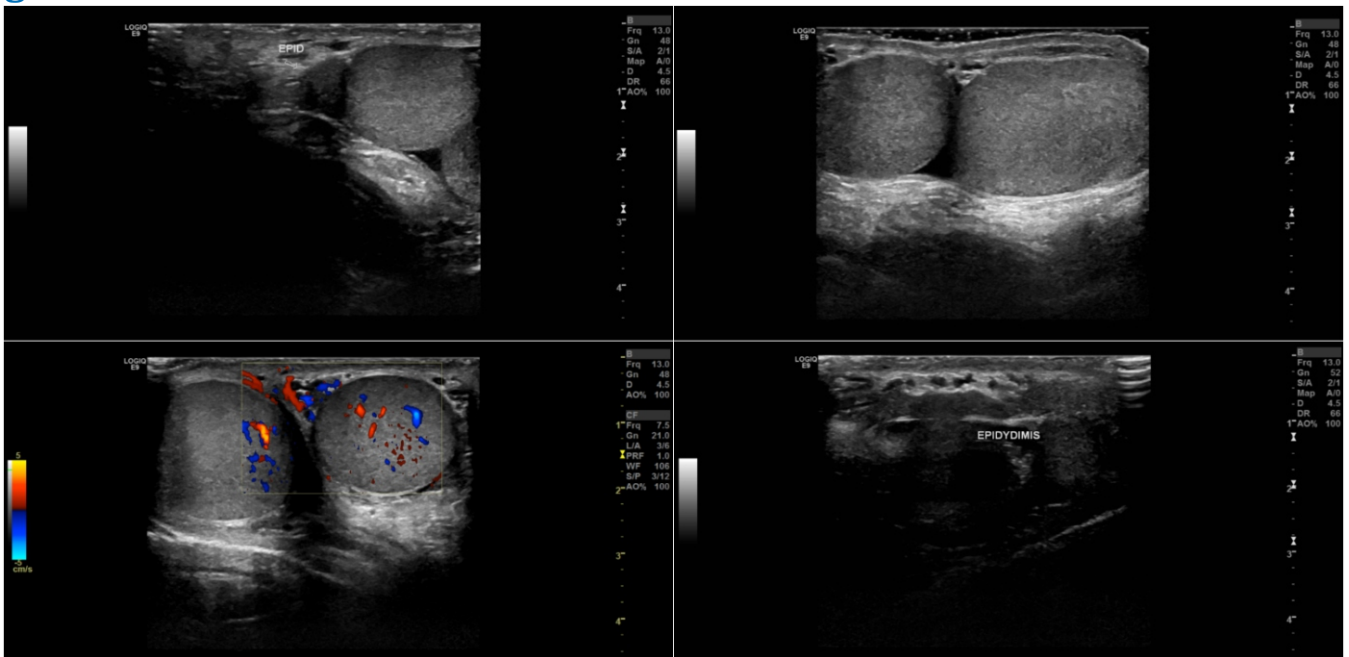


Figure 1: Ultrasound imaging showing similar echogenic characteristics and vascularization between the ipsilateral testes, as well as the epididymis to the supernumerary testicle. Bottom left shows the left epididymis in its entire length, adjacent to both ipsilateral testes and connected to their respective efferent ductules.

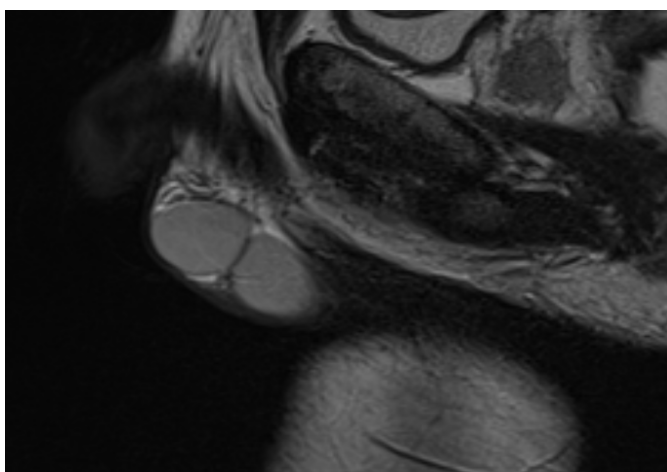


Figure 2: Sagittal T2 shows the supernumerary testicle in the left hemiscrotum, ventrocranial to the native testicle. Both are comparable homogeneously isointense.

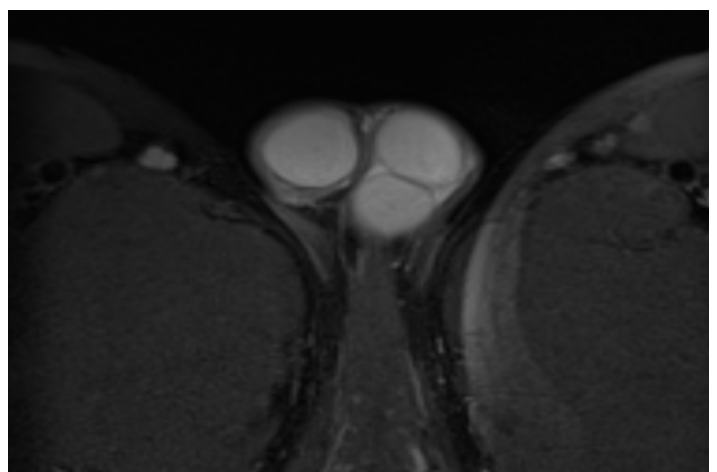


Figure 3: Axial T2 with fat saturation, shows supernumerary testicle in the left hemiscrotum. The two ipsilateral testes and the contralateral one are homogeneously iso- to hyperintense.

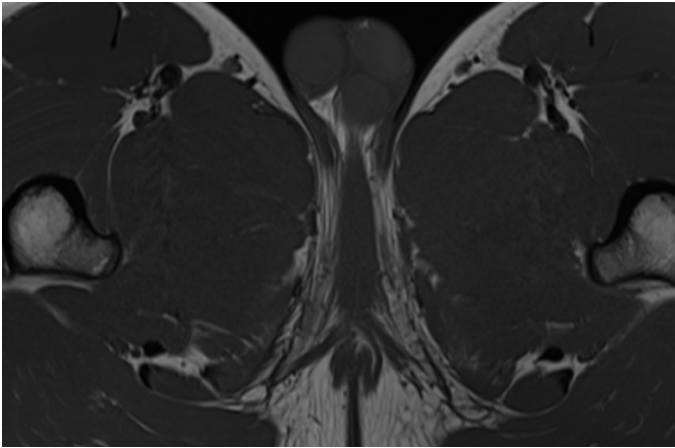


Figure 4: Axial T1, the two native testes and the supernumerary testicle have a homogenous T1-hypo-intense signal.

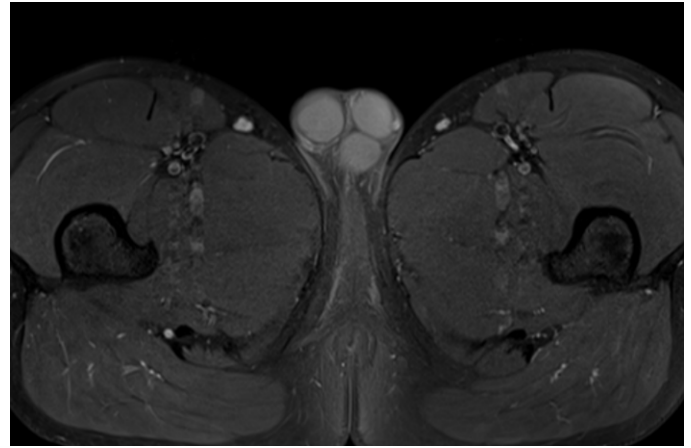


Figure 5: Axial T1 with fat saturation, after administration of intravenous Gadolinium contrast, shows a supernumerary testicle in the left hemiscrotum. There are comparable intensities and homogenous contrast-enhancement between the two ipsilateral testes, and the contralateral one.

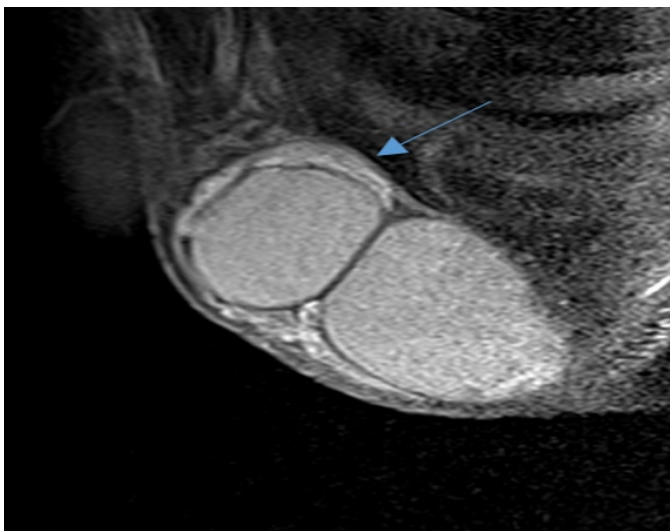


Figure 6: Sagittal T1 with fat saturation after administration of Gadolinium contrast. Both ipsilateral testes enhance in a similar fashion. The head of the epididymis is adjacent to the supernumerary testicle (arrow) and is connected to its efferent ductules.

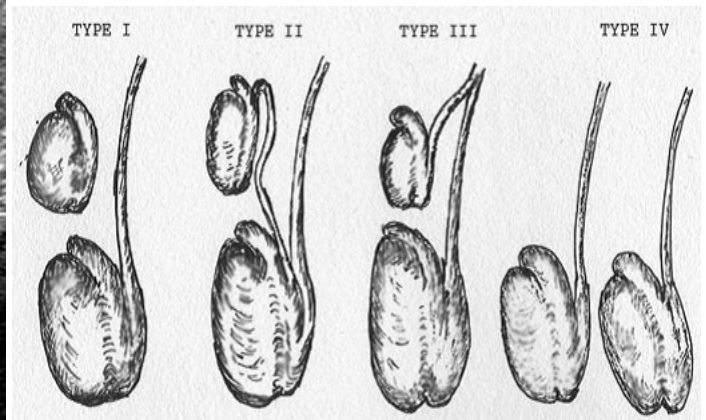


Figure 7: classification by Leung, from Nayak et. al. 2001

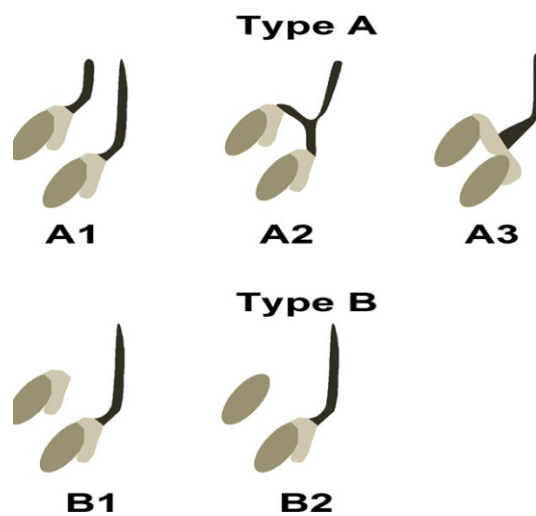


Figure 8: classification by Bergholz, from Bergholz et. al. 2007

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