International Journal of Current Research and Review
DOI: 10.7324/IJCRR.2018.1027

Case Report

IJCRR
Section: Healthcare Sci. Journal
Impact Factor 4.016
ICV: 71.54

Penile Agenesis- An Extremely Rare Urogenital Anomaly

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ABSTRACT

Aim: Penile agenesis, defined as aphallia, is an extremely rare congenital anomaly. Though this condition is diagnosed clinically, it may be associated with various ano-rectal and systemic malformations. Until date, very few cases of aphallia have been reported globally. The most important dilemma in such cases lies in the treatment associated with gender reassignment.

Case Report: We report a rare of case aphallia as a solitary presentation, in a neonate born in a rural hospital of West Bengal and the dilemma imposed on the parents regarding the management.

Discussion and Conclusion: The confusion regarding the surgical intervention associated with the psyco-sexual influence is a major hindrance.

Key Words: Aphallia, Congenital anomaly, Ano-rectal

INTRODUCTION

Penile agenesis is a very rare genitourinary malformation noticed in 1 in 30 million live births.[1]Aphallia occurs from the absence or failure in development of genital tubercle. Aphallia was first reported by French Surgeon, Saviard in 1701 as "Child who had no rod", but the detailed description was reported by Imminger in 1853. Literature review suggests that till date 100 cases have been reported.[2] Moreover, Penile agenesis may be solitary or associated with wide array of malformations like genitourinary, gastrointestinal tract and developmental defects of caudal axis.[3] In the present instance a solitary malformation of penile agenesis in a genotypic male new-born and the dilemma associated with the treatment is described.

CASE REPORT

A full term new born was delivered in a gynaecology and obstetric department of a rural hospital of West Bengal by emergency caesarean section. The baby was born of non-consanguineous marriage and the antenatal history of the mother was uneventful and uncomplicated. The birth weight of the child was 3.5 kg .On general examination of the child, there was no penis. The scrotum was well developed with bilateral normally descended testis. Urethral opening was noticed in the anterior aspect of scrotum. (Figure 1) No other external malformations were noticed. Ultrasonography of whole abdomen revealed no renal abnormality and no female internal genitalia was observed. Karyotyping workup confirmed that the baby was 46XY. No fistula or aberration of urethral tract was noticed in Cystourethrogram. The baby was diagnosed as a case of aphallia and the treatment of reconstruction surgery were discussed with the parent. The parent were counselled regarding the constraints of treatment in this rural set-up and transferred to a higher set-up for reconstruction surgery.

Figure 1: Showing urethral opening on the scrotum.

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ISSN: 2231-2196 (Print) **ISSN:** 0975-5241 (Online)

Received: 15.11.2017 **Revised:** 10.12.2017 **Accepted:** 30.12.2017

DISCUSSION

Aphallia develops as a result of failure of development of genital tubercle into phallus with incomplete separation of urogenital sinus from hindgut.[1] The urethral opening may be situated at sites like in the perineum, near pubis, anterior aspect of scrotum or most commonly just anterior to the anus and in the rectum. Penile agenesis associated with other malformations is much more prevalent (54%) than the solitary variety. In this instance we report a solitary variety of aphallia with urethral opening situated on the scrotum.[3]Aphallia must be differentiated from micropenis, rudimentary penis, penile amputation, concealed penis and pseudohermaphroditism.[4] The confusion lies in the surgical management of such patients. Genital reassignment surgery was the main stay of treatment consisting of bilateral orchiectomy, vaginal reconstruction, urethral transposition and labial reconstruction with adequate postoperative outcome. [5]Moreover, lifelong hormonal therapy initiated during puberty is an important part of treatment. Recently phallus reconstruction has also been successful in few such cases.[6]So, there is great dilemma between both the treatment modalities. The newer concept of gender imprinting in brain since in-utero and the psychosocial consequences of gender reassignment is an important perspective in such cases.[7] Thus the treatment should be individualised according to age of presentation, psychosocial adjustment of both patient and parents. The treatment is a multidisciplinary approach involving a paediatric suregeon, urologist, endocrinologist and psychologist.

Present case report is unique in this that the patient presented at birth and there are no other genitourinary malformations. The parents were counselled regarding both the surgical interventions and referred to a tertiary care centre. Moreover, recent advances in radiological techniques T2-Weighted MRI(Magnetic resonance imaging) can not only detect such anomalies at an early pregnancy but also ascertain the prognosis. But, it is not advisable in Indian scenario where the bias for male gender is still very high.

CONCLUSION

This case highlights that though penile agenesis is a rare urogenital finding yet the main dilemma lies in the intervention. Thus, treatment should be individualised depending on the age of presentation and psychosexual orientation of patient. Pre-surgical counselling remains an indispensible part of the treatment.

ACKNOWLEDGEMENT

Authors acknowledge the immense help received from the scholars whose articles are cited and included in references of this manuscript. The authors are also grateful to authors/ editors/publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

Conflict of interest and Funding: There is no conflict of interest and no funding from external agency.

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