Internal Fat Prolapse from Ischiorectal Fossa Masquerading as Currarino Syndrome

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Abstract

Pelvic magnetic resonance imaging (MRI) is the latest addition to already existing tools for evaluation of anorectal malformation (ARM). It provides detailed information about pelvic floor musculature as well as position of rectal pouch. However, lack of knowledge about normal pelvic floor anatomy can lead to misinterpretation of pelvic MRI which can create confusion and change in approach for surgery. A 14-month-old male child with diagnosis of ARM was evaluated with pelvic MRI. There was finding of abnormal protrusion of fat through the ischiorectal fossa which was misinterpreted as Currarino syndrome which created confusion immediately before posterior sagittal anorectoplasty. Pelvic MRI is highly informative while evaluating a case of ARM. However, a detailed knowledge of pelvic floor anatomy is mandatory to avoid wrong interpretation and misdiagnosis.

Keywords: Anorectal malformation, Currarino syndrome, magnetic resonance imaging, presacral mass

Introduction

Currarino syndrome is a rare entity characterised by a triad of anorectal malformation (ARM), presacral mass and sacral deformity.\(^1\) Pelvic magnetic resonance imaging (MRI) is being used more frequently in case of high ARMs. It is helpful in identifying the level of blind-ending rectal pouch as well as detecting its communication with genitourinary tract. Pelvic MRI provides detailed information about pelvic floor muscles as well as external anal sphincter, which helps in predicting the functional outcome of surgery. Any abnormal mass or defect in the spine can also be detected easily by pelvic MRI. However, it has not yet established its place in routine imaging protocol of ARM.\(^2\) In this case report, we are presenting one such experience with pelvic MRI in a case of ARM in which prolapse of fat from the ischiorectal fossa was misinterpreted as presacral mass making a false impression of Currarino syndrome.

Case Report

A 14-month-old male child, who was born with absent anal opening, underwent high divided sigmoid colostomy at day 2 of life and was readmitted for definitive procedure, i.e., posterior sagittal anorectoplasty (PSARP). On admission, the patient was evaluated with distal colostogram which was suggestive of high ARM with recto-urethral fistula which was 1 mm wide and 4.2 mm long [Figure 1]. In view of high ARM and longer pouch perineum distance as well as for assessment of pelvic floor musculature, pelvic MRI was advised. Pelvic MRI along with screening of the whole spine was done with 3 Tesla MRI scanner. There was presence of blind-ending rectal pouch terminating at the level of the pubococcygeal line. There was anterior protrusion of ischiorectal fossa fat compressing the anorectal pouch anteriorly, which was giving a false impression of some presacral mass, hence confusion with Currarino syndrome [Figure 2]. Further, there was finding of a tiny tract extending from the distal rectum towards distal prostatic urethra suggestive of recto-urethral fistula. The levator ani muscle was thinned out and the external anal sphincter was not visualised. On screening of the spine, the conus was found to be terminating at second lumbar level. There was no tethering of the conus. Distal coccygeal vertebra appeared to be deformed and displaced towards the right side.

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To confirm the opening of fistula in the urethra, cystourethroscopy was performed in which fistula opening was identified just distal to verumontanum at 6 O’clock. The finding of fat protrusion in pelvic MRI caused confusion immediately before surgery. There was a doubt of Currarino syndrome, so approach for the surgery needed to be revised. However, after clinical discussion, it was decided to proceed with PSARP. After catheterisation and positioning the patient in prone jackknife position, painting and draping was done. Anterior and posterior limits of sphincter muscle complex were identified with the help of electrocautery. Midline posterior sagittal incision was given extending from the tip of the coccyx till the anterior limit of anal dimple. The incision was deepened in midline with subsequent visualisation of parasagittal muscle fibres and sphincter muscle complex, preserving the limit of the ischiorectal fossa. The coccyx was identified and a plane was created just anterior to the coccyx with blunt dissection. Subsequently, blind-ending rectal pouch was identified. The fat in the right ischiorectal fossa was seen bulging towards the midline [Figure 3a], correlating with the finding of pelvic MRI. However, there was no obvious presacral mass lesion, thereby ruling out Currarino syndrome. The rectal pouch was further dissected laterally, posteriorly and medially to identify the recto-urethral fistula [Figure 3b]. The fistula was then divided and closed with 5-0 absorbable suture in interrupted fashion. The rectal pouch was further dissected to gain sufficient length and anorectoplasty was performed in standard manner. The prolapsing ischiorectal fat was left intact. Post-operatively, the patient was started on oral diet on day 2 of surgery. Foley’s catheter was removed on day 7 of surgery, following which the patient voided in good stream without difficulty. Subsequently, the patient was discharged and is being planned for restoration of bowel continuity in the next follow-up 6 weeks later.

**Discussion**

Routine imaging protocol of ARM includes distal colostogram, radiograph of the pelvis and spine, ultrasound of the kidneys, bladder and spine and voiding cystourethrogram. However, none of these investigations provide accurate information about pelvic floor musculature and external anal sphincter which are important for achieving faecal continence. Ultrasonography of the perineum can provide information about distal rectal pouch and pouch perineum distance, but it is difficult to identify pelvic musculature and sphincter muscle complex. Similarly, pressure-augmented distal colostogram is routinely used to identify the distal limit of rectal pouch as well as recto-urethral fistula if present.

Recently, pelvic MRI is being used more frequently in children with ARM. It helps in evaluation of the actual position of rectal pouch. It also provides detailed information about the size and development of pelvic floor musculature. Moreover, it also helps in identifying any spinal dysraphism or tethering of cord which further affects bowel and bladder control.

In the index case, we performed MRI to have better idea of pelvic floor anatomy as well as actual position of rectal pouch. The patient was posted for surgery, i.e. PSARP, after receiving the MRI report. However, there was confusion due to abnormal fat protrusion. We proceeded with PSARP, but
we could not see any abnormal mass. There was an abnormal bulge of fat through the right ischiorectal fossa which was correlating with MRI finding. One possible explanation of this abnormal protrusion of fat is weak pelvic floor musculature due to high ARM.

In cases of high ARM, a detailed study of pelvic floor anatomy is warranted. An assessment of pelvic floor muscles, particularly levator ani and external anal sphincter, helps us in prognosticating these babies for faecal continence after PSARP.[7] As pelvic MRI can provide us with this information, it should be included in imaging protocol for evaluation of high ARM along with other existing tools. At the same time, a detailed knowledge of pelvic floor anatomy is mandatory to correctly interpret MRI to avoid any misinterpretation.

Conclusions
Pelvic MRI is an emerging tool for evaluating a case of ARM. It is an excellent imaging modality for assessing pelvic muscles and sphincter complex as well as location of blind-ending rectal pouch. However, a detailed knowledge of pelvic floor anatomy is mandatory to avoid wrong interpretation and misdiagnosis.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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