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RESEARCH ARTICLE

CERVICAL AGENESIS – A RARE CLINICAL PRESENTATION.

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Abstract

Cervical agenesis is a rare mullarian anomaly occur 1 in 80,000 births. According to Modified American fertility society it is classified under class II b.

Case report:- A 40yrs old Nulligravida presented to Gynaec OPD with c/o urinary retention for 1 day, right loin pain for 10 months, with primary amenorrhoea. On examination secondary sexual characters tanner IV, per abdomen-a 28 weeks size mass arising from pelvis, local examination External genitalia were normal. Per speculum, vagina healthy, Cervix is not visualized. We Planned for laparotomy under general anaesthesia. We removed uterine body and fundus, both fallopian tubes and ovaries. Histopathological examination shows cervical agenesis with haematometra with subserous fibroid with haematosalpinx.

Discussion:- Mullarian anomalies, present in multiple ways from infancy to young adulthood. Patients may present with mucocolpos, hematocolpos or haematometra, primary amenorrhoea, pelvic pain, repeat pregnancy loss, and infertility. One of the rarest anomalies is cervical agenesis.

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Introduction:-

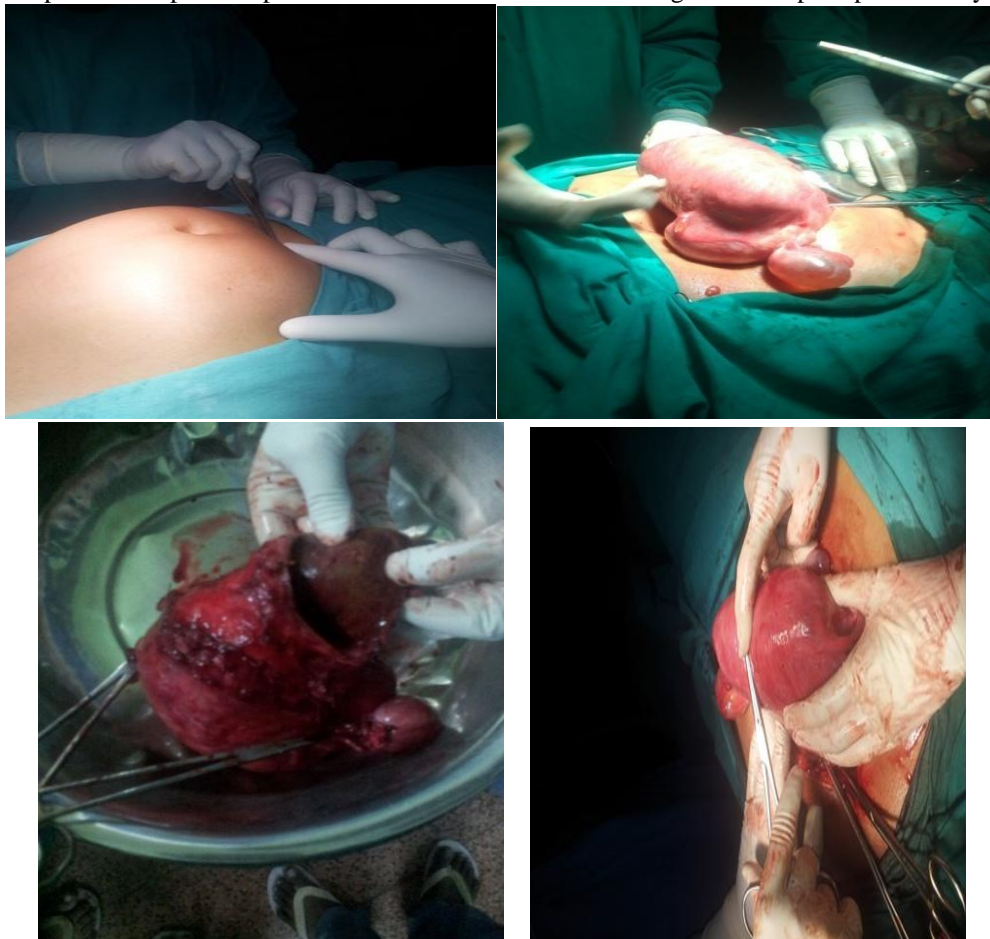
Cervical agenesis is a rare mullarian anomaly occur 1 in 80,000 births. According to Modified American fertility society it is classified under class II b which results from disordered vertical fusion of mullarian duct. It is usually associated with normal functional uterus and tubes. Presence of functional endometrium with outflow tract obstruction (cervical agenesis) and patent tubes is to be associated with endometriosis in 77% cases.

Clinical presentation is usually with primary amenorrhoea, cyclical abdominal pain and endometriosis or pelvic infection may result from the chronic hematometra as was seen in our patient.

Case report:-

- ❖ A 40yrs old Nulligravida presented to Gynaec OPD with c/o urinary retention for 1 day and right loin pain for 10 months.
- ❖ History of primary amenorrhoea present. Her marital life was 20 years. Her sibling also has similar history.
- ❖ On examination patient general Condition is good, thyroid, breast were normal. Secondary sexual characters were tanner IV, Cardiovascular system and respiratory system were normal.

- ❖ Per abdomen - a 28 weeks size mass arising from pelvis, which was cystic, non tender, smooth surface, horizontally mobile and vertical movements were restricted. No hepatosplenomegaly. Right renal angle tenderness present.
- ❖ Local examination External genitalia were normal.
- ❖ Per speculum , vagina healthy , Cervix is not visualized..
- ❖ On vaginal examination cervix not palpable, a cystic thick walled structure noted which is mobile with abdominal mass.
- ❖ Per rectus a mass is felt, rectal mucosa – normal.
- ❖ Diagnosed as hematometra with? cervicalagenesis ?cervical atresia.
- ❖ USG abdomen revealed grossly distended uterus with echogenic collection suggestive of haematometra with posterior wall fibroid of 4x3cm size, Cervix not visualized by TVS. Kidneys shows Bilateral moderate Hydronephrosis.
- ❖ CT abdomen and pelvis shows bicornuate Uterus with septum with haematometra with posterial wall fibroid with mild bilateral hydronephrosis.
- ❖ We Planned for laparotomy under general anaesthesia.
- ❖ Per operative findings were bladder edematous drawn up, Adhesions noted between Greater omentum and parietal peritoneum. About 2 liters of black tarry fluid aspirated from uterine cavity. Bilateral fallopian tubes were? agglutinated? atretic, a 3x3 cm cyst noted in Right Ovary. Left ovary is cystic. Subserous fibroid on right side of the uterus seen about 3x4cm, Cystic vesicles seen on uterine surface.? Endometriotic. We removed uterine body and fundus , both fallopian tubes and ovaries . Abdominal lavage was done. Blood loss about 1.5 liters noted. Intra operatively 1 unit of compatible blood transfusion given.
- ❖ Cut section shows Uterus was filled with tarry black coloured blood .
- ❖ Histopathological examination shows cervical agenesis with haematometra with subserous fibroid with haematosalpinx. Post operative period uneventful. Patient was discharged on 12th postoperative day.



Discussion:-

Mullarian anomalies, although rare, present in multiple ways from infancy to young adulthood. Patients may present with mucocolpos, hematocolpos or haematometra, primary amenorrhoea, pelvic pain, repeat pregnancy loss, and infertility. One of the rarest congenital anomalies is cervical agenesis, or the absence of cervix. These patients present with primary amenorrhoea, debilitating pelvic pain, and most commonly have a large pelvic mass consisted of obstructed outflow of menstrual blood after the onset of puberty. The incidence of cervical agenesis is unknown, but a review of literature lists less than 200 cases since 1942.

There has been a renewed interest in the clinical management of congenital cervical agenesis and dysgenesis with the publication of over 10 articles on the subject since 2000. Grimbizis et al. published a complete review of the literature reporting 116 cases of transverse cervical defects since 1900. The authors here have outlined recommended surgical alternatives for patients with cervical anomaly observed.

Embryology of the female reproductive tract:-

Two mullarian ducts develop into the fallopian tubes Uterus, cervix, and upper two thirds of the vagina. Complete formation and differentiation of the mullarian ducts into a functioning female reproductive tract depend on completion of three phases of development as follows;

1. Organogenesis : one or both mullarian ducts may not develop fully, resulting in abnormalities such as uterine agenesis or hypoplasia or unicornuate uterus.
2. Fusion:-
 - a. lateral fusion forms the uterus, cervix, and upper vagina. Failure of fusion results in anomalies such as bicornuate or didelphys uterus.
 - b. Vertical fusion is fusion of the ascending sinovaginal bulb with the descending mullarian system. At about the 20th week of gestation, the cervix forms as a result of condensation of stromal cells at a specific site around the fused mullarian ducts. Complete vertical fusion forms a normal patent vagina, whereas incomplete vertical fusion results in an imperforate hymen.
3. Septal resorption occurs after the lower mullarian ducts fuse. Initially, a central septum is present, which subsequently must be resorbed to form a single uterine cavity and cervix. Failure of resorption is the cause of septate uterus.

Anatomical variations of congenital cervical anomalies:-

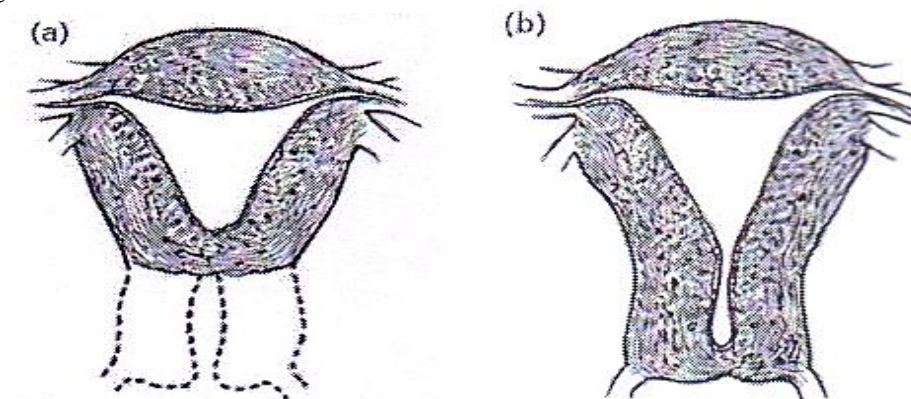
Two basic categories of cervical anomalies have been observed

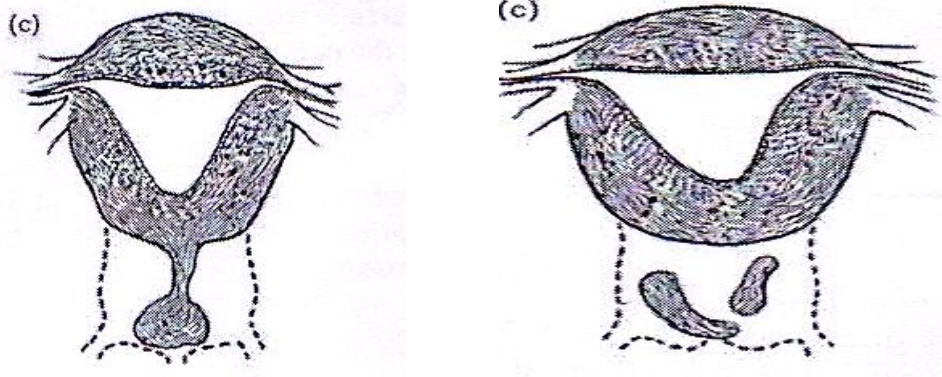
First one is, cervical agenesis

Second one is cervical dysgenesis

Can be described as four subtypes

- a. Cervical body consisting of a fibrous band of variable length and diameter
- b. Intact cervical body with obstruction of cervical os
- c. Stricture of the mid portion of the cervix
- d. Fragmentation of cervix.





In cervical agenesis with functioning uterine corpus had to endometriosis , cryptomenorrhoea ,and sepsis. In order to avoid these complications many authors recommended hysterectomy .in this case patient had hypothyroidism which explain hyperoestrogenic state. This may explain the presence of subserous fibroid. The adhesions, endometriotic vesicles may be due to reflux of endometrial fluid into the peritoneal cavity. SO we carried out removal of whole uterus, tubes and ovaries .

Conclusion:-

In cervical agenesis without residual endometrial tissue hysterectomy is the treatment of choice.

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