CASE REPORT

ANORECTAL MALFORMATION IN A TEENAGER IN GHANA- A CASE REPORT

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Abstract

Anorectal malformation is a congenital anomaly comprising a wide spectrum of diseases, which can affect male and female and can involve the distal anus and rectum as well as the urinary and genital tracts. The late presentation of patients with anorectal malformations especially in adolescence is known but rare. This is particularly true in females with rectovestibular fistulas. We present the case of a 17-year old female who underwent a posterior sagittal anoplasty on this account with satisfactory outcome.

Key Words: Anorectal malformation, Rectovestibular fistula, Anovaginoplasty, Colostomy

Introduction

Anorectal malformation is a congenital anomaly comprising a wide spectrum of diseases, which can affect male and female and can involve the anus and rectum as well as the urinary and genital tracts. The approximate incidence of anorectal malformations is stated as 1 in 5,000 births. In females, recto-vestibular fistulas are most commonly encountered while recto-urinary fistulas are the most common anomalies in males[1].

Delayed presentations of patients with these malformations are known especially in developing countries, majority being outside the neonatal period but within infancy or early childhood[2,3]. Presentation as adolescents, teenagers or young adults is a rare event and is associated with undesirable sequelae. Multi-staged surgical procedures are also required to tackle these anomalies at this late presentation[4,5].

Reasons adduced for late presentations are varied and these include poor neonatal services at birth, poverty, poor social support and wrong diagnosis and/or treatment[4].

We present the case of a female teenager who was first seen at our out-patient department at 17 years of age. She had been passing faeces and flatus via a rectovestibularfistula. She was managed successfully with a posterior sagittal anovaginoplasty after a diverting colostomy.

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CASE REPORT

A 17-year old female was seen at our facility with the complaint of passage of faeces from an abnormal opening close to her vagina. She had noted this for as far back as she could remember. She recalled that her mother made attempts to get medical help but this failed to materialise before her demise. She was however continent of faeces and did not suffer from any chronic illness. Menarche was attained at the age of 15 years and menstrual cycles have been regular since then. She hailed from Anloga (Volta region of Ghana) and was engaged in peasant farming. Her presentation was occasioned by preparations for marriage.

Physical examination confirmed the presence of a vestibular fistula. There was no anal opening (Figure 1). Her gluteal muscles were well developed and the natal cleft and anal dimple were prominent. The rest of the systemic examination was unremarkable. Sacral X-ray was normal.

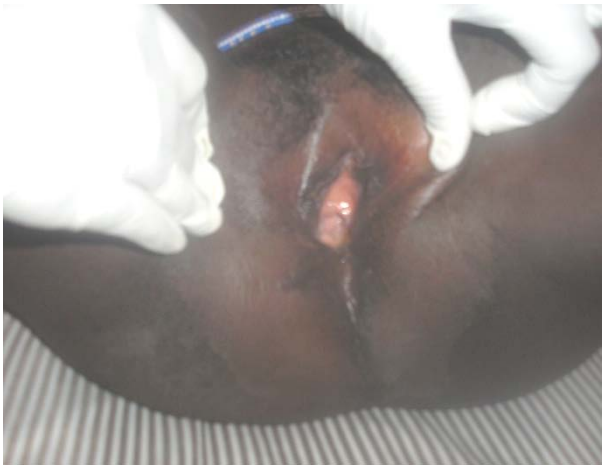

Figure 1: 17 year old girl with no anus but faeces in the vagina

A staged surgical treatment was then scheduled starting with a divided sigmoid colostomy. Six months later, a minimal anorectoplasty was done through a posterior sagittal approach. The fistula was first circumferentially mobilized and the rectum was then meticulously separated from the vagina before creating a neoanus. The procedure was complicated by wound dehiscence and retraction of the neoanus, necessitating a revision anoplastytwo weeks after the initial procedure (Figure 2).

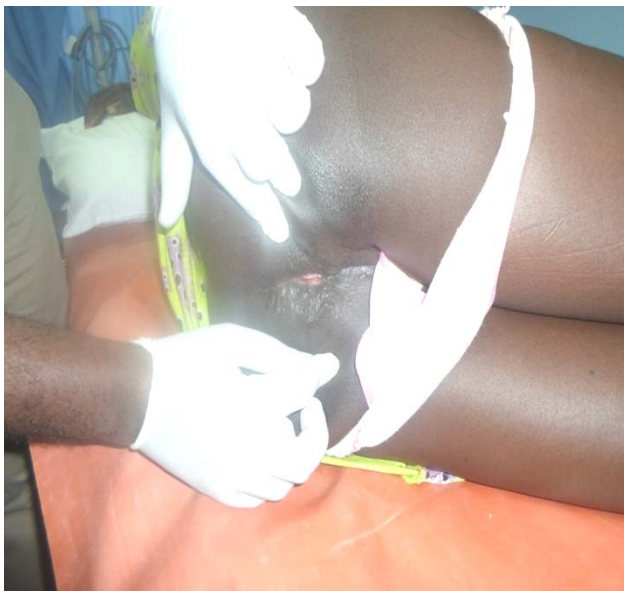

Figure 2: Anal orifice after surgery.

Serial anal dilatation was commenced on the 14th post-operative day and the colostomy closed 4 months afterwards. Out-patient follow up has been regular, revealing normal bowel opening habits and faecal continence. During her last clinic visit, two years after her first presentation, she was married and 7 months pregnant.

DISCUSSION

Meticulous inspection of the perineum of a female newborn is sufficient for the diagnosis of an anorectal malformation (ARM) with a rectovestibular fistula. The fistula is usually seen as a third orifice apart from the urethral and vaginal openings. It is the commonest form of the condition in females[1].

Surprisingly late presentations of such cases have and are still being reported[6,7,8,9]. This is usually in developingcountries and is linked with poor neonatal services at birth and poverty[4]. Poor social support and wrong diagnosis and/or treatment advice have also been cited as causes of late presentation to the appropriate care giver[2]. In the case presented there was evidence of poor social support, coupled with ignorance and illiteracy. The reason for her eventual presentation which was linked to concerns about marriage has also been previously documented in the literature[9].

ARM could be syndromic[1]. Even the non-syndromic varieties often present with associated anomalies represented by the VACTERL association (Vertebral; Anorectal; Cardiac; Tracheo-Esophageal fistula; Renal; Limb). There was no clinical or radiological evidence of anomalies in other systems in the case of our patient. Constipation, the major pre-treatment complication in such cases[10,4], was also absent.

Although Sanchez Martin et al[6] reported treating all but one of their cases of vestibular fistula in older girls with a single stage procedure followed post-operatively by administration of parenteral nutrition, we opted for a multi-stage approach. The existence of a megarectum at this age and our limitation in administering parenteral nutrition necessitated this approach. We also wanted to minimise the risk of post-operative infection and dehiscence[1]. Late presenters, like our patient, are more likely to have multiple procedures[11,12,4]

We used the posterior sagittal approach for the definitive anovaginoplasty as described by Pena, an approach which has been reported to have favourable outcomes even in late presentations[1,2]. We attributed the wound dehiscence and subsequent retraction of the neoanus post operatively to sub-optimal post-operative nursing care. The revision surgery however had an uneventful post-operative period. Serial anal dilation was prophylactic to forestall stenosis. Underestimation of this simple but vital post procedure practice carries with it the development of megarectum and faecal incontinence from chronic constipation[1]. As expected, there were no issues with continence following the closure of the colostomy. Our patient has since married and is now pregnant for 7 months.

CONCLUSION

The late presentation of ARM with rectovestibular fistula is avoidable since the diagnosis can be made by simple inspection of the perineum at birth. A 17-year old female was treated successfully, following a divided colostomy, using the posterior sagittal approach described by Pena. This approach is associated with favourable anatomical and functional outcomes and it also addresses the psycho-social challenges associated with this condition.

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