Triorchidism: An Incidental Finding And Review Of Literature


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Abstract. Polyorchidism is a rare congenital anomaly with less than 100 cases reported in the world literature. Several theories have been proposed regarding the production of polyorchidism, but the exact explanation is still not known. In majority of the reported cases, the patients are asymptomatic and have a painless groin or testicular masses. In others, it may occur as maldescent or cryptorchidism, or discovered in relation to indirect inguinal hernia, hydrocele, varicocele, epididymitis, infertility, testicular malignancy or testicular torsion. The majority has triorchidism and the supernumerary testis is most frequently found on the left side. In the present report, a rare case of triorchidism was detected incidentally in connection with the usual patient care service of a teaching hospital. The supernumerary testis was found in the right scrotal sac. It is described here with review of literature.

Key words: Polyorchidism, Supernumerary testis, Three testes, congenital anomaly of testes/gonads.

Introduction:
Polyorchidism or incidence of more than two testes is a very rare anomaly that has been referred in both medical literature as well as mythology. In the mythological literature there are more fantasy than fact and they describe that the man with this condition is supposed to possess increased sexual power and virility. Again, triorchidism is the commonest variety of polyorchidism and the supernumerary testis is most commonly found on the left side (Leung 1988 Sep). Rifkin et al (1983) reviewed that the first reported case of extra testicle was established by Lane in 1895. Out of less than 100 reported cases in world journal, only a few cases have been reported in our country (Kulkarni et al, 1992; Singh and Sobti, 1988; Mewara et al, 1984). Here we report an incidentally detected peculiar case of unilateral polyorchidism which presented with a malignant growth in the external genitalia with normal spermatogenesis.

Cast Report:
A 64 year old man presented himself at North Bengal Medical College, Darjeeling, West Bengal, India for treatment of a small fungating mass on the penis. He was observed not to be cachectic. There was also no other obvious clinical finding. His routine haematological and biochemical parameters were within normal limits. Skiagram of the chest was found normal. The case was clinically diagnosed as a case of squamous cell carcinoma of penis without any metastasis.

As the patient had already completed the formalities of his reproductive function in his family, he was operated for complete amputation of the penis shaft along with the fungating mass followed by perineal urethroscopy and emasculation.

During operation, it was found that the fungating cauliflower growth involved the distal penile shaft. On opening the right scrotal sac, two testes were detected and they were attached to a single epididymis. The smaller one (measuring 1.5 cm ‘ 1cm) was found at a higher level than the larger one (measuring 2.5cm ‘ 1.5cm), which was more directly related to the epididymis from where the vas was found to arise. Each had independent testicular vessels. The single testis of the left side showed no commendable variation from the normal (measuring about 4 cm ‘ 2.5 cm). (Fig.1)

Histopathologically, the penile neoplasm revealed the picture of squamous cell carcinoma. Surprisingly all the three testes presented normal histological features of seminiferous tubules with evidence of normal spermatogenesis.

Discussion:
Polyorchidism is an uncommon congenital anomaly defined as the presence of more than two histologically proven testes. In this type of unusual abnormality of the genital tract, most patients with supernumerary testicles are asymptomatic and have painless groin or scrotal masses (Leung, 1988; Aragona et al, 1994). Sometimes, the primary accompanying disorders and/or anomalies include cryptorchidism, maldescended testis, infertility, indirect inguinal hernia, torsion, epididymitis, malignancy, hydrocele or varicocele (Mastroeni et al, 1997; Figler et al 1966; Ozok et al, 1992; Shah et al, 1992; Mianne et al, 1990; Gandia et al, 1987; Mewara et al, 1984 Steyaert et al, 1966). The majority have triorchidism and though the present case shows two testes in the right scrotal sac, the supernumerary testis is most frequently found on the left side (Leung, 1988; Haddock and Burns, 1987; Sujka et al, 1987). Most common associated anomalies (about 80%) are maldescent of the testis or cryptorchidism and indirect hernia. Rest are associated with torsion, Infertility, malignancy,
hydrocele, epididymitis or varicocele (Mastroeni et al 1997; Besner et al, 1996; Leung, 1988). In about 7% of supernumery testes, malignant degeneration and/or transformation occurs (Sujka et al, 1987).

Polyorchidism is usually identified during orchidopexy and repair of indirect inguinal hernia (Gracia et al, 1992; Shah et al, 1992). But high accuracy or preoperative ultrasonographic evaluation of scrotal masses differentiates this rare benign entity from more ominous abnormalities such as neoplastic involvement of the scrotal content and thus it prevents unnecessary surgical exploration of sonographically normal, uncomplicated and orthotopic supernumery testicle (Ghiacy, 1996; Thum, 1991; Tammela et al, 1989; Yoshida et al, 1989; Rifkin et al, 1983).

If at all surgery is done, testicular biopsy must be carried out to reveal histological pattern (Ozok et al, 1992), like our case. However, in children testicular preservation should not be based only on the sonographic findings, and surgical exploration is mandatory whenever the nature of the testicular mass remains unclear (Aragona et al, 1994).

Laparoscopy may allow identification of polyorchia, especially in case where the proximal testicle is intraabdominal. Inguinal exploration alone may result in failure to recognise a higher duplicated gonad (Besner et al, 1996).

Incidence of polyorchidism has been identified in the age group of 4 to 75 years (Singer et al, 1992) and bilateral polyorchidism has been noted only in 5 cases (Mastroeni et al, 1997; Sozer et al, 1989; Singh and Sobti, 1988; Baker et al, 1987; Snow et al, 1985). Some exceptional cases like, three homolateral testis (Gracia et al, 1992) and intra-abdominal polyorchidism (Burgers and Gearhart, 1988) were also reported. Holder (1925) described a patient with 5 testes-2 in each scrotal sac and 5th in a small scrotal pouch in the perineum. One case of intrascrotal supernumerary testicle along with hypospadiasis was reported by Zur Kenntnis (1983).

Normal spermatogenesis is usually absent in the majority of supernumery testis (Thum, 1991; Butz and Croushor, 1978). But, the case reported here shows normal histological picture as well as normal spermatogenesis in all the three testes.

The exact explanation for the production of polyorchidism is not known, although several theories have been proposed including-anomalous appropriation of cells, initial longitudinal duplication of genital ridge or transverse division of genital ridge (Mastroeni et al, 1997; Jorion et al, 1990).

Leung (1988) described the anatomical variations on the possible embryological basis (Fig-2).

Type-I : supernumerary testis lacks an epididymis or vas and has got no attachment to the usual testis. (Division of genital ridge only).

Type-II : the supernumerary testis drains into epididymis of usual testis and they share a common vas. (Division of genital ridge occurs in the region where the primordial gonads are attached to the metanephric ducts, although the mesonephros and metanephric ducts are not divided, i.e.-incomplete division).

Type-III : the supernumerary testis has its own epididymis and both epididymis of the ipsilateral testes draining into one vas. (Complete transverse division of mesonephros as well as genital ridge).

Type-IV : complete duplication of testes, epididymis and vas. (Vertical division of genital ridge and mesonephros).

According to Leung 1988, the Type-II is the commonest and Type-II & Type-III together account for more than 90% cases of polyorchidism.

Review of literature (Thum, 1991; Sujka et al, 1987) allows groups of anatomical variations-

1. The double testes of a side are connected with their independent epididymis, so also vas. It can be explained embryologically by complete splitting of not only the genital ridge but also the whole mesonephros for the wolffian duct along the dorso-ventral line of cleavage.

2. On the contrary, the double testis of a side are not connected with a single epididymis or vas. This anomaly is explained by horizontal splitting of genital ridge to form gonad. Similarly, Singer et al (1992) suggested an anatomical as well as functional classification of polyorchidism.

Type-I : supernumerary testes attached to the draining epididymis and vas deferens with reproductive potential (Leung Type-II, III & IV).

Type-II : testes with lack of such an attachment without having any reproductive potential (Leung Type-I).

Each of these two types are again subdivided into two groups, Gr.-A and Gr. B, depending upon their location in the scrotum (orthotopic) or outside the scrotal sac(ectopic) respectively. Combining this classification with a knowledge of potential complications, they proposed a management strategy.

The case presented here is characterised by
presence of two testicles of unequal size within the right scrotal sac, disposed in two different levels with single epididymis and vas, which can be explained by horizontal (transverse) splitting of genital ridge only. But this case deserves a special notice, because it was associated with normal spermatogenesis in all the three testes with no other anomaly or abnormality and polyorchidism was detected incidentally during the treatment of carcinoma of penis.

References:


Fig. 1. Cut section of the scrotum showing three testicles.

Fig. 2. Animation showing Leung’s Classification of polyorchidism.