Unicornuate uterus with two cavitated, non-communicating rudimentary horns?

Dear Sir,

I read with interest the publication of Nezhat and Smith (1999) and believe they were misinterpreting both the clinical case they reported, and the embryology of the female genital tract. At that time, I did not reply nor make any comment. However, I recently read another Letter to the Editor by Nisolle and Donnez (2000) who also believe that the interpretation by Nezhat and Smith is not correct.

Nisolle and Donnez suggest that it might be a variant of a double uterus with blind hemivagina but they wonder: ‘Was it not possible to perform drainage of the collection through the vagina trying to preserve the blind uterus?’ In their reply, Nezhat and Smith (2000) point out again the singularity of the case, that there were two endometrial cavities in the right side, and that there was no vaginal cavity, recurring once more for its explanation to a Mullerian embryological hypothesis which I also think is incorrect. Since from my point of view, neither Nisolle and Donnez’s explanation, nor that of the former authors is totally correct; nor do the latter mention other similar cases reported by us (Acién, 1986; Acién et al, 1987, 1988, 1991); plus, there is no reference to our embryological observations on the female genital tract and out embryological hypothesis about the human vagina (Acién, 1992). Therefore, I would like to analyse the case reported by Nezhat and Smith and suggest an embryological explanation.

From my point of view, this case could be clearly included in group II of our embryological classification of female genital malformations and it is likely to be a double or didelphys uterus which in its right hand side had cervical
atresia of the corresponding hemiuterus and below, a blind hemivagina. All this would explain the right renal agenesis. I enclose a rough sketch of how the anatomy of this patient’s genital tract would then appear (Figure 1A).

We have pointed out that: (i) renal agenesis is systematically associated with either agenesis of all the derivatives of the urogenital ridge, or with a unilateral blind hemivagina (with haematocolpos or with atretic blind vagina of the Gardnerian type); (ii) the vagina must be derived both from the mesonephric or Wolffian ducts (from which also derives the ureteral bud) and from the Müllerian tubercle; (iii) the vagina is first lined by cells of the Müllerian tubercle (the epithelium that lines the blind hemivagina is often a Müllerian type) and later, by metaphasic induction or more probably by epidermization from the urogenital sinus, the vagina becomes lined by a squamous epithelium; and (iv) the proper development of the Müllerian ducts depends upon the ‘inductor’ function of the mesonephric or Wolffian ducts.

Therefore, an agenesis or an early and extensive injury of the right mesonephric duct could properly explain this patient’s malformation: (i) absence or failure in the fusion of the Mullerian ducts; (ii) misdevelopment of the right Mullerian duct (cervical atresia or stricture in its middle part); and (iii) misdevelopment of the lower segment of the mesonephic duct and of its opening in the urogenital sinus (atresia or blind hemivagina and renal agenesis).

Another possibility (although less likely), to explain the anatomy of this patient’s malformation is shown in Figure 1B, which more closely resembles that of the original publication. In this case there would only be right vaginal rudiment, atresia or atretic hemivagina. However, the embryological origin would be the same: agenesis or early and extensive lesion (from above) of the right mesonephric or Wolffian duct. It also explains the anomalous location and vascular supply of the ovary. Figure 2 shows how (in my opinion) the embryological development of the malformation has taken place in this case.

Complex malformations of the female genital tract due to mesonephric anomalies (with absence of an opening in the urogenital sinus and of the formation of the corresponding ureteral bud) are not rare, and if we consider them taking into account the embryological hypothesis above pointed out, many inappropriate surgical operations and reoperations could be avoided. Shortly, we will operate on an 18 year old patient with strong dysmenorrhea, who had a right adnexectomy performed because of endometrioma in a different hospital and province some months ago. She was also found to have a double uterus. We saw her after she came to the emergency service of our Hospital during her holidays presenting with strong dysmenorrhea and having haematometra of the right hemiuterus (as shown in transrectal ecography). She has already been diagnosed as having a didelphys uterus with cervico–vaginal atresia on the right handside and right renal agenesis, and we plan to perform a right hemihysterectomy, which in our opinion is the operation she should have undergone initially, since there is no permeable right cervix nor, therefore, right haematocolpos.

The presence of haematocolpos would be the situation in which it would only be necessary to perform a wide transvaginal opening in the haematocolpos (with resection of the intervaginal septum) without any other surgery.

The unicorunate uteri due to an isolated Mullerian anomaly (group IIIa of our classification), with rudimentary horns cavitated or not, and communicating or not, usually show such horns fused, and in these cases there is no atretic blind hemivagina nor therefore, ipsilateral renal agenesis.


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