Neoplasm of a supernumerary undescended testis: A case report and review of the literature

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ABSTRACT

INTRODUCTION: Polyorchidism, defined as the presence of more than two testicles, is a rare congenital abnormality of the male genital tract. There is no consensus regarding the management of supernumerary testis (SNT) due to its rareness. To the best of our knowledge, this is the first report of leiomyoma in SNT.

PRESENTATION OF CASE: We report the case of a 41-year-old man with two right testes sharing a common vas deferens and separate epididymides. Orchiectomy of right testes was performed. Pathology examination found that the superior testis was the site of a leiomyoma and the lower testis was the site of an intratubular germ cell neoplasia (IGCN). Orchiectomy and biopsy were later performed for the left testis. Histology was compatible with IGCN. We opted for follow-up to avoid risks of hypogonadism.

DISCUSSION: Polyorchidism is usually asymptomatic and found incidentally during surgery for inguinal hernia, undescended testes as in our case, torsion, hydrocele or testicular tumor. If the SNT is scrotal, and there is no other indication for surgery, most authors recommend conservative management with regular ultrasound follow-up. If nonscrotal SNT is found incidentally during surgery, orchiectomy could be performed because of increased risk of malignancy. Treatment of IGCN includes surveillance, orchiectomy, or low-dose external radiation.

CONCLUSION: Different factors come into account for polyorchidism management: the drainage system, the fertile potential of the supernumerary gonad, and its localization. In cases of uncomplicated polyorchidism, a conservative treatment, with US or MRI follow-up seems to be a rational choice without surgical complications.

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1. Introduction

Polyorchidism, defined as the presence of more than two testicles, is a rare congenital abnormality of the male genital tract. Triorchidism is the most common variant. Two important classifications of polyorchidism are the embryological one of Leung and the vascular one of Bergholz. Usually asymptomatic, it can be revealed by an abdominal mass, a testicular torsion or an inguinal hernia. Its management is controversial depending on different factors. To the best of our knowledge, we report the first case of leiomyoma in supernumerary testis. We summarize also the classifications and management of polyorchidism. Our work has been reported in line with the SCARE criteria [1].

2. Presentation of case

A 41-year-old man presented to our institute for bilateral inguinal swelling and infertility for 15 years. He stated that his scrotum was always empty since childhood. There was no other medical history.

Physical examination found a palpable right testis in the right lower abdomen along with an empty scrotum. The left testis was not palpable. Ultrasonography (US) showed both undescended right and left testes respectively in the right and left inguinal canals. Both testes had heterogeneous echogenicity. Right testis measured 45 × 30 × 13 mm while left testis measured 24 × 20 × 10 mm.

Due to malignancy risk we decided to do an orchiectomy with testicular biopsy one side at a time. Pre-operative serum tumor markers (Lactate dehydrogenase, beta human chorionic gonadotropin, alpha fetoprotein) were normal.

He was operated upon through right inguinal incision. There was no hernia sac. Two structures, identified as two right testes, were sharing a common vas deferens and a double separate epididymides. The superior testis measured 80 × 50 × 35 mm and was highly dysmorphic and suspicious, while the lower testis measured...
40 × 30 × 25 mm and seemed macroscopically normal. The superior
testis drains into the epididymis of the lower testis. Orchiectomy
for the two right testes was performed (Figs. 1 and 2). Pathology
examination confirmed the testicular nature of the two structures
and found that the dysmorphic superior testis was the site of a large
leiomysarcoma and the lower testis was the site of an intratubular germ
cell neoplasia (IGCN).

We planned for surgical exploration directed to the left-sided
cryptorchidism by an inguinal approach. An eutrophic and macro-
scopically normal left testis was found. A complete separation
between the epididymis and the testis with a mesentery between
them was noted (Fig. 3). Testicular biopsies followed by a left
orchiopexy in dartos were performed. Histopathological exami-
nation of the specimen was compatible with IGCN. We opted for
surveillance with follow-up and regular testicular self-examination
and US.

3. Discussion

Polyorchidism is rare with around 200 cases reported in the
literature to date [2]. Leung described the first classification by
embryological development with functional implications as fol-
low:

- **type I:** supernumerary testis (SNT) without epididymis and vas
derference.
- **type II:** SNT shares common epididymis and vas deference with
ipsilateral testis.
- **type III:** SNT has its own epididymis but shares a common vas
derference with the ipsilateral testis.
- **type IV:** complete duplication of testis, epididymis and vas.

According to Leung, types II and III together represent more than
90% of the cases [3].

Bergholz and Wenke proposed an innovative classification. A
testis being drained by an outflow path was coded as **type A.**
Undrained testes without connection to a draining vas defer-
ens were coded as **type B.** Type A testes were further divided
into the subgroups A1 (own epididymis and vas deferens),
A2 (own epididymis but common vas deferens), A3 (common
epididymis and vas deferens), A4 (own vas deferens but com-
mon epididymis) and AX (no further description of appendixes).
Type B testes were divided into the subgroups B1 (own ep-i-
didymis), B2 (no epididymis, thus testicular tissue only) and BX
(no further description of appendixes) [4]. Our case was com-
patible with Type III according to Leung and A2 according to
Bergholz.
Most cases of polyorchidism are asymptomatic and are found incidentally during surgery for inguinal hernia (30%), undescended testes (15–30%) as in our case, torsion (13%), hydrocele (9%) or testicular tumor [2]. In a series of 140 polyorchidism cases, Bergholz [4] reported 10 cases of testicular tumor (7.14%): 2 were benign (rete testis adenoma) and 8 were malignant (3 seminomas, 2 choriorcincinomas, 2 teratomas, 1 embryonal carcinoma). 7 of these 8 cases were undescended SNT. To the best of our knowledge, this is the first report of leiomyoma in SNT.

There is no consensus regarding the management of SNT due to its rarity. If the SNT is scrotal, confirmed by ultrasound or magnetic resonance imaging (MRI), and there is no other indication for surgery, most authors recommend conservative management with regular ultrasound follow-up [2,4,5]. If nonscrotal SNT is found incidentally during surgery, orchiectomy could be performed because of increased risk of malignancy [2,4]. However, type A testes are best treated with orchiectomy to prevent future torsion and allow follow-up by US and self-examination. Biopsy of the SNT will prove the histology and provide an evaluation of the degree of spermatogenesis. For our patient, the two right testes were managed by orchiectomy. Histopathological examination corresponded to leiomyoma for the supernumerary testis and to intratubular germ cell neoplasia (IGCN) for the usual right and left testes. Testicular leiomyoma is rare and may be confused with malignancy. ICGN has been reported to be less than 1% in the normal male population, and 0.43% to 0.8% in autopsy studies, with most cases being unilateral [6].

Treatment of ICGN includes surveillance, orchiectomy, or low-dose external radiation (LDRT). For patients with unilateral ICGN and a normal contralateral testis, orchiectomy is the preferred treatment option. ICGN in solitary testis is more controversial, with some advocating surveillance with testicular self-examination and others recommending early LDRT [6].

For our patient, we opted for conservative management with regular US and follow-up to avoid risks of hypogonadism.

4. Conclusion

The diagnosis of polyorchidism is often occasional. Different factors come into account when choosing their management: the drainage system, the fertile potential of the supernumerary gonad, and its localization. Surgery should only be considered appropriate if there is an associated pathology. Indeed, in cases of uncomplicated polyorchidism, a conservative treatment, with US or MRI follow-up seems to be a rational choice without surgical complications.

Conflicts of interest

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Ethical approval

Ibn El Jazzar Teaching Hospital ethic comittee, Kairouan, Tunisia.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Boussaфа H: concept or design, data collection, data analysis or interpretation, writing the paper.

Naouar S: concept or design, data collection, data analysis or interpretation, writing the paper.

Ati N: data collection, data analysis or interpretation.

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Ben Khelifa B: data collection.

Salem B: data collection, data analysis or interpretation, writing the paper.

El Kamel R: writing the paper.

Registration of research studies

This is no research study.

Guarantor

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