Men in dilemma: The tale of male pseudohermaphroditism

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ABSTRACT

Introduction: The development of the reproductive system is rather complicated. The presence of the Y chromosome with sex-determining region Y protein (SRY) influence the mesonephric to evolve as a male phenotype. Otherwise, paramesonephric will continue growing and result as female phenotype. Rarely, failure of regression of paramesonephric duct may occur, despite of presence of. SRY protein, as seen in cases of Persistent Müllerian Duct Syndrome (PMDS). The genetic inheritance in PMDS is attributed to the mutations in MIS or the MIS receptor. Although the incidence of PMDS is very low globally, once diagnosed, it caters complex management in improving patient health and quality of life. **Case Description:** We present a case of a 32-year-old married man with four years of subfertility presenting with an apparent strangulated inguinal hernia later discovered to have PMDS. We embarked on a multidisciplinary team approach with combined surgery in an attempt to restore his quality of life, including his future fertility outcome. **Discussion:** In our case, as he underwent orchidopexy for bilateral testis and his Johnson score was three; therefore, there is a value to repeat TESE in this patient after six months. This recommendation was postulated as his testis are placed back in the scrotum, leading to proper temperature; the testicular tissue might regain its function to produce sperm. Microsurgical TESE would be the procedure of choice for this patient. However, his chances are slim and thorough counselling should be done before the procedure. If favourable amount of sperm managed to be harvested during the repeat TESE, sperm banking can be offered and ICSI would be the next step for the hope of a successful pregnancy.