46, XX Male or Ovotesticular DSD (SRY-negative) without SRY, Is It Possible to have Testicular Tissues?

Ndour D[1*], Faye PM[2], Sagna A[3], Gueye D[3], Gassama O[4], Signate AS[2] and Ndiaye O[2]

[1]Faculty of Medicine of Pharmacy and Odontology, Cheikh Anta Diop University, Senegal

[2]Service Neonatology Children's Hospital, Albert Royer, Senegal

[3]Service Hospital Surgery, Albert Royer, Senegal

[4]Hospital Gynecology and Obstetrics Service Aristide Le Dantec, Senegal

[*]**Corresponding author**: Ndour D, Faculty of Medicine of Pharmacy and Odontology, Cheikh Anta Diop University, Senegal, Tel: 00221775121402; E-mail: daoudandour@hotmail.com

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Abstract

The 46, XX male syndrome, or the 46, XX testicular Disorder of Sexual Differentiation (DSD) is rare. We report a clinical case of a male infant 46, XX, DSD with SRY negative. This observation suggests that genes other than SRY play an important role in determining sex. From this case we will present a review of the literature to take stock of the subject.

Keywords: XX DSD; Karyotype; SRY-negative

Introduction

foetal sex depends on a number of cellular and hormonal signals that interact in a order but still poorly to contribute to the establishment of the genital apparatus and a male or female phenotype [1].

Chronologically, it takes place in four major sequential steps:

- Establishment of the genetic sex at the time of fertilization.
- combination of a maternal oocyte (23, X) and a paternal spermatozoon, either (23, X) or (23, Y) will give a homogametic egg (46, XX) of female genetic sex or heterogametic (46, XY) of male genetic sex.
- establishment of the gonadal sex which represents the pathway of the gonads. It encompasses the determination and subsequent of the bipotential foetal gonad into testis or ovary. the internal structures will be set up from two parallel channel systems present in the embryo: the and Müller ducts.
- hormonal production by the testis of two capital hormones for the virilisation of the male foetus: the anti-Müllerian hormone (AMH) and the testosterone.
- AMH will allow the regression of the Müller ducts while the testosterone is responsible for the development of the ducts.
- development of phenotypic sex results in of the external genital organs and the urogenital sinus.

last stage of sexual

Pregnancy was complicated by pre-eclampsia requiring hospitalization for surveillance. child was born through spontaneous eutocic vaginal delivery to 35 weeks of amenorrhea and 4 days; with an Apgar score of 7 then 9 respectively at 1 and 5 minutes of life.

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Figure 2: genital bulges had a scrotum-like appearance in which gonad-like formations were found when palpated.

neonatologists, surgeons, geneticists and biologists who can improve management while insisting on the role of psychological support to families.

From a therapeutic point of view, our patient is expected to have a correction of the hypospad, and long-term monitoring to detect puberty disorders, appearance of gynecomastia at puberty, but also, and above all, impairment of fertility.

Conclusion

Although abnormalities of sexual development with testicles associated with a karyotype 46, XX are rare, but this clinical case illustrates all their interest for the clinician.

From a practical point of view, the approach to etiologic diagnosis