True Hermaphroditism: Review Article

I Wani

Citation

I Wani. *True Hermaphroditism: Review Article*. The Internet Journal of Pediatrics and Neonatology. 2007 Volume 9 Number 1.

Abstract

True hermaphroditism is a rare condition . This is a form of ambigious genitilia where there is presence of both ovarian and testicular tissue in a same individual.The author is presenting a brief review of true hermaphroditism with emphasis on clinical perspective.

INTRODUCTION

True hermaphroditism is a characteristic intersex disorder in which a gender possess both testicular and gonadal tissue[1]. The presence of both müllerian and wolffian structures in these patients has been explained by the inappropriate timing of adequate mullerian-inhibiting factor effect, as well as insufficient synthesis of testosterone by Leydig cells[2]. Several variables are decisive in management of true hermaphrodites. Factors kept in consideration at this juncture are age at the time of diagnosis, location of gonads, their histology, the phenotypic and functional aspects of the external genitalia.

GENERAL DISCUSSION

The term 'Hermaphrodite is derived from the Greek mythological God "Hermaphroditos" ,son of Hermes and Aphrodite ,whose body after being merged with nymph Salmakis assumed a more perfect form with the both male and female attributes [3] . True hermaphrodtism is one of the inclusion in the spectrum of intersex disorders. This disorder is a manifestation of discordance between genetic, gonadal and phenotypic sex. Other catogeries in this spectrum are masculinised genetic females and gonadal dysgenetics

PRESENTATION

Regarding appearance of external genitalia , the gender may posses genitalia ranging from normal male to normal female [4] .In many of these ,there are ambiguous genitalia manifesting phallic chordee , hypospadiasis and cryptorchidism [5,6] The most common congenital anamoly present with the true hermaphrodites is hypospadiasis, associated often with bifid scrotum. Hypospadiasis is commonly seen in the phenotypic males .In 90 % of true hermaphrodites there is a breast development. Vicarious menstruation in the form cylical hematuria due to the presence of endometrium in the urinary tract [7,8] .Inguinal hernia is present in 50% of all true hermaphrodites [9] .Presence of an intersex should be suspected in gender with ambiguous genitalia or with severe hypospadiasisis without a two palpable gonads [10] .Ovatestes is the most common gonad, followed by the ovary and testis is last of all.Development of the ipsilateral ductal system is consistent with that of gonad.Uterine horn is completely absent on the side of functional testis. The fallopian tube usually develops normally in continuity to a functional ovary. The normal looking appearance of external genitalia in true hermaphroditism attributes to overlooking of diagnosis at birth in many cases. Further misery in late diagnosed cases is added at puberty by manifesting opposite secondary characters, ammenohroea in pts raised as girls or breast development in pt, raised as boys [11,5]

Theses are classified on the basis of histology and location of gonads:

Lateral –Testis and contralateral ovary(30%);

Bilateral: Testicular and ovarian tissue identified on both sides ,usually as ovatestis (20%);

Unilateral: Ovatestes on one side and testis or ovary on other side (50%) [9]

The distribution of ovarion and testicular within ovatestes varies from case to case with one moiety predominating and other assuming a polar or hilar distribution [4] .It is recommended that gonadal biopsies for confirmation of diagnosis taken from pole to pole with tissue chunk

preferably from deep into the hilum of gonad [12,4] .

MALIGNANT CHANGES

Malignant tumors occurs in 1.91% of true hermaphrodite commonly in patients with a 46XY karyotype , in dysgenetic and undescended testes [4] . ,In male hermaphrodite there is increased incidence seminomas, gonadoblastoma and teratomas [13] . This malignant degeneration usually occurs in true hermaphrodites in phenotypic males and potentially in mosaics 1,11 and 17

REARING OF HERMAPHRODITE

A high index of suspicion is necessary is mandatory to lessen stigma of late diagnosis .True hermaphroditism manifests in adulthood if non diagnosed in infancy [4] .Gender assessment is considered an increasingly controversial subject [14,15] .After confirmation of diagnosis relevant multidisciplinary team of specialist assign sex rearing of child with guardian consultation on the basis of genitalia , gonadal and genetic factors. The choice of rearing hermaphrodite as male or female sex is governed by phallic size..Phallic size of more than 1.5 cm. and its inherent potential of acquiring sexually mature characteristics is considered the single most important factor in deciding the choice of male sex of rearing [16,4] . To rear hermaphrodite as female sex, likely choosing factor is phallic length of less than 1.5 cm., the benefit of small phallus favors an adequate and functional vagina. Gonadal factors are also important in the choice of rearing with adequate functional should be present to sustain the sex of rearing and gonads producing contraindicatory hormones should be excised [5] .Assigning gender for rearing in hermaphrodite with parents consultation and opinion of specialist is called “paternalistic approach” [17] . This approach is based on the assumption that children are gender neutral at birth and can be molded to male or female characteristics by psychosocial and ‘normalizing surgery' in infancy [18] .In the intrauterine life, genetic and endocrinal influences on the brain are proposed in the genesis of development of hermaphrodite compared to pseudohermaphrodites. There is not complete lack of germinal epithelium in true hermaphrodites accounting for ovulation and the spermatogenesis which has been reported only in 12% of cases [19] .Phenotypic males with microphallus may have three month trial of intramuscular enanthate to get aware of phallic growth potential [20] .Drawback with androgen therapy n childhood is that it delays only not prevents the development of psychosocial problems associated with microphallus.

References

1. Donahoe PK, Crawford JD, Hendren WH. True hermaphroditism: a clinical description and a proposed function for the long arm of the Y chromosome. J Pediatr Surg 1978; 13: 293-301.
2. Williams C, Hughes IA. Unusual dual genital duct remnants in true hermaphroditism. J Med Genet 1988;25:206-8.
3. Fallat ME. Ambiguous genitalia and intersex anomalies. In: Ziegler MM, Azizkhan RG, Weber TR. ed. Operative Paediatric Surgery. New York: McGraw-Hill Companies; 2003: 839-61.
4. van Niekerk, W.A.: True hermaphroditism: An analytic review with a report of 3 new cases. Am. J. Obstet. Gynecol., 126: 890 905, 1976.
5. Olsson CA, Tessier PA, Brown ML, Austen G Jr. True hermaphroditism. J Urol 1971; 105: 586-90.
6. Luks FI, Hansbrough F, Klotz DH, Kottmeier PK, Tolete-Velcek F. Early gender assignment in true hermaphroditism. J Pediatr Surg 1988; 23: 1122-6.
7. Federman,D.D.: Abnormal Sexual Development. A Genetic and Endocrine Approach to Differential Diagnosis. Philadelphia: W.B. Saunders Co., pp. 5866, 1967.
8. Raspa, R.W., Subramaniam, A.P., Romas, N.A.: True hermaphroditism presenting as intermittent hematuria and groin pain. Urology, 28 (2): 133 136, 1986.
9. Grumbach, M.M., Conte, F.A.: Disorders of sex differentiation, in Williams, R.H. (ed): Textbook of Endocrinology, Philadelphia, W.B. Saunders, Co., ed 6, chapt. 9, p. 473,1981.
10. Salvatierra, O. , Skaist, L. B. , Morrow, J.W. : True hermaphroditism discovered 10 years after hypospadias repair: Report of 2 cases. J. Urol., 98: 111, 1967.
11. Wiersma R. Management of the African child with true herma-phroditism. J Pediatr Surg 2001; 36: 397-9.
12. Donahoe PK, Crawford JD, Hendren WH. True hermaphroditism: a clinical description and a proposed function for the long arm of the Y chromosome. J Pediatr Surg 1978; 13: 293-301
13. Jones, H.W., Scott, W.W.: Male hermaphroditism (non familial masculinizing). In: Hermaphroditism, Genital Anomalies and Related Endocrine Disorders, 2nd edition. Baltimore, Williams 8 Wilkins Co., p. 97,1971.
14. Nihoul-Fékété C. The Isabel Forshall Lecture. Surgical management of the intersex patient: an overview in 2003. J Pediatr Surg 2004; 39: 144-5.
15. Reiner WG. Sex assignment in the neonate with intersex or inadequate genitalia Arch Pediatr Adolesc Med 1997; 151: 1044-5.
16. Luks FI, Hansbrough F, Klotz DH, Kottmeier PK, Tolete-Velcek F. Early gender assignment in true hermaphroditism. J Pediatr Surg 1988; 23: 1122-6.
17. Blizzard RM. Intersex issues: A series of continuing Conundrums. Pediatrics 2002; 110: 616-21.
18. Dreger AD. "Ambiguous sex" - or ambivalent medicine? Ethical issues in the medical treatment of intersexuality. Hastings Cent Rep 1998; 28: 24-35.
19. van Niekerk, W.A.: True Hermaphroditism. In: The Intersex Child. Pediatric and Adolescent Endocrinology. Edited by N. Josso. New York: S. Karger, vol. 8, p. 80,1981
20. Money, J., Hampson, J.G., Hampson, J.L.: Hermaphroditism: Recommendations concerning assignment of sex, change of sex, and psychologic management. Bull. Johns Hopkins Hosp., 97: 284, 1955.

Author Information

Imtiaz Ahmed Wani
Post-Graduate Scholar, Post Graduate Department of Surgery, S.M.H.S Hospital