

Correspondence: Persistent Mullerian Duct Syndrome with Ovarian Endometriosis

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Dear Editor,

With much interest, we have read the study of Nerune SM et al., "Persistent Mullerian Canal syndrome with ovarian endometriosis-a rare case" [1]. We appreciate the authors' attempt to emphasise the Persistent Mullerian Duct Syndrome (PMDS) as a disorder of sexual development, but the evidence of the clinical case was in favour of ovotestis.

In the present study, the patient was 19 years old with a male phenotype and a left painful scrotum, the right testicle and external genital organs were normal. The general physical examination revealed the presence of bilateral gynecomastia. Radiological and surgical exploration was done for the pelvic solid-cystic mass. Nonetheless, the anatomo-pathological study revealed the coexistence of a testicular and ovarian tissue knowing that the patient did not benefit from a karyotype. In fact, the sum of these exams is in favour of an ovotestis and far from being a PMDS.

A Persistence of Mullerian Duct Syndrome (PMDS) is defined as an internal pseudo-hermaphroditism characterised by the presence of a uterus, fallopian tubes and the upper 1/3rd of the vagina in a normally virilized patient with karyotype 46, XY [2].

Three types of PMDS have been identified [3,4]:

- Type 1 (60-70%) female type: bilateral cryptorchidism with two bilateral intra-abdominal testicles position similar to the ovaries.
- Type 2 (20-30%) male type: presence of a testicle, uterus and tubes in the same hernia sac (hernia inguinal uterus).
- Type 3 (10%) male type: both testicles are located in the same hernial sac with Mullerian structures (transverse testicular ectopia or cross-testicular ectopia).

The pathophysiology of PMDS is characterised by either an insufficient amount of Mullerian Inhibitory Factor (MIF) or an insensitivity of the target organ to MIF [5].

Ovotestis, which is a true hermaphroditism, is characterised by the simultaneous presence, in an individual, of testicular tissue and ovarian tissue. This type of disorder in sexual development can be classified in relation to the localisation of the gonads [6,7]:

- Lateral ovotestis (20%) is characterised by the presence of a testicle on one side and the ovary on the other side.
- Bilateral ovotestis (3%) with a mixed gonad on both sides.
- Unilateral ovotestis (50%) with a mixed gonad on one side and a normal one on the other side (ovary or testicle).

The discussed clinical case is characteristic of an ovotestis, of the type "lateral ovotestis"

In conclusion, this case report has been deprived of scientific value due to its false definition of the development disorder which must be considered as an ovotestis and not a persistent of Mullerian duct syndrome.

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