TRANSVERSE TESTICULAR ECTOPIA WITH PERSISTENT MULLERIAN DUCT SYNDROME: IS IT SO RARE?

MUHAMMAD SHAHAB ATHAR, NANDLAL KELLA*, JAVED AHMAD, MUHAMMAD TALAT MEHMOOD, MUHAMMAD SAJJAD ASHRAF, SHERA MOTI, NAIMA RASOOL**

Department of Paediatric Surgery, Dow University of Health Sciences & Civil Hospital, Karachi
*Department of Paediatric Surgery, Liaquat University of Medical & Health Sciences, Jamshoro**
*Department of Surgery, Dow University of Health Sciences & Lyari General Hospital, Karachi***

ABSTRACT

Objective: *To review all cases of Transverse testicular ectopia (TTE) associated with persistent mullerian duct syndrome (PMDS).*

Study Design: *Case Series study.*

Setting & Duration: *Department of Paediatric Surgery, Civil Hospital Karachi, SU VII Lyari General Hospital and Liaquat University of Medical Sciences, Hyderabad from 2001 to 2008.*

Methodology: *All patients diagnosed as transverse testicular ectopia (TTE) associated with or without persistent Mullerian duct syndrome (PMDS), who were who were managed in the Department of Paediatric Surgery of above mentioned centers were included.*

Results: *A total of 13 patients of transverse testicular ectopia (TTE) were managed. Out of them, 12 had associated persistent mullerian duct syndrome (PMDS). Mean age was 4.53 years. 10 patients presented with inguinal hernia of one side with non palpable undescended testis (UT) of opposite side, while one had bilateral inguinal hernia with left non palpable undescended testis, and another as obstructed right inguinal hernia and left non palpable UT. One patient presented with left non palpable undescended testis found to have both testes on right side. Gonadal biopsies were taken in 12 cases. No instance of Mixed Gonadal Dysgenesis was recognized in any of the cases. In all but one case it was possible to fix the ectopic testis trans-septally in the respective side of scrotum.*

Conclusion: *This is the largest series reported with 12 cases of TTE with PMDS and one case of TTE alone.*

KEY WORDS: *Undescended Testis, Testicular Ectopia, Crossed Ectopia, Persistant Mullarian Duct Syndrome*

INTRODUCTION

Transverse testicular ectopia (TTE) is a rare form of testicular ectopia. In this condition, the two testes are located on one inguinal side.[1] TTE has been classified as.[2]

Type I - Accompanied only by hernia;
Type II - Accompanied by persistent mullerian duct

Correspondence:
Dr. Muhammad Shahab Athar, Assistant Professor,
Department of Paediatric Surgery,
Dow University of Health Sciences, Karachi.
Phone: 021-9215740.
E-mail: drshahab@yahoo.com

Type III - Associated with disorders other than persistent mullerian remnants.

The condition classically presents as inguinal hernia which is mostly right sided with contralateral cryptorchidism. Most of the cases are discovered incidentally on groin exploration for inguinal hernia.[3] In most of these cases the patient is otherwise a normal male with a 46, XY karyotype.

The first case of TTE with PMDS was reported in May 2002 as a rare case by this team.[4] Since then several cases have been managed at the two centers, and some cases have been reported by other centers in the country as case reports.[5,6, 7,8] This article is written so as to describe in an detail all cases that we managed over a period of 8 years. It will provide great insight into various aspects of the anomaly.

METHODOLOGY

All patients with transverse testicular ectopia associated with or without persistent Mullerian duct syndrome, managed at the Department of Paediatric Surgery Civil Hospital Karachi during 2001 to 2008, SU VII Lyari General Hospital. Karachi between 2007 and 2008, Department of Paediatric Surgery, Liaquat University of Medical Sciences Hyderabad during 2005 to 2008 were included in this series. We have also included 3 cases previously published by us as case reports. No case has been excluded. Data regarding the age, presentation, and operative findings the surgical procedure performed was recorded.

RESULTS

Total 13 patients of transverse testicular ectopia were managed at the two centers. Out of them, 12 had associated persistent Mullerian duct structures (Type II), while one patient, a 6 month old infant presented with left Un-descended Testis without inguinal hernia (Type I), On examination both testes were found in right hemiscrotum (right sided crossed testicular ectopia). Ultrasound examination did not reveal PMDS or any other abnormality. Patient was not explored, so it can not be confirmed whether he has associated PMDS or not and was lost to follow up.

Age range was 2 months to 15 years mean age was 4.53 years. Nine cases presented with right inguinal hernia and left non palpable undescended testis, 2 cases with left Inguinal hernia and right non palpable undescended testis, one case with bilateral inguinal hernia and left non palpable undescended testis, and one case presented with left undescended testis and both testes were in right hemiscrotum. Two patients were related (Uncle and nephew).

Testicular biopsy was done in 12 cases, and no case of mixed gonadal dysgenesis was found. Ultrasound was done in all cases to locate the non palpable testis and it failed to visualize the testis and PMDS structures. It was possible to fix the ectopic testis trans-septally in the respective side of scrotum in 10 cases, while in one case orchidectomy was performed due to atrophic testis.

One case did not need surgery as he had both testes in right scrotum without hernia, In a 2 month old baby with obstructed hernia and advanced intestinal obstruction the left testis could not be pulled down to the scrotum, and condition of the patient did not allow abdominal exploration.

In 10 cases Mullerian remnants were left as such while in one case splitting of the rudimentary uterus was done. and in another case Mullerian structures were excised. Laparoscopy was done in two cases to locate non palpable testis, in the first case the Mullarian structures could not be recognized, and these were found on exploration during second stage orchidopexy, In the second case PMDS structures were identified at laparoscopy.

DISCUSSION

Transverse testicular ectopia (TTE) with Persistent Mullerian Duct Syndrome (PMDS) is a rare condition. This was first reported by von Lenhossek.[9] The first case published in English literature was reported in 1907 by Halstead[10] Few cases of TTE with out PMDS have also been reported.[11]

Until our first case report in 2002[4], there were only 5 cases reported from the country[12,13,14] and about 11 cases were reported in the English language literature. Since than some more cases have been reported locally, and a total of 116 cases have been reported in Pub Med up to September 2008.

PMDS is thought to result from the failure of synthesis, release or a receptor defect of Mullerian inhibiting factor (MIF). It is likely that the mechanical effect of the persistent mullerian duct structures produces cryptorchidism by preventing normal testicular descent producing TTE.[15]

The condition classically presents as inguinal hernia which is mostly right sided with contralateral cryptorchidism. Nine of the cases had right inguinal hernia, two had left inguinal hernia, one patient had a bilateral inguinal hernia. Most of these cases are discovered incidentally on groin exploration for inguinal hernia. Twelve cases were discovered in this way although suspicion of TTE and PMDS was there in some of the cases.

Other associated abnormalities that have been reported include True hermaphroditism, hypospadias, pseudohermaphroditism, and scrotal anomalies. In 2% to 97% of patients with crossed testicular ectopia, disorders of the upper and lower urinary tract have been reported.[16] These or other abnormalities were not detected in any of the cases in this study.

Twelve of the cases belonged to type II, and one presented as TTE without hernia. If crossed testicular ectopia is suspected, ultrasonography and subsequent magnetic resonance imaging have been recommended for diagnosis.[17,18]

In this series Ultrasound examination failed to diagnose

TTE and PMDS. In clinically suspected of TTE, ultrasonography may be able to identify the ectopic testis on contralateral side.

It is recommended that the diagnosis of PMDS must be confirmed with testicular biopsies and chromosomal studies. The PMDS must be distinguished from mixed gonadal dysgenesis in which the mullerian structures are generally present, a testis is present unilaterally and there is a contralateral streak gonad. These patients usually show chromosome mosaicism of XO/XY. In contrast, TTE and PMDS is characterized by a normal 46 XY karyotype and normal masculinization of external genitalia.[19]

Testicular biopsies in 12 of the cases did not show any case of mixed gonadal dysgenesis, although some atrophic changes were found in few cases while one testis was found to be completely atrophic and another testicular biopsy showed tuberculosis.[6]

There is near consensus that surgical treatment should include bilateral orchidopexy through the same or separate inguinal canals, former is usually feasible in the majority of cases. In the past excision of Mullerian structures was recommended with orchiopexy or orchidectomy, this is no longer recommended because there is no report of malignancy arising from the retained Mullerian structures, and the removal of the uterus may damage the vasa deferentia, which are in close proximity.[20] Hysterectomy is indicated only when mullerian structures limit intra-scrotal placement of the testes.[21] Alternatively the rudimentary uterus may be divided in midline to allow placement of the two testes in scrotum.[22] In this series excision of PMDS structures was performed in one case to facilitate scrotal placement of testes, in another case rudimentary uterus was split in the middle for the same reason.

Successful laparoscopic assisted orchidopexy has been reported in TTE with PMDS.[23] In this series laparoscopy was done in two cases to locate non palpable testis, in the first case the Mullerian structures could not be recognized, and these were found on exploration for second stage orchidopexy, In the second case the PMDS structures we were identified.

CONCLUSION

This is the largest series reported with 12 cases of TTE with PMDS and one case of TTE alone.

REFERENCES

1. Karnak I, Tanyel F C, Akren Z. Transverse testicular ectopia with persistent Mullerian duct syndrome. Journ Pediatr Surg 1997; 32: 1362-1364.

2. Loeff D S, Imbeaud S, Reyes H M. Surgical and genetic aspects of persistent Mullerian duct syndrome. Journ Pediatr Surg 1994; 29: 61-65.

3. Lam W W, Le S D, Chan K L, Chan F L. Transverse testicular ectopia detected by MR imaging and MR venography. Pediatr Radiol 2002; 32: 126-9.

4. Mehmood M T, Ahmad J, Athar S M. Transverse Testicular Ectopia and persistent Mullerian Duct Syndrome: A rare association Journ Coll Physicians Surg Pak 2002; 12(5): 320-21.

5. Kella N, Rathi P K, Memon S, Ahuja P, Memon A B S. Persistent Mullerian Duct syndrome with Transverse Testicular Ectopia Journ Coll Physicians Surg Pak 2005; 15(2): 119-20.

6. Kella N, Rathi P K, Qureshi M A. Tuberculosis of transverse testicular ectopic testis associated with persistent mullerian duct syndrome. J Coll Physicians Surg Pak 2008;18(4): 250-51.

7. Shamim M. Persistent Mullerian duct syndrome with transverse testicular ectopia presenting in an irreducible recurrent inguinal hernia. J Pak Med Assoc 2007; 57(8): 421-23.

8. Shamsuddin S, Mirza T H, Khan N, Ibrahim M, Imtiaz T. Transverse testicular ectopia J Ayub Med Coll Abottabad 2007; 19(1): 56-7.

9. Von Lenhossek M N. Ectopia testis transversa. Anta ANZ 1886; 1: 376.

10. Halstead A E. Ectopia testis transversa. Surg Gynecol Obster 1907; 4: 129-32.

11. Leung A K, Wong A L, Kao C P. Duplication of the testis with contralateral anorchism. South Med Journ 2003; 96(8): 809-10.

12. Mughal S A, Magan U. Persistent Mullerian Duct Syndrome with Transverse Testicular Ectopla in Two Brothers. Pak J Paed Surg 1999; 1-2: 25-31.

13. Mandhan P, Hussain I, Naqvi R, Ahmed J, Ali M A, Aziz A. Persistent mullerian duct syndrome report of two boys with associated transverse testicular ectopia. JPMA 1999; 49: 74-6.

14. Jan I A, Anwar-ul-Haq, Aziz S. Transverse testicular

ectopia with persistent mullerian duct syndrome. Journ Coll Phys Surg Pak 1998; 8: 239-40.

15. Thompson S T, Grillis M A, Wolkoff L H. Transverse testicular ectopia in a man with persistent Mullerian duct syndrome. Arch Pathol Lab Med 1994; 118: 752-755.

16. Tolete-Velcek F, Bernstein M O, Hansbrough F. Crossed testicular ectopia with bilateral duplication of the vasa deferentia: an unusual finding in cryptorchism. Journ Pediatr Surg 1988; 23: 641-3.

17. Nam Y S, Baik H K, Kim S J, Lee H K, Park H K. Transverse testicular ectopia found by preoperative ultrasonography. Journ Korean Med Sci 1998; 13(3): 328-30.

18. Lam W W, Le S D, Chan K L, Chan F L. Transverse testicular ectopia detected by MR imaging and MR venography. Pediatr Radiol 2002; 32: 126-9.

19. Martin E L, Bennett A H, Cromie W J. Persistent mullerian duct syndrome with transverse testicular ectopia and spermatogenesis. Journ Urol 1992; 147: 1615-1617.

20. Vandersteen D R, Chaumeton A K, Ireland K, Tank E S. Surgical management of persistent mullerian duct syndrome. Urology 1997; 49: 941-5.

21. Buchholz N P, Biyabani R, Herzig M J U. Persistent mullerian duct syndrome. Eur Urol 1998; 34: 230-232.

22. Pappis C, Constantinides C, Chiotis D, Dacov Vouletakis C. Persistent mullerian duct structures in cryptorchid male infant: surgical dilemmas. Journ Pedia Surg 1979; 14: 128-31.

23. Dean G E, Shah S K. Laparoscopically assisted correction of transverse testicular ectopia. Journ Urol 2002; 167: 1817.