Mullerian Anomaly Presenting as Chronic Pelvic Pain

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DEAR EDITOR,

Pelvic pain is a common gynaecological problem. A non-communicating rudimentary horn with a functional endometrial cavity may cause retrograde expulsion of menstrual debris and lead to endometriosis and infertility. We report a case of a bicornuate uterus with both non-communicating uterine cavities and dysgenesis at isthmic level and associated endometriosis who reported to us for chronic pelvic pain and had to undergo surgical removal of the horn and subsequently hysterectomy for resolution of symptoms.

A nulliparous thirty three year old female presented with chronic cyclical pelvic pain and a history of primary amenorrhoea and primary infertility. Pain was not alleviated by medical therapy and was interfering with her social life. Physical examination revealed an overweight woman (BMI- 28) with normal secondary sex characters. Per speculum examination revealed a normal looking cervix. Pelvic examination revealed anteverted, normal sized uterus. Investigations for amenorrhoea and infertility were done and reported normal. MRI showed uterus bicornis unicollis with hypoplastic right uterine horn and larger left endometrial cavity with well developed uterine musculature around it and apparently communicating with cervix .

With a provisional diagnosis of a bicornis unicollis uterus with rudimentary horn, hysterolaparoscopy was decided. Hysteroscopy was tried but access to main uterine cavity could not be attained due to suspected false passage or stricture at isthmic level. Laparoscopy revealed dense adhesions due to Stage IV endometriosis. Omentum was stuck to anterior surface of uterus and pouch of Douglas was obliterated. Adhesiolysis was done. A bicornuate uterus with a normal left horn and congested smaller right horn was seen. Both fallopian tubes were unhealthy looking, thick oedematous and congested and stuck behind uterus. The non-communicating right horn was considered to be the cause of the patient's severe dysmenorrhea due to intracavitary retention of menstrual effluent and retrograde menstrual flow to the tubes. A decision of laparotomy was taken followed by removal of right uterine horn along with removal of both tubes was done while preserving the other apparently well developed uterine cavity and ovaries. Since the communication could not be established between the well-developed uterine horn and normal looking cervix, she was later planned for repeat hysteroscopy and reconstruction of passage between cervix and uterus. Histopathology revealed a rudimentary horn with endometrial and myometrial tissue with functional but patchy endometrium. The tubes showed changes of endosalpingiosis with areas of hemosiderin-laden macrophages.

Patient was lost to follow up after the surgery and reported after 8 months with persisting cyclical pain. She was reinvestigated and a second look MRI was done. It suggested non-communication of the well-developed uterine cavity with cervix at isthmic level with collection in the uterine cavity. The pressure of the fluid in cavity caused keyhole appearance at cervical level.

Patient was taken for laparotomy after informed consent for hysterectomy. Pre-op hysteroscopy revealed well-formed normal looking cervix but blindly ending endocervical canal. Uterine cavity could not be visualised. On laparotomy, no communication could be established between uterine cavity and internal os. The diagnosis of non-communicating

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horn with dysgenesis at isthmic level was made and hysterectomy was done. Histopathology revealed patchy functional endometrium. Patient is being followed up in OPD and is now relieved of her pain.

The incidence and prevalence of mullerian duct anomalies varies widely. A prevalence of 0.4% has been reported in women who were investigated because of nonobstetric indications while a prevalence of 8-10% has been reported in women investigated because of recurrent pregnancy loss.[1]

The traditional hypothesis of mullerian development maintains that the müllerian ducts are fused in a caudal-cranial direction. Chang et al[2] challenged the classic hypothesis and supported an alternative embryologic hypothesis which states that fusion and resorption begins at the isthmus and proceeds simultaneously in both the cranial and caudal directions.

Sadik[3] reported a case of a complex mullerian malformation with a hypoplastic non cavitated uterus and two rudimentary horns, pathogenesis of which could not be clearly defined but could involve sequential embryological errors in the development process. They suggested need for inclusion of rare cases with abnormal mullerian development and opined that fusion of mullerian duct occurs first followed by resorption of the septum that begins at isthmus and extends cranially and caudally.

Nezhat[4] reported a case of unicornuate uterus with two cavitated non-communicating rudimentary horns and Stage IV endometriosis. This variant has not been described by the AFS classification and requests some elucidation of the embryology of mullerian system as the anomalies may result from failure of lateral or vertical fusion or failure of resorption. Crosby et al[5] have suggested that canalization follows fusion and it can begin in any location along the line of fusion and can proceed in any direction.

Acien et al[6] pointed out possible discrepancies in the classic hypothesis of Müllerian development and suggested that the fusion and reabsorption processes in the development of the Müllerian ducts can be affected to different degrees in the superior-convergent and inferior-divergent portions, thus resulting in atypical or transitional cases without a classification.

In our case, similar process could have occurred explaining dysgenesis at the level of isthmus which is very rare and can be included in class IV of Modified Rock and Adam-AFS classification.[7] This classification embraces a broader vision of anomalies without encountering over simplicity or conflicting observation as in other classifications and correlates anatomic anomalies with embryologic arrests.

Rare Mullerian anomalies should be kept in mind while evaluating cases of chronic pelvic pain. Complex unusual configurations and combination of defects are rare but possible and should be considered taking into account the alternative embryological hypothesis to ensure quick and appropriate management for the patient

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