Endometriosis in a case of Endometrial Agenesis and Cervical Atresia

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Maldevelopment of the Mullerian ducts occurs in a variety of forms and although each anomaly is distinctive, some generalization may be made. Disorders of vertical fusion of Mullerian duct is associated with cryptomenorrhoea, endometriosis, infertility.

Ms. S. N. a 16 year old unmarried girl, presented in the out patient department in December 1999 with complaints of primary amenorrhoea, history suggestive of cryptomenorrhoea since 3 years and past history of evaluation under anaesthesia (EUA) followed by attempted hymenectomy in May 1997. She gave history of 2 exploratory laparotomies, first in September 1997 for right adnexal mass which was diagnosed as endometriosis. A second laparotomy with right ovarian cystectomy was done in August 1998 for chronic pain in abdomen.

On examination, vital parameters were stable. She was 148 cms tall and weighed 42 kgs. Secondary sexual characters were well developed. Per abdominal examination showed a midline infraumbilical hypertrophic scar. Per speculum examination not done. Per vaginum examination showed blind vaginal pouch of normal depth, cervix not felt. On rectal examination, a uterine nodule felt and ultrasonography of pelvis showed a small uterus, and normal left ovary. A hypoechoic mass with few internal echoes was seen in the region of right adnexa. Right ovary was not visualised separately.

Evaluation under anaesthesia and diagnostic laproscopy were performed. Operative findings were –
EUA – blind vaginal pouch, normal vaginal depth, uterus normal size deviated to right, cervical atresia as cervix not felt.

Diagnostic laproscopy revealed less than normal size uterus. Right side adnexa was not visualized due to dense adhesions due to endometriosis. In view of absence of hematometra a diagnosis of absence of functioning endometrium in uterus was made. Patient was put on Nafarelin nasal spray 200 microgram 3 times a day for 3 months for Right ovarian endometriosis. During therapy patient had no cyclical lower abdominal pain.

One month after stopping Nafarelin nasal spray she again became symptomatic. Re-evaluation pelvic USG showed uterus less than normal size, no endometrial echo. Left ovary normal size with single follicle of 12x11 mm. In the region of right adnexa a hypoechoic mass with few internal echoes seen. Right ovary not seen separately from the mass. No free fluid in Pouch of Douglas.

Exploratory Laparatomy done in August 2000 Laparotomy findings were as follows; Left tube and ovary normal, uterus less than normal size. Cervix atretic. Right ovarian endometrioma adherent to sigmoid colon Sigmoid colon separated from the endometrioma. Total Abdominal Hysterectomy and Bilateral Salpingo oophorectomy performed.

Histopathology report confirmed the diagnosis. Uterus streak like projection. Only basal layer of endometrium seen. Area of cervix showed a white homogenous fibrous appearance with focal haemorrhage and cystic change suggestive of endometrioma. No evidence of cervical gland. Impression : Cervical atresia and endometriosis

Subsequently patient was put on hormone replacement therapy for 6 months after surgery which she tolerated well without any features of pelvic endometriosis.

Pelvic endometriosis develops secondary to implantation of retorgrade menstruation. This case was unusual in that there was cervical atresia and absence of functioning endometrium and still there was pelvic endometriosis.