Case Report

Familial Occurrence of Imperforate Hymen in A Muslim Family- Treated by Hymen Sparing Surgery in one and Hymenectomy in Other

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Abstract

Background: Although imperforate hymen occurs in approximately 1% of female newborns, familial occurrence has been reported rarely.

Cases: We report one family in which imperforate hymen was diagnosed in two siblings. Both cases presented with cyclical abdominal pain and hematocolpos. The cases are described in detail and both are post menarche.

Conclusion: Imperforate hymen usually occurs sporadically but the occurrence can be familial. It is advised to screen all newborn females for vaginal patency specially if there is history of effected child in the family.

Key Words: Imperforate hymen, Gynecological anomalies, Hymen sparing surgery

Case Report

A thirteen years old girl reported with her mother in gyne OPD in 2005 with complaints of primary amenorrhea and regular cyclic abdominal pain for six months. The intensity of pain had increased considerably for the last four days and she had taken different pain killers to alleviate the problem. On examination she was looking apprehensive and restless. There was a doughy swelling in the abdominopelvic region and a bluish membrane at vulva. Rectal examination revealed a tense cystic mass anteriorly, suggestive of hematocolpos. Pelvic ultrasound illustrated a large amount of fluid in uterus and hematocolpos of 10 x 12 cm.

The patient and mother were counseled about the problem and different surgical treatment options (hymen sparing surgery, hymenectomy after cruciate, plus, X shaped and central oval incision followed by Foley catheterization). The mother was worried about the integrity of the girl's genitalia and effect on her fertility. The option of hymen sparing procedure was preferred because there is importance of integrity of hymen in the Muslim culture. However the mother gave consent for the best treatment according to the situation.

Under general anesthesia the hymen was incised by vertical incision and about 800 ml thick chocolate colored fluid was drained under oxytocin infusion. Few oblique sutures were applied to prevent reunion of aperture. No further excision was done because the area of incision was sufficient for drainage of blood. In order to prevent any adhesions, the patient was advised to apply estrogen cream on the hymenal area for two weeks and was also advised to follow up in her next menstruation. She reported back in the next menses after one month and was found to have normal menstruation. Out of younger siblings of the family the second one had normal menstruation.

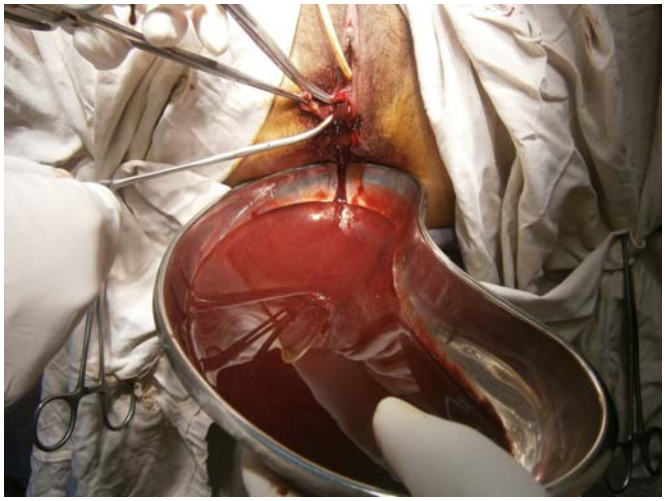

Figure 1. Blood Flowing From Vagina after Incision of Hymen

After 7 years, in 2013, the same mother reported to gyne OPD with her third daughter of 12 years crying with pain. She presented with a history of primary amenorrhea and abdominal pain for one month. This was her second episode of abdominal pain and as the mother had her one daughter with an imperforate hymen, she had an ultrasound report which spoke clearly of the same occurrence.

Examination revealed a lower abdominal tender bulge, bluish membrane at vulva and on rectal examination a tense cystic bulge was felt anteriorly. Ultrasound revealed hematocolpos of 10 x 10 cm with marked hematometra. The mother again opted for hymen sparing surgery because the elder daughter fared well in terms of menstruation. The hymenal membrane was incised by vertical incision under general anesthesia and about 900 ml thick, chocolate colored fluid was drained under oxytocin infusion. This was followed by placement of two oblique sutures in hymen. The patient was sent home with advice to apply estrogen cream to prevent adhesions and follow up after next menstruation. She reported back after nine weeks with same symptoms. On examination the orifice had reunited and bluish membrane was visible at introitus.

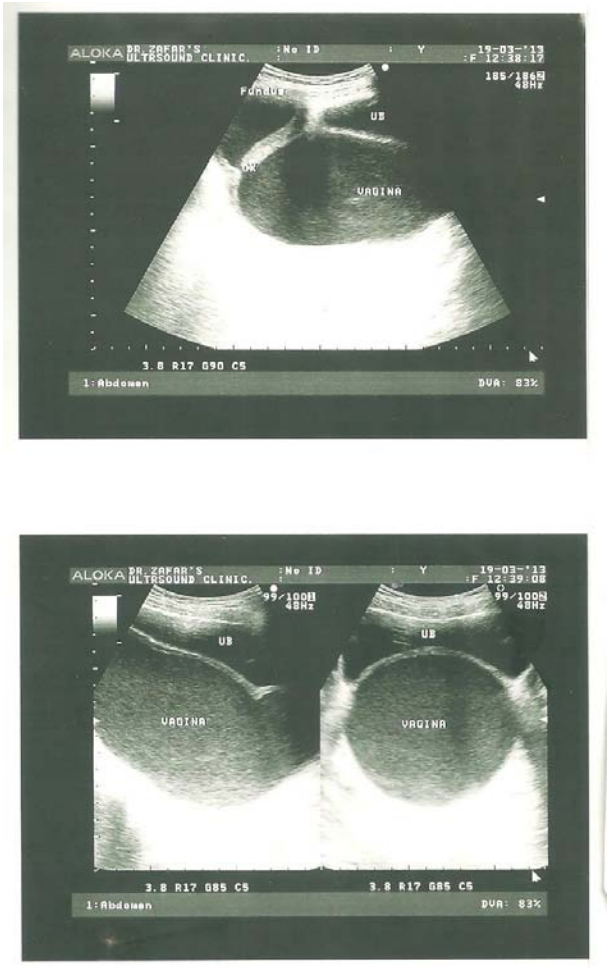

Figure 2. Ultrasound Pelvis on First Consultation: S_1

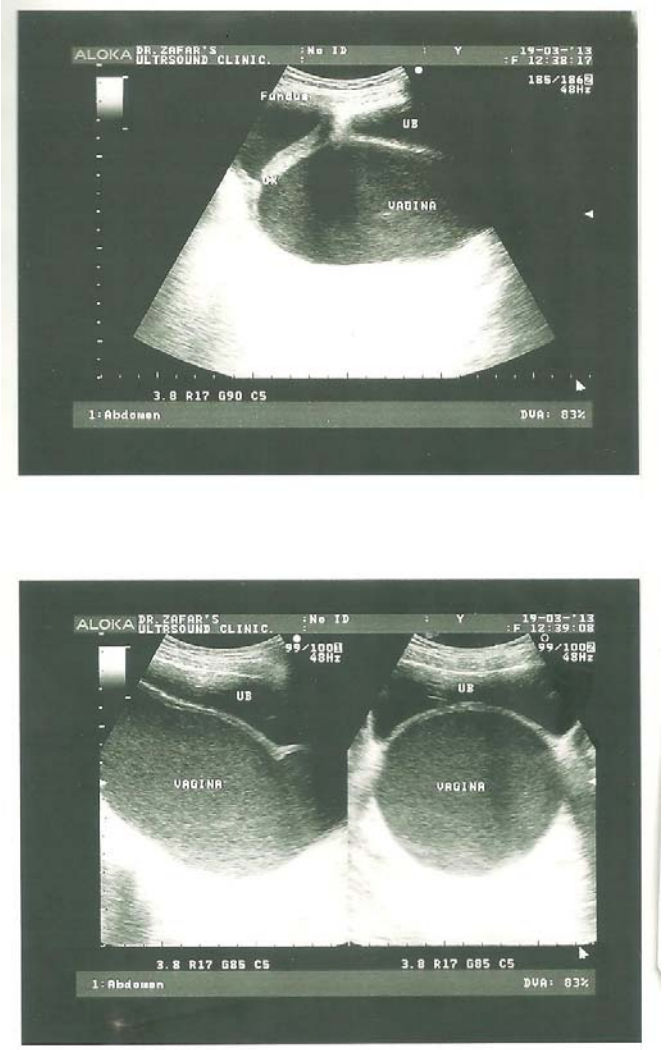

Figure 3. Ultrasound Picture on Second Consultation: S_2

Ultrasound showed hematocolpos. Under anesthesia hymen was incised and again chocolate colored fluid drained. Simple hymenectomy was performed and patient was advised oral antibiotics and estrogen cream for local use and follow up after four weeks. She reported back with normal menstruation after six weeks.

Both girls are unmarried and I hope for normal reproductive outcome because they were diagnosed and treated at an early stage.

Discussion

The uterus and vagina are formed embryologicaly by the dynamic process of differentiation, migration, fusion and canalization. A wide variety of anomalies occur of both uterus and vagina or separately by disruption of any process of derivation. Selective agenesis of lower vagina and vaginal atresia is a separate entity and is usually associated with normal Müllerian derivatives.

I have come across a few patients of imperforate hymen but it is the first time I am seeing two siblings with the same genital tract anomaly from the same family. Careful examination and appropriate treatment is the key to success for patient care and normal reproductive outcome. Detailed counseling of mother and patient is required. If no harm has occurred then there is no difference of reproductive outcome between normal and affected patients.

The anomaly of Imperforate hymen is seen occasionally and occurs in 0.1 % of females. Familial occurrence of imperforate hymen is extremely rare and has been reported only three times in the English literature. Although no genetic markers or mutations have been proven as etiological factors and both the recessive and the dominant modes of transmission have been demonstrated. In order to treat at early stage the evaluation of female siblings of the effected patients is very helpful.

Salkalkale R and Samarakkody U also reported familial occurrence of imperforate hymen in which they described two cases in a family across two generations, one presenting with chronic cyclical abdominal pain, and the other acutely. There were no significant reproductive or systemic abnormalities in either case.[1]

Usta IM and coworkers reported two families in which imperforate hymen were diagnosed in three siblings of each family. The patients were two postmenarchal young women and one premenarchal girl.[2]

Basaran M, Usal D and Aydemir C reported about the consequences of hymen sparing surgery for imperforate hymen because of integrity of hymen changes in different cultures and religious groups. The procedure they adopted was giving simple vertical incision and application of few oblique sutures to prevent refusion and this resulted in uneventful postoperative recovery.[3]

Hymenectomy may not be needed in all cases if adequate opening for menstrual discharge is warranted. Kurdoglu Z, Kurdoglu M and Kucukaydin Z reported spontaneous rupture of imperforate hymen in an adolescent girl with hematocolpometra just before a planned hymenectomy operation. The patient was managed conservatively with a satisfactory outcome.[4]

Dickson CA, Saad S and Tesar JD reported two cases of imperforate hymen in pediatric emergency. One presented with abdominal pain, urinary obstruction and constipation and the other one with syncope and bilateral lower abdominal pain. Both patients had hematocolpos and underwent hymenectomy and experienced no further symptoms.[5] The primary amenorrhea combined with abdominal colic should always suggest to the surgeon and the pediatric specialist the possibility and existence of a genital malformation.

Acar A, Balci O, Karatayli R, Capar M and Colakoglu MC presented a prospective study of treatment of 65 women with imperforate hymen by a central incision followed by insertion of 16F Foley catheter for two weeks. Hymenal orifice remained open and intact in all women except for two. Closure of artificially created hymenal orifice in these two women was treated with local estrogen cream which was successful. This technique is less invasive than others and prevents many social problems related to virginity by preventing destruction of the integrity of the hymenal structure.[6]

Ben Temime R, Najar I, Chachia A, Attia L, Makhlouf T and Koubaa A discussed a series of 13 cases of imperforate hymen. The mean age was 14 years. All patients were single and had primary amenorrhea. They presented with pelvic pain in 9 cases and bladder urinary retention in 4 cases. Secondary sexual characters were present and normal in all cases. Inspection of the vulva could establish the diagnosis in all cases. Pelvic ultrasounds showed the hematocolpos in all cases. The latter was associated to a hematometra and a Pouch of Douglas liquid in 2 cases. Hematocolpos was evacuated by hymenectomy under oxytocin infusion in all cases. Eight patients were treated by cruciform incisions and five patients were treated by radial incisions of the hymen. The volume of hematocolpos varied from 250 ml to 2000 ml. One patient underwent surgery twice for restenosis of the imperforate hymen.[7]

The physiological importance of this problem in adolescent age must be dealt with empathy and reassurance. I described two sisters of a family with imperforate hymen. One presented with chronic cyclical abdominal pain for six months and the other presented acutely in the second episode of cyclical abdominal pain with an ultrasound report. There were no significant reproductive or systemic abnormalities in either case.[10] The level of education of the mother is important because in our setup patients usually go to hakims and quacks instead of visiting the appropriate doctor. As the family was well educated, they consulted with the concerned specialist. The diagnosis was early and treatment of the said condition which caused no harm to the reproductive structures and I hope for a normal reproductive outcome of these patients.[8,9] The importance of integrity of hymen changes is different the Muslim culture. Preservation of hymenal tissue to have the perception of integrity of female genitalia is an

important aspect to be considered as the treatment option. As the mother was anxious and concerned about this aspect, so a simple longitudinal incision was adopted as a treatment and in both cases. There was enough space after the incision but in the second daughter surgery was required due to reunion of membrane. Both patients now have normal menstrual cycles. I have still not evaluated their other siblings because of the shyness of the girls.

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