INTRODUCTION

Anorectal malformations (ARM) have a reported incidence of approximately 1 in 5,000 births. In males, the more complex malformations connect internally to the lower urinary tract, urethra or bladder. However, in females, the vestibular fistula represents the most common type of anorectal defect with associated malformations. Adult presentation of this condition is rare since most cases are diagnosed and corrected at birth.

The delayed diagnosis of such malformations will be associated with complications due to the effects of constipation or in the delay and alteration of surgical management. In addition to the physical problems of this condition, there is a possibility of development of intractable constipation or there could be social and psychological morbidity due to lack of control over defecation and passage of flatus. Furthermore, female patients may experience adverse effects during pregnancy and delivery.

The following is a case of a late presentation of an adult female with the anorectal malformation with a vestibular fistula, with no associated anomalies, presenting late due to inadequate medical facilities. She was successfully treated by a procedure in which the rectum was mobilized and brought through external sphincter.

CASE REPORT

A 23 years old, unmarried lady belonging to rural Sindh presented in the surgical OPD with a complaint of anorectal malformation since birth and passage of fecal matter through the fistula. According to her mother, a local doctor advised that a definite procedure would be performed when she would be 2 years of age. Thereafter, she remained untreated. She learned to pass stool through the fistula when the urge was felt once a day and did not have incontinence as such but she did not have any control over flatus. Later, she developed normal menstrual cycles and secondary sexual characteristics at puberty. Now, as her parents wanted her to get married, she was brought to the hospital for a reconstructive procedure.

On examination she had vestibular fistula through which fecal matter was coming out with an associated anorectal malformation (Figure 1 and 2). A lumbosacral X-ray was done which showed no abnormalities. The patient also underwent an ultrasound, which was normal. Rest of the clinical and para-clinical parameters were within normal limit.

Intraoperatively the fistula was observed. Muscle stimulators were used to localize the sphincters. During the procedure, the rectum was dissected out anteriorly from the vagina and was brought out through the external anal sphincter after a cut back incision, through the proposed site of external sphincter (Figure 3).

Postoperatively, the patient was incontinent for both feces and flatus for one month. She was advised to perform pelvic floor exercises. After one month, she became continent with the help of the pelvic floor exercises. However, after two months she developed anal stenosis for which anoplasty was successfully performed.

Key words: Anorectal malformation. Vestibular fistula. Late presentation. Anal transposition. Anal stenosis.

ABSTRACT

Vestibular fistula represents the most common type of anorectal defect seen in girls with anorectal malformation. Adult presentation of this malformation is rare. The following is a case of adult presentation of anorectal malformation with a vestibular fistula in a 23 years old female, who came because of cosmetic and fertility concerns before her marriage. The patient complained of having an anorectal malformation since birth and passage of fecal matter through the fistula. The patient was continent for feces, but had leakage of flatus. She had no other associated anomaly. The patient subsequently underwent a surgical procedure where anal transpositioning was done with an intact skin bridge. Post-operatively, these developed an anal stenosis which was successfully treated through anoplasty, and the patient is now continent both for feces and flatus for the last 6 months.
performed. She is now on regular anal dilatations and pelvic floor exercises and she is continent both for feces and flatus for last 6 months.

DISCUSSION

Vestibular fistula was described as a fistulous tract between the bowel and the low female genital tract by Bryndorf and Madsen in 1960. It represents the most common type of anorectal defect seen in girls with anorectal malformation.

All significant malformations are readily apparent on routine neonatal examination. For neonates born with an anorectal anomaly, early treatment is crucial. However, as in this patient, illiteracy, lack of neonatal care and inappropriate medical facilities may lead to the persistence of this condition to adulthood. Kumar et al. described 2 cases of anovestibular fistula presenting in adulthood.

Anorectal malformations are associated with other congenital abnormalities, seen commonly in multi-anomaly sequences such as VACTERL. It is imperative to perform a thorough systemic examination together with radiological tests on a patient presenting with a vestibular fistula. However, anorectal malformations are most commonly associated with other genitourinary abnormalities such as described by Adejuyigbe et al. in a case of a 15 years old Nigerian girl with vestibular fistula associated with distal vaginal atresia.

Delayed presentation of an anorectal malformation (ARM) could be a result of a number of possible reasons. Some common reported causes in a developing country such as ours include inadequate treatment, delayed diagnosis, wrong treatment advice as well as social factors, such as lack of money or social support. The mode of presentation in these patients included constipation, ARM discovered during evaluation for infertility, or as was the case in this patient, before a young girl’s impending marriage.

The delayed diagnosis of such malformation will be associated with complications. These may be due the effects of constipation or in the delay and alteration of surgical management. It also leads to recurrent genital and urinary tract infections and may even lead to infertility. Delayed presenters also have inadequate weight gain and increased parental anxiety for a greater period of time.

In this patient, some of the complications of delayed presentation did manifest themselves. There was an obvious cosmetic concern associated with the malformation, which prompted the patient's family to seek medical help before her impending marriage. There were also concerns regarding the fertility and her ability to successfully carry a pregnancy to term. In addition, the difficulty of passing stool through the fistula as well as the leakage of flatus greatly diminished her quality of life.

Repair of ARMs can itself lead to several complications. Sinha et al. described constipation, anal excoriation and occasional soiling as some of the major postoperative complications in their study population. Other studies report recurrence of fistula, mucosal prolapse and sexual abnormalities in males in the form of retrograde ejaculation and abnormal penile erection.

Furthermore, despite optimal surgical management, no adequate repair for poorly developed muscles or nerves has been developed. In this case described by Adejuyigbe et al., which was subsequently treated by surgery, the girl was eventually able to conceive and carry the pregnancy to term. Similarily, Iwai et al. also mentioned 4 female patients who had undergone repair for ARM, and later had been able to conceive and deliver children through normal vaginal route. This is an especially important point in this being the major concern for the patient and her family.
Early detection and management is the key to better outcome due to multiple reasons. Surgery done in infant pelvis is found to be more convincing as at this stage the sphincter muscles are soft and easily manageable in comparison to the adult. Also the cortical integration of the somato-sensory input is usually lost with advancing age. Older aged females already have become so much matured in the defecating pattern that their toilet training becomes very difficult and they have to be kept on long-term bowel management program.

This case report highlights the existence of adult patients with anorectal malformations in our environment due to various reasons. This should also increase the awareness among surgeons regarding the possibility of successful management of such patients allowing them to lead a normal life.

REFERENCES