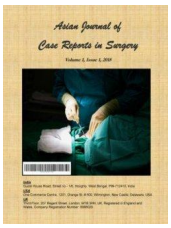

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Testicular Vanishing Syndrome – A Rare Clinical Entity

Mukhtar Mehboob[1*], Muhammad Samsoor Zarak[2] and Hamaiyl Sana[2]

[1]Mohtarma Shaheed Benazir Bhutto Hospital, Quetta, Pakistan.
[2]Bolan Medical College, Quetta, Pakistan.

Authors' contributions

This work was carried out in collaboration between all authors. Author MM wrote the protocol and wrote the first draft of the manuscript. Authors MSZ and HS managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Vanishing Testes Syndrome (VTS) also known as Testicular Regression Syndrome (TRS) is a condition in which there is absence or incomplete development of testis of varying degrees in 46XY subjects with normal external genitalia. This situation is observed in overall 5% of patients presented with cryptorchidism. We relate a case of VTS in an adult here, in which the subject had absent left testes on clinical examination, but MRI showed presence of atrophic testes at the level of deep inguinal ring. Orchidectomy of left testis was performed and the left testicular artery was followed which had a blind ending. The blind end was also resected and both of the specimens of the parenchyma of testicular tissues and blood vessel were sent for biopsy.

Keywords: Testicular vanishing syndrome.

1. INTRODUCTION

Vanishing Testes Syndrome (VTS) is a rare birth defect [1]. Various etiological conditions have been suggested but the exact cause of the condition is not known. Some believes it may be genetical. Recently, a novel heterozygous missense mutation (v355m) in *SF*1 gene is found in one boy with a micro penis and Testicular Regression Syndrome (TRS) [2]. VTS is found to

**Corresponding author: E-mail: mehboob87300@yahoo.com;*

occur between 12 and 14 weeks of gestation, when the testes are lost during the phase of male differentiation. The penis and scrotum are formed, that follow a normal pattern of development, but one or both testicles may be present [3] the unilateral absence of testis is usually observed in normal 46 XY male. VTS is more common than testicular agenesis in patients with non-palpable testis [4].

A case series of a time span of 8-year of non-palpable testes, was reported at the Children's Hospital of Philadelphia. The study showed that 41% of 447 affected boys (181 patients) had atrophic remnants or absent testis. It, therefore, seems that this syndrome is a common phenomenon among testicular abnormalities [5]. lOU and co-workers reported a series of patients with non-palpable testes, of which 64% had cryptorchidism. Of these, 22.5% had complete absence of testis, vas deferens and epididymis, 15% had only a blindly-ending vas deferens, and 67.5% had a blindly ending vas deferens with blood vessels in the inguinal canal [6].

The present case report is presented to highlight the unilateral presentation of VTS on left side and its association with hernia and infertility.

2. PRESENTATION OF CASE

A 25 year old man, presented in outpatient department with complaint of swelling in left inguinal region for last 10 years. He also had history of infertility since his married life of two and a half years. The general physical examination was unremarkable. The examination of left inguinal region revealed a reducible swelling, where impulse was noted on coughing. His left testes was absent in the left hemi-scrotum and not palpable in the inguinal region. His left side of scrotum was not developed, while a normal size right testis was placed inside the right hemi-scrotum.

The Color Doppler Ultrasound Study of the scrotum revealed that left testis was not visualized in any of its normal locations or even in an ectopic location but showed a normal size right testis with parenchymal echotexture. MRI of pelvis was performed, which indicated evidence of abnormal signals below the anterior abdominal wall at the level of deep inguinal ring on left side; and measured 1.2x0.8 cm. Semen analysis showed the total count and active form of sperm were reduced while the abnormal form was found to be higher than normal.

Preliminary investigations were performed. Detail discussion about the nature of disease and informed consent for surgery was taken. During surgery, left inguinal canal was explored and spermatic cord was identified. The deep inguinal ring was widened. Left testis was neither visualized nor palpated in abdomen deep in the inguinal canal. Left testicular artery was followed that had a blind end below the deep inguinal ring with a nodular swelling of 1.2 x 0.8 cm. Blind end was resected and sent for biopsy. The hernial sac arising from deep inguinal ring was dissected, ligated and excised. Herniorrhaphy was performed. The wound was closed anatomically. The recovery was smooth. The biopsy report revealed presence of a fibro vascular nodule with associated hemosiderin-laden macrophages and dystrophic calcification. Residual testicular tubules were also found.

3. DISCUSSION

Absent, non palpable testis with a blind ending spermatic cord in urological literature is referred to as the "vanishing testis syndrome" (VTS) while in pathological literature it is called as "Testicular Regression syndrome"(TRS) [7,8]. One of the characteristic features of TRS is the absence of testis, which might be unilateral or bilateral, with or without partially developed epididymis and spermatic cord structure in 46XY patient with normal external genitalia [9]. The pathological standards for the diagnosis are, first the presence of a calcified and vascularize fibrous nodule with or without hemosiderin and secondly a minimum of a vascularized fibrous nodules in the vicinity [9]. Most of the patients with TRS exhibit features of intersex or severe micro penis in association with complete unilateral or bilateral regression of testicular tissue. The degree of masculinization of the internal and external genitalia depends on the duration of testicular function, prior to its loss.

In 1969 Abeyaratne suggested a sequence of events that might be responsible for VTS. He proposed that a vascular catastrophe like torsion or vascular occlusion of the cord in intrauterine life during its descent might be a very like cause of VTS [10]. However no histopathological evidence of this vascular sequence was obtained. Another interesting cause presented by Honore in 1978 says that the unique pre dominance of absent testis on the left side most likely suggests a vascular cause [11]. This can be attributed to the anatomic arrangement of the left spermatic vein which drains into the left renal

vein along with the left kidney. The possibility of bending of the spermatic vein is real and could cause secondary obstruction with hemorrhagic infarction and/or ischemic atrophy. The above suggested reason seems to be the most likely cause in our case. The findings of hemosiderin deposits, dystrophic calcification and giant cells support these theories. Early intrauterine life presence of testis can be proved by the presence of spermatic cord structure. In our patient the characteristic findings on pathological examination favours this theory.

A better way to handle the case of non-palpable testis on physical examination is Laparoscopic examination, which is also favored by most surgeons. Inguinal exploration is also done in cases where spermatic vessels can be seen through the deep inguinal ring [12,13]. It is very much stressed upon to identify the entire vascular supply and lymph drainage of the testes due to the fact that testicular vein and Pampaniform plexus ought to be present for testis to be found there. Thus the presence of testicular veins demonstrate the presence of epididymis and vas deference, an indication for presence of testis [9].

Although in cases of cryptorchidism, surgery is needed as a secondary intervention to perform orchiedopexy in case of testis that are present but not palpable. But there is rather a disagreement whether to remove the remnant structure or leave it there in cases of VTS. The remnant structures were present in about 11% of cases presented and among these cases about 26% had cellular atypia [12,14]. In our patient we removed the nodule and surrounding tissues, which revealed no cellular atypia. A single case report was found regarding the malignant transformation of testicular remnants in VTS. Therefore considering the risks of malignancy the remnant should be removed. The malignant potential is found in testicular or para-testicular remnants that might have germ cells. Some authors have gone so far as to suggest that the contralateral testis should be fixed surgically so as to keep the said testis safe from torsion and by doing so ensuring the fertility of affected patients with VTS [15,16].

Some studies revealed adverse psychological effects in men or children with absent testis. As a solution for this testicular prosthesis implantation is considered to be a favorable option for the said subjects.

4. CONCLUSION

The early diagnosis of VTS is necessary to prevent the psychological trauma while surgery reduces the risk of malignancy in future.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard written ethical permission has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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