Treating painful haematuria with hysterectomy in a male with congenital adrenal hyperplasia (CAH)

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ABSTRACT

Introduction: Congenital adrenal hyperplasia (CAH) is an autosomal recessive disorder which is mainly caused by deficiency of 21-hydroxylase enzyme. This enzyme deficiency causes accumulation of steroid precursors which will be shunted into the androgen synthesis pathway resulting in androgen excess. CAH causes genital virilization of genotypic XX female babies. The usual medical practice consists of hormone replacement therapy and corrective feminising genitoplasty surgery. However, some patients are lost to follow-up and raised as males. Few of these patients have troublesome symptoms severe enough to convince them to seek medical attention in later life. Case Presentation: A 19-year-old phenotypic male with 46XX CAH was referred to the gynaecology department due to recurrent severe suprapubic pain with haematuria. CAH was diagnosed during infancy but the patient defaulted follow-up due to socio-financial issues. Clinical phenotype was male and genital examination revealed severely virilized genitalia consistent with stage 4 on Prader Scale. Imaging studies confirmed the presence of uterus with low confluent persistent urogenital sinus (PUGS). After a series of multidisciplinary assessments, the patient decided to keep his sex allocation as a male. Total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAHBSO) was performed and the symptom of recurrent painful haematuria was cured. Gender affirming surgeries are planned later. Conclusion: This case is a rare case reported in the literature. This report highlights the management of XX CAH who presented late with recurrent painful haematuria and managed by hysterectomy.

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"No one left behind" – Incorporating HPV screening test into Orang Asli outreach program

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ABSTRACT

Introduction: One of the challenges in the implementation of WHO global strategy to eliminate cervical cancer is reaching the marginalized community for screening program. In Malaysia, Orang Asli community often have lack of access to social services and health coverage. Healthcare services including child immunization & antenatal care are covered by special unit under JKN of few states like Pahang, Kelantan & Perak with additional coverage run by mobile outreach team under NGOs. Objectives: As a pilot project, to assess the feasibility of incorporating HPV self-testing & mobile colposcopy as cervical screening program into mobile outreach for Orang Asli run by an NGO. Methods: Representatives from ROSE Foundation joined a regular Orang Asli outreach team run by IMAM Relief & Response Team (IMARET) in August 2020 to Pos Balar, Gua Musang. The team of 37 volunteers mobilized on ten 4x4 vehicles in a 5-day mission running a medical (OPD) clinic, dental clinic, health education activities with children and HPV screening using HPV self-test kit provided by ROSE Foundation, sponsored by ETIQA Care. Results: 20 eligible women attended the booth & performed the HPV self-testing without difficulty. 19 of them tested negative for HrHPV DNA & only 1 test was invalid. Among challenges encountered were false belief, incorrect age (inconsistent with Mykad), traditional health restriction and cultural differences. Conclusions: Despite the geographical and cultural challenges, HPV self-test among marginalized Orang Asli women is feasible. It can be implemented with usage of mobile colposcopy & cold coagulation for treatment of CIN.