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CASE REPORT

Polyorchidism with varicocele: a case report and review of literature

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Summary

Polyorchidism is defined as the presence of more than 2 histologically proven testes. We report the case of a 20-year-old man with polyorchidism, presenting with right scrotal pain and right scrotal ovoid mass. Scrotal examination revealed two ovoid, mobile lumps with testicular sensation in the right side of the scrotum. Scrotal colour Doppler ultrasonography and magnetic resonance imaging confirmed the presence of double testes with common epididymis and common vas. Microscopic varicocele ligation was performed, and then, two ipsilateral testes were sutured together. The testes were then returned to the scrotum with fixation.

Introduction

Polyorchidism is a rare congenital anomaly that is not well known by most urologists. Polyorchidism is often complicated by inguinal hernia, hydrocele, maldescent testis, varicocele, spermatic cord torsion or neoplasms. It is found predominantly on the left side. But in our case the accessory testicle was present in the right scrotum. To the best of our knowledge, we report on the second right-sided polyorchidism with varicocele in the literature.

Case report

A 20-year-old man was admitted to our outpatient clinic with a history of right scrotal mass and right testicular pain. On palpation, left testis seemed normal. On examination, two masses of equal size were palpable in the right scrotum and without firm or hard areas. Both masses were of equal texture similar to his normal left testis. Other system examinations were normal. Physical examination revealed normal secondary sexual characters. Laboratory analysis, including tumour markers (AFP, βhCG and LDH), was all within normal ranges. His spermiogram showed oligoasthenospermia. The mean values on two semen evaluations in 20 days showed a moderate oligoasthenospermia. Semen analysis one revealed 15 million sperm per ml, 30% motility[8% fast forward], 4.0 ml volume, and semen analysis two revealed 17 million sperm per ml, 35% motility[10% fast forward], 3.2 ml volume. Follicle-stimulating hormone and testosterone levels were normal. Abdominal ultrasonography and chest radiography were normal. Scrotal colour Doppler ultrasonography demonstrated a small testis ($3.4 \times 2.9 \times 2.3$ cm) on the right with an additional mass upper of equal echogenicity and almost equal size ($3.1 \times 2.4 \times 1.8$ cm). Multiple dilated tortuous veins were seen in right supratesticular regions, around the posterior and inferior aspects of the testes with maximum vein diameter 5.2 mm, showing flow in rest that augmented by Valsalva manoeuvre. Ultrasonography suggested the presence of two different homolateral testes with separate blood supply. There was no focal abnormal echogenicity suggesting malignancy. Magnetic resonance imaging confirmed two separate testes in the right hemiscrotum. The

patient was informed about surgery, and his written approval was obtained before the surgery. Exploration of the right inguinal region was performed under spinal anaesthesia. Inguinal region was opened, and testes were delivered. Two testes surrounded by a common tunica vaginalis were found. The distal testis was measured as $3.7 \times 3 \times 2.5$ cm and the proximal testis $3.1 \times 2.5 \times 2$ cm. Two right testes have common epididymis and vas (Figs 1 and 2). Biopsies were performed in both testes, revealed normal spermatogonial proliferation. No signs of malignancy were present. Microscopic varicocele ligation was performed at subinguinal level, and then, two ipsilateral testes were preserved with suturing together creating a single mass. The testes were then returned to the scrotum with fixation to prevent torsion. After 1 year, the mean values on two semen evaluations in 20 days improved (22 million sperm per ml, 60% motility, 25% fast forward, 3.0 ml volume). After 1 year, scrotal ultrasonography, tumour markers and physical examination were normally.

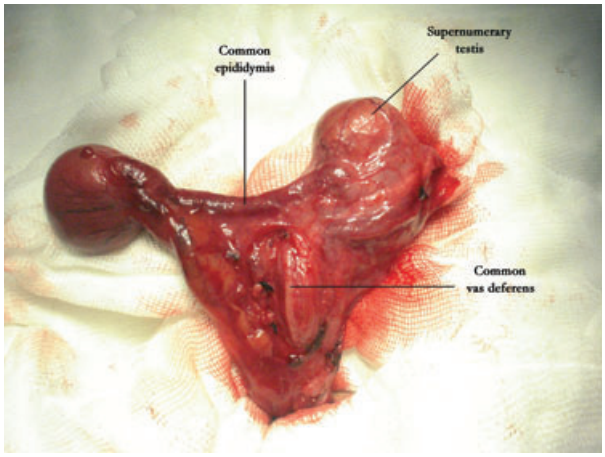

Fig. 1 Intraoperative photograph of the two right testes. There was a single epididymis and vas deference. Type A3 polyorchidism according to Bergholz's classification.

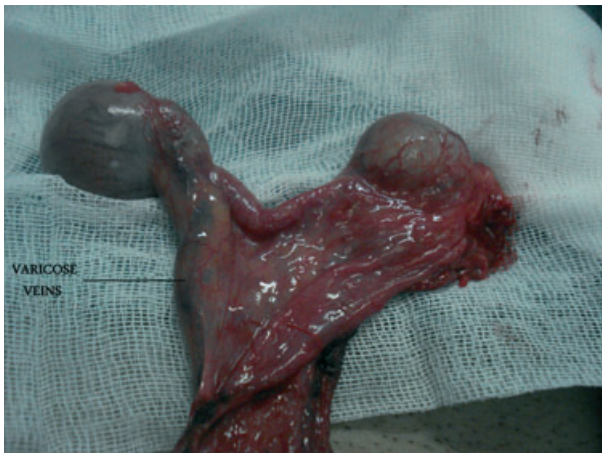

Fig. 2 Intraoperative photograph of the two right testes and varicose veins.

Discussion

Polyorchidism is a rare congenital anomaly. It is defined as the presence of more than 2 histologically proven testes. Although three testes are the most common forms, as many as five have been reported (Woodward *et al.*, 2003). The patient usually presents for evaluation of a scrotal mass. Supernumerary testicles are present usually within the scrotum. It can be located in the scrotum, inguinal region or in the abdomen. Among 75% of cases, the supernumerary testes were intrascrotal, 20% of the testes were inguinal and 5% were retroperitoneal (Bostwick, 1997). The first case was described in 1670, but the first histologically proven case was reported in 1895 (Wolf & Youngson, 1998). To date, approximately 150 histologically proven cases have been reported in the literature.

The exact cause of polyorchidism is unknown, although developmental errors in the formation of the testes are suspected. The testes develop embryologically from a mesodermal band called the urogenital ridge. The epididymis and vas deferens develop from mesonephric ducts. After 3 months of development, urogenital union of the urogenital ridge and mesonephric ducts begins, and they undergo transverse division (Cookson *et al.*, 2001). Embryological theories responsible for polyorchidism include duplication of the genital ridge or division of the genital ridge (Wilson & Litter, 1953; Nocks, 1978; Thum, 1991; Singer *et al.*, 1992). The most likely is that polyorchidism results from transverse division of the urogenital ridge between the fourth and sixth weeks. The theory of genital ridge duplication only explains some of the variations found. If a more medial duplicated ridge existed, it would be less likely to communicate with the mesonephric tubules and therefore would lack an outflow path, which only occurs some of the time with polyorchidism (Nocks, 1978; Lawrentschuk & MacGregor, 2004). This leaves only division of the genital ridge that accounts for all variations. As the primitive gonadal ridge is an elongated structure, transverse fragmentation could take place at any level and extend onto the mesonephric duct (Nocks, 1978; Lawrentschuk & MacGregor, 2004). This would explain the inequality of the two testes (Wolf & Youngson, 1998; Cookson *et al.*, 2001). If the separation was located caudal to the persisting group of mesonephric tubules, the superior gonad would develop spermatogenically active tubules and the caudal gonad would lack an

outflow path (Nocks, 1978; Lawrentschuk & MacGregor, 2004).

Although there are no definite statistics regarding the types of polyorchidism in the literature, it appears that duplication of the testes with a single epididymis and vas deferens is more common (Giyanani *et al.*, 1987). The more common anomalies associated with polyorchidism are maldescent of one of the supernumerary testes (15–50%), inguinal hernia (30%) and testicular torsion (13%) (Giyanani *et al.*, 1987). Other associations include hydrocele, epididymitis, varicocele, infertility, cryptorchidism and malignancy (Giyanani *et al.*, 1987).

Singer *et al.* (1992) suggested an anatomical as well as functional classification of polyorchidism. Type 1: supernumerary testes attached to the draining epididymis and vas deferens with reproductive potential. Type 2: testes with lack of such an attachment without having any reproductive potential. Each of these 2 types is again subdivided into two groups (A and B) depending on their location in the scrotum (orthotopic) or outside the scrotal sac (ectopic), respectively. In our case was type 1A according to Singer's classification. On the basis of embryologic development, Thum classified polyorchidism into three types. Type 1: The supernumerary testis lacks an epididymis and vas. Type 2: The supernumerary testis is linked to the regular testis by a common epididymis and shares a common vas with it. Type 3: The supernumerary testis has its own epididymis but shares the vas with the regular testis (Thum, 1991). In our case was type 2 according to Thum's classification. The other classification was proposed by Bergholz and associates. They are based upon reproductive potential of the supernumerary testis. They were suggested, a testis being drained by a deferent duct is coded as type A; otherwise, type B. Type A divided into three subgroups, and type B divided into two subgroups. A1: The drained supernumerary testis has its own epididymis and vas. A2: The drained supernumerary testis can have its own epididymis but shares a common deferent duct with its neighbour. A3: The drained supernumerary testis can share a common epididymis (and duct, subsequently) with its neighbour. B1: The undrained supernumerary testis does have its own epididymis. B2: The undrained supernumerary testis does not have its own epididymis, thus consisting of testicular tissue only (Bergholz *et al.*, 2007). In our case was type A3 according to Bergholz's classification.

The accessory testicle usually presents as a scrotal mass predominantly left-sided (Wolf & Youngson, 1998). But in our case, the accessory testicle was present in the right scrotum. Although most undescended testes are to be found on the right side, polyorchidism is frequently seen on the left side and those findings are one of the questions requiring an explanation. The larger size of the left testis might subdivide more readily than the right testis during development (Shah *et al.*, 1992).

There are characteristic sonographic features of polyorchidism, and the diagnosis is often made on the basis of sonography findings (Amodio *et al.*, 2004). Magnetic resonance imaging can be used for confirmation but may be more helpful in case complicated by cryptorchidism or neoplasia. The high accuracy of a preoperative ultrasonographic evaluation of scrotal masses differentiates and thus prevents unnecessary surgical exploration of sonographically normal, uncomplicated and orthotopic supernumerary testicles (Amodio *et al.*, 2004).

Previously, some authors suggested orchiectomy of the supernumerary testis in adults, even if orthotopic and functional, because of the risk of malignancy. There is no evidence in the literature to support this opinion. More recently, conservative management has been advocated. Conservative approach has made popular because of the availability of more sophisticated imaging modalities in the form of ultrasound and magnetic resonance imaging. The low incidence and its frequent association with proved risk factors, such as cryptorchidism, make it difficult to estimate its true malignant potential. Thus, a careful diagnosis is necessary in every suspicious testicular or scrotal finding. Owing to the described increased association with testicular malignancy, patients with polyorchidism should undergo regular clinical and ultrasound examinations. Testicular tumours were found in 6.4% of cases (Bergholz & Wenke, 2009). Nearly, all malignancies were associated with abdominal and inguinal maldescended testes. Polyorchidism and cryptorchidism in combination appear to be most important risk factors for testicular malignancy (Bergholz & Wenke, 2009). The management and evaluation of polyorchidism have two significant factors. First is the reproductive potential of polyorchid. The second concern is the potential for malignancy in the polyorchid testicle. Khedis and associates suggested that patients with polyorchidism can divide into two major groups; 1-Simple polyorchidism: Supernumerary testes are in orthotopic position without any signs of malignancy and without any association with other abnormalities. They were suggesting conservative management in this group. 2-Complicated Polyorchidism: Supernumerary testes are associated with other abnormalities. This group was divided into three subgroups. 2a-Polyorchidism plus signs of malignancy (Ultrasonographic Features, Elevated Tumor Marker Levels). They were advised to perform radical orchiectomy of all testes on the suspicious side, but in specific cases removal of the suspicious testis only is feasible: young patient motivated to close observation, small tumour. 2b-Polyorchidism plus torsion. They were advised orchidopexy if the testis is not necrotic; in other situation, they were advised orchiectomy.

2c-Polyorchidism plus cryptorchidism. If the patient is young, they were advised orchidopexy. In contrast, in adults, a supernumerary testis in the ectopic position implies an unnecessary risk of malignancy. Hence, they were advised excision of the supernumerary functional ectopic testis in adults (Khedis *et al.*, 2008). Bergholz and Wenke proposed the following protocol for supernumerary testes. Nonscrotal type A or B (according to Bergholz's classification) supernumerary testes should be managed by orchiectomy because of the increased risk of malignancy. Scrotal type A polyorchidism detached by imaging and without suspicion of malignancy or need for surgery owing to concomitant symptoms should be managed by watchful waiting and regular follow-up. Scrotal type A polyorchidism detected perioperatively or on imaging complicated by symptoms requiring surgery or suspicious for malignancy, and all scrotal type B (undrained) cases detected perioperatively or on imaging require biopsy with intraoperative frozen section and histological evaluation followed by orchiectomy(if there are signs of malignancy) or orchiopexy(to prevent testicular torsion) (Bergholz & Wenke, 2009).

Conclusion

Polyorchidism or supernumerary testis is a rare congenital anomaly. To the best of our knowledge, we report on the second right-sided polyorchidism with varicocele in the literature. In the absence of any concomitant disorder and if testicular tumour can be ruled out by imaging techniques and tumour markers, surgical exploration with biopsy is unnecessary. Supernumerary testis may be found incidentally during a groin exploration. In this condition, frozen section examination should be performed in the operation. If testicular tumour can be ruled out, orchidopexy should be performed to minimise the chances of torsion, malignancy and infertility. The psychological and cosmetic considerations need to be taken into account as well. Although the surgeons mention orchiopexy for prevention of torsion, the decision to fix the two testes together to make a single functional mass should be emphasised. So they can appear as a single testicular mass on inspection. There is a reservation that suturation of testes together side by side may cause problem in selfexamination of testes by the patient (Lawrentschuk, 2010). They claimed that the tumour arising from the suture sides cannot be palpated and it will delete the diagnosis. We disagree with this reservation. In our case, the suture line could be easily examined circularly with palpation. We also think that became it is sutured with a thin line and ultrasonography control is performed annually, this will not create a problem in follow-up of the patients in the future.

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