

## HUGE ABDOMINAL MASS SECONDARY TO A TRANSVERSE VAGINAL SEPTUM AND CERVICAL DYSGENESIS

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### SUMMARY

Transverse vaginal septum is a rare occurrence. When it co-exists with cervical dysgenesis, it is even rarer. Primary amenorrhea with cyclic pain is one manifestation in post-pubertal women. A case of transverse vaginal septum with vaginal atresia and cervical dysgenesis is presented. Presentation as a huge abdominal mass and severe anaemia posed diagnostic challenges. A two-staged management is described as well as variation in management of the septum. The involvement of the patient in her follow up is also stressed.

### Keywords

Primary amenorrhea, abdominal mass, vaginal septum, cervical dysgenesis

### INTRODUCTION

Transverse vaginal septum is a rare condition that results from incomplete fusion between the vaginal components of the Mullerian ducts and the urogenital sinus.<sup>1</sup> The septum varies in thickness and may be located at any level in the vagina, although most are found in the upper and mid-vagina.<sup>2,3</sup> Clinical presentation depends on whether it is complete or partial. With complete septa, menstrual blood accumulates and distends structures above the septum after puberty, resulting in hematocolpos and haematometra. Such patients usually present with cyclic lower abdominal pain and ultrasonic findings of haematocolpometra. Occasionally a lower abdominal mass (haematometra) is palpable. Incomplete septa allow partial egress of menstrual blood and such patients usually complain of cryptomenorrhea, dysmenorrhea and dyspareunia.

Cervical dysgenesis is an even rarer condition often associated with atresia of the vagina.<sup>1</sup> Clinical diagnosis is usually difficult before surgery. We present the case of a 20 year old woman with primary amenorrhea who had the rare combination of a low transverse vaginal septum and cervical dysgenesis presenting with wasting, severe anaemia and a huge, painful abdominal mass.

### Case Report

A 20-year old nulliparous woman was referred from a district hospital in late December 2008 as Severe Anaemia and Abdominal swelling of 6 years' duration. She had been transfused three units of whole blood. At the medical emergency where she was initially admitted, positive findings included extreme wasting, moderate pallor, ascites and a firm, tender multi-lobulated abdominal mass of 32 weeks' size that arose from the pelvis. Abdominal paracentesis yielded hemorrhagic ascites which analysis showed markedly raised LDH (4302U/L) and total protein 200.0g/l, but reduced albumin (26.0g/l). She was managed propped up in bed with oxygen by nasal tube, intravenous infusions and two units of whole blood and scheduled for abdominal CT scan. Her haemoglobin was 7.3g/dl. The liver and renal function tests as well as electrolytes were within normal ranges.

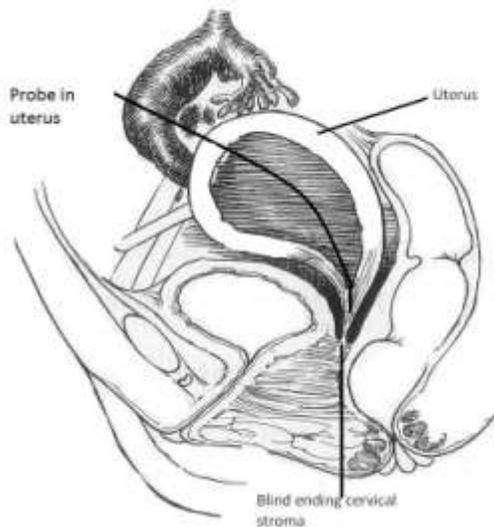
A day later the history of 'never menstruated' was fortuitously obtained during discussions with the mother. An abdominal ultrasound scan subsequently done showed a large haematometra and fluid in the abdomen. There was no hematocolpos. The liver, kidneys, spleen, ureters and urinary bladder were all normal on ultrasound. The patient was thus referred to our department for further management. We found her to be slightly pale, small for her age, wasted and very dyspneic on lying supine. She had normal female features and well developed breasts (Tanner Stage 5). Abdominal findings were essentially as described. She had normal female external genitalia. The vagina was shallow, about 1.0cm deep and limited by a transverse septum that was not bulgy (Figure 2). A rectal examination confirmed a pelvic mass but no bulge in the vagina.

A diagnosis of Transverse Vaginal septum with probable vaginal atresia was made. A two-stage surgery was planned: laparotomy to drain the haemorrhagic ascites/haematometra and definitive vaginoplasty for the septum.

She was administered Depo-Provera injection, transfused two more units of whole blood and covered with broad-spectrum antibiotics to make her fit for surgery. Her HIV antibody screen was negative. At laparotomy a week later, findings were chocolate-coloured ascites of 400ml and an enlarged uterus of 32 weeks containing 3.5 litres old menstrual blood that was drained through a low vertical uterine incision. A probe passed through the uterine incision towards the cervix ended blindly. The tubes were distended with blood and matted with the ovaries and broad ligaments. A biopsy was taken of the endometrium and myometrium for histopathology.

Histopathologic report read 'chronic inflammation of the endometrium and myometrium; no malignancy noted'. She made good progress and after 3 weeks the uterus was not palpable. She was discharged home with an Hb of 10.7g/dl.

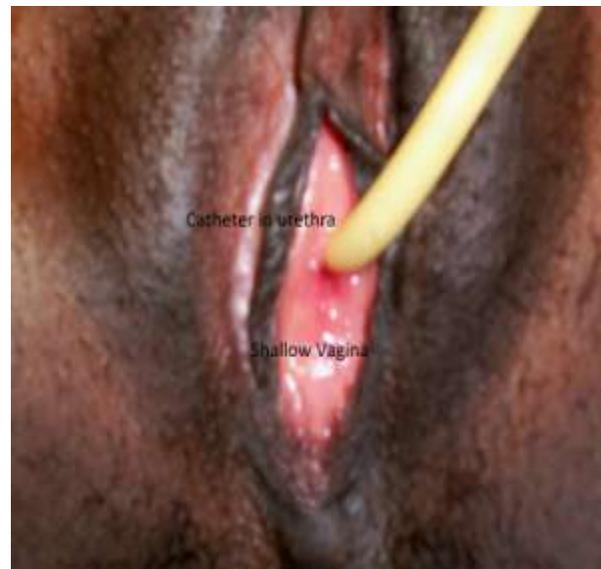
Three months after the first surgery, she was re-admitted for excision of the vaginal septum. Two surgical teams undertook this operation. One team worked on the vagina to excise the septum. The vagina was atretic and dissection was done in the loose areolar space between the bladder and rectum towards the cervix. Cervical stroma was identified with a small dimple but without any os or canalization. The other team performed a re-laparotomy passing a probe through a stab incision at the uterine fundus towards the cervix to help the vaginal team. A canalization of the cervix was done through the identified dimple using mosquito artery forceps and with the help of the probe in the canal abdominally as a guide (as shown in Figure 1).



**Figure 1** Graphic presentation of a probe in the uterus towards the blind cervix

The non-canalised part of the cervix was about 0.5mm thick. The excised edges of the septum were undermined and sutured together without tension. The neo-vagina was packed with gauze in chlorhexidine solution. The fundal incision was closed with a figure-of-8 stitch and abdomen cleaned and closed.

The vaginal pack was removed after 24 hours and replaced with a male condom filled with gauze to keep the vagina patent. This was changed every other day for two weeks. Daily vaginal dilatation with metal boggies was started in the 3<sup>rd</sup> week. Vaginal examination at 4 weeks showed a patent vagina that admitted 2 fingers freely and to a depth of 6.0cm. The need for regular vaginal dilatation was stressed to the patient who was taught self-dilatation and discharged home.



**Figure 2** Vulval appearance of patient before 1<sup>st</sup> surgery.

Patient reported weekly for 4 months for vaginal examination and dilatation in theatre in addition to what she did in the house daily. She has subsequently been seen monthly for it. The vagina has remained patent admitting two (2) fingers freely to a depth of 6.0cm, after 15 months, although a circular cicatrisation can be felt where the septum was excised. The cervical canal has also remained patent. Patient has had normal monthly menses since the last surgery. Karyotyping results received shows 46XX. Follow up continues.

## DISCUSSION

We have presented a rather rare combination of a transverse vaginal septum and cervical dysgenesis. The major diagnostic challenge in this case was the lack of adequate history taking initially when the patient presented: the history of 'never menstruated' was not ob-

tained on admission. Diagnosis of such cases is usually arrived at from the history of cyclic pain and primary amenorrhea with or without the clinical finding of a bulging vaginal septum.<sup>4</sup> This patient had primary amenorrhea, and though she admitted to having cyclic pain earlier on, over the years she had gotten used to the pain such as not to complain about it again. The finding of no bulge in the vagina on rectal examination raised the suspicion of vaginal atresia. This was confirmed by ultrasound findings of no hematocolpos. A two-staged surgical procedure was adopted for this patient for two reasons: she was unfit for the relatively longer procedure of excision of the septum and was in respiratory distress from the abdominal mass that had to be relieved. She was put on Injection Depo-Provera to thin out the endometrium and reduce further bleeding into the mass while waiting for surgery.

When transverse septum is diagnosed after onset of puberty, often a large segment of the vagina is absent, making anastomosis of the upper and lower segments difficult.<sup>5</sup> A Z-plasty method of bridging this gap has been described.<sup>6</sup> A simpler flap method has also been described.<sup>7</sup> Neither the Z-plasty method nor the simple flap method was used in this case. A simple excision and dissection through loose tissue was done to create a neo-vagina. To prevent stenosis during the phase of epithelialization, a male condom filled with gauze was inserted in the vagina and changed every other day for two weeks, after which daily dilatation with metal bobbies was done for another two weeks before the patient was discharged. She continued with daily self-dilatation in addition to being seen in theatre once a week for four months and later monthly. Canalization procedures for the cervix have been documented to be successful especially where the cervical body is intact

but with obstruction of the os in carefully selected cases, such as this patient.<sup>8</sup> The 'neo-cervical canal' created has remained patent for fifteen months without need for any dilatation

## ACKNOWLEDGEMENTS

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