High resolution MRI of anorectal malformation in the newborn: case reports of Currarino syndrome and anocutaneous fistula

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Received: 26 March 2001/Accepted: 2 May 2001

Abstract
To show the accuracy of a high-resolution magnetic resonance imaging (MRI) technique with a phased array coil in diagnosing anorectal malformation, we present two neonates with Currarino syndrome and anocutaneous fistula, respectively. Anatomy was visualized correctly with this technique, but conventional MRI did not show the complete extent of the disease. The reported high-resolution MRI findings concerning these conditions are scanty.

Key words: Currarino syndrome—Magnetic resonance imaging—High resolution—Anorectal malformation—Fistula—Teratoma.

Magnetic resonance imaging (MRI) is the most accurate imaging modality for the classification of anorectal malformation because of its excellent soft tissue contrast resolution and multiplanar imaging capability [1]. The role of imaging in the diagnosis of Currarino syndrome has been extensively described, but information concerning the MRI features of this relatively rare hereditary syndrome, which consists of anorectal malformation or constipation, sacral defect, and presacral mass, is scanty [2–7]. Most anorectal malformations show a communicating tract between the rectum or anal canal and the genitourinary tract or the perineum, called a fistula or ectopic anal orifice [1]. MRI is unreliable in detecting these fistulas because they are often too small to visualize [8]. To demonstrate the accuracy of a high-resolution MRI technique (HR-MRI) in comparison with a conventional MRI technique, we describe two patients, one with Currarino syndrome and another with anocutaneous fistula, in whom the anatomy was correctly diagnosed with this HR-MRI technique.

Case reports

Case 1

A 3-day-old girl presented with rectal blood loss and symptoms of bowel obstruction. Her family history showed a sister with Currarino syndrome. Physical examination showed no abnormalities. Digital rectal examination could not be performed. A plain X-ray of the sacral bone showed a bony defect. To confirm the diagnosis of Currarino syndrome, abdominal ultrasound and conventional MRI of the pelvis and spine were performed. The ultrasound study was normal. The conventional MRI confirmed the sacral bone abnormality and visualized an anterior meningocele, but no other presacral mass was seen (Fig. 1). Associated spinal cord anomalies such as tethered cord were excluded in the same session. The patient underwent a second MRI examination on the same day with a HR-MRI technique to better visualize the presacral mass. HR-MRI with a phased array coil showed a meningocele and a presacral mass with a heterogeneous signal intensity on T1- and T2-weighted sequences suggestive of a teratoma (Fig. 2). HR-MRI also visualized a normal position and aspect of the anal sphincter. The patient underwent surgical resection of the presacral heterogeneous mass. The anterior meningocele was left in situ. Pathology confirmed the diagnosis of teratoma. Despite symptoms of bowel obstruction, no anal or rectal stenosis was found during digital examination under general anesthesia.
Case 2

A 1-day-old newborn boy presented with an anorectal malformation. The anal canal was imperforated and there was a fistula opening into the perineum anterior to the normal position of the anal verge. HR-MRI with the same technique used in case 1 visualized a low malformation with a normal position and aspect of the anal sphincter muscles, with an anocutaneous fistula running just anterior in the external sphincter complex (Fig. 3). There were no abnormalities in the spine and presacral region. Because of these MR findings, a simple dilatation of the fistula rather than reconstructive surgery was performed.

Discussion

Currarino syndrome can be associated with a spectrum of abnormalities such as malformation of the spinal cord and genitourinary tract [4, 5]. Early diagnosis is necessary for its surgical treatment and to prevent complications such as meningitis, neurologic impairment, and malignant degeneration of a presacral teratoma [3, 9]. Accurate imaging of the complex anomalies of this syndrome is essential for the exact visualization of the abnormalities. Conventional radiography can show a scimitar bony sacral defect, which is highly suggestive for Currarino syndrome and requires further investigation. However, the sacral defect may not always be present [9], as one or two characteristics of the triad may be missing [10]. Barium enema studies may show anorectal malformation but do not directly visualize the pelvic floor, anal sphincter anatomy, and, possibly, the tumor. Although ultrasound can accurately exclude associated renal anomalies, it is less reliable in demonstrating the level of

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Fig. 1. Case 1. Sagittal T2-weighted turbo spin-echo MR image using a conventional low-resolution MRI technique with a body coil (repetition time/echo time = 3039/120 ms, field of view = 90 mm, slice thickness = 3 mm, 256 × 163 matrix, Number of signal averages (NSA) = 8; 0.5-T Gyroscan T5-II, Philips Medical Systems, Best, The Netherlands) showed a defect in the sacral bone with an anterior meningocele (arrows) but missed a small heterogeneous presacral mass.

Fig. 2. Case 1. A Sagittal T2-weighted turbo spin-echo MR image using an HR-MRI technique with a quadrature phased array coil (repetition time/echo time = 4500/126 ms, turbo factor = 16, field of view = 100 mm, slice thickness = 3 mm, 512 × 128 matrix, NSA = 10, voxel size = 0.5 mm³; 1.5-T Gyroscan NT, Philips Medical Systems, Best, The Netherlands) shows a presacral mass smaller than 20 mm with a heterogeneous signal intensity (short arrow), a sacral defect, and an anterior meningocele (long arrow). B Transverse T2-weighted turbo spin-echo MR image using the same technique as in A shows a sacral defect, an anterior meningocele (long arrows), and a presacral heterogeneous mass attached to the meningocele (short arrows).