



Discover Generics

Cost-Effective CT & MRI Contrast Agents



[VIEW CATALOG](#)

AJNR

Retrorectal cyst-hamartomas and sacral dysplasia: MR appearance.

L S Williams, A M Rojiani, R G Quisling and J P Mickle

AJNR Am J Neuroradiol 1998, 19 (6) 1043-1045

<http://www.ajnr.org/content/19/6/1043>

This information is current as
of September 3, 2025.

Retrorectal Cyst-Hamartomas and Sacral Dysplasia: MR Appearance

Lorna Sohn Williams, Amyn M. Rojiani, Ronald G. Quisling, and J. Parker Mickle

Summary: Retrorectal cyst-hamartoma, an uncommon lesion, arises from hindgut embryonic remnants and may be associated with sacral anomalies. Such a lesion is presacral, multicystic, and lined with glandular or transitional epithelium. Malignant transformation of these lesions has been reported. We describe the clinical, pathologic, and imaging findings in an infant.

Retrorectal cyst-hamartomas, or tailgut cysts, are unusual lesions arising in the retrorectal, presacral space. They most likely originate from vestigial remnants of the embryonic hindgut (1). Although quite rare, retrorectal cyst-hamartoma is a known entity that has been described in the surgical and pathologic literature (1, 2). CT findings have been reported in the radiologic literature, but the MR imaging appearance has not been described (3, 4). We report the clinical features and MR imaging findings of a retrorectal cyst-hamartoma associated with sacral dysplasia.

Case Report

A 4-month-old boy was seen at an outside institution after several days of irritability and poor feeding. His birth and neonatal history were normal. External examination revealed a sacral dimple. A lumbar puncture showed pleocytosis (WBC count of 710 mm^3 , 90% polymorphonuclear leukocytes) and Gram stain of the CSF was positive for gram-negative coccobacilli, which was cultured and identified as *Proteus mirabilis*. The patient was given intravenous antibiotics (ampicillin, gentamicin, and ceftriaxone sodium) and transferred to our institution, where neuroimaging studies were performed to assess a possible cord abnormality.

MR images revealed a well-circumscribed, thick-walled, $2.0 \times 2.5 \times 2.0$ -cm multicystic mass posterior to the rectum and anterior to the sacrum. Heterogeneously low signal was seen on T1-weighted sequences (600/15/5 [TR/TE/excitations]) and high signal was noted on T2-weighted sequences (6500/21/2) (Fig 1A and B). The mass did not enhance. The lesion appeared contiguous with but did not obviously invade the rectum. The surrounding soft-tissue planes appeared intact. A tethered cord was seen at the level of L3. The lumbar vertebral bodies were unremarkable. All levels of the sacrum below the S1 level were dysplastic.

A lumbar myelogram was performed at the L4–L5 level using a 25-gauge spinal needle and 2 mL of intravenous, non-ionic contrast material (Omnipaque 180) to evaluate for possible subarachnoid communication, given the patient's history of meningitis. The myelogram did not show any obvious communication between the thecal space and the rectum or surrounding soft tissues. Postmyelographic CT scans of the entire pelvis, obtained with contiguous 5-mm-thick sections, showed a presacral mass similar in configuration to that seen on the MR images. The mass was of mixed CT density, consistent with both fluid and soft-tissue attenuation. No calcification or myelographic contrast was apparent within the lesion.

At surgery, densely scarred epidural tissue was identified at S1 through a laminectomy to the level of L3. When the dura was opened, a low-lying spinal cord with matted cauda equina was identified. In the distal conus and cauda equina, a 2.0×3.0 -cm cystic mass was seen densely adherent to the neural structures. This cyst insinuated itself ventrally into the retrorectal space, where it was also tightly adherent to the rectum and surrounded by fibrotic-appearing tissue. The cyst was easily removed, the filum terminale was identified and sectioned, and the wound was then closed.

The specimen, received in multiple fragments, consisted of soft tissue with multiple cysts. The epithelial lining of these cysts varied from stratified squamous to transitional (cloacogenic) epithelium to columnar epithelium, ciliated or pseudostratified with rare foci of mucin-producing cells. Individual cysts showed multiple epithelial types within the same lining. Smooth muscle was present in the adjacent soft tissue and was also seen in close relationship to the cyst wall, following its contour (Fig 1C and D). These findings are typical of retrorectal cyst-hamartoma.

The patient was discharged on the fifth postoperative day and remains well 4 months after surgery.

Discussion

Retrorectal cyst-hamartomas are thought to be congenital lesions originating from the embryonic hindgut. At 35 days' gestation, the most caudal portion of the hindgut develops a true tail (1). The primitive gut extending into the hindgut has been called the tail gut or postanal gut, and it normally regresses by day 56. Retrorectal cyst-hamartomas are believed to be vestigial remnants of the tail gut (1–4). A cyst occurring in this region is classified by its relation to the sacrum, dorsal being posterior to the sacrum and ventral being anterior (3). Retrorectal

Received June 27, 1997; accepted after revision January 2, 1998.

From the Departments of Radiology (L.S.W., R.G.Q.), Pathology (Neuropathology) (A.M.R.), and Neurological Surgery (J.P.M.), University of Florida College of Medicine and Brain Institute, Gainesville.

Address reprint requests to Amyn M. Rojiani, MD, PhD, Department of Pathology (Neuropathology), University of South Florida, 12901 Bruce B. Downs Blvd, MDCII, Tampa, FL 33612.

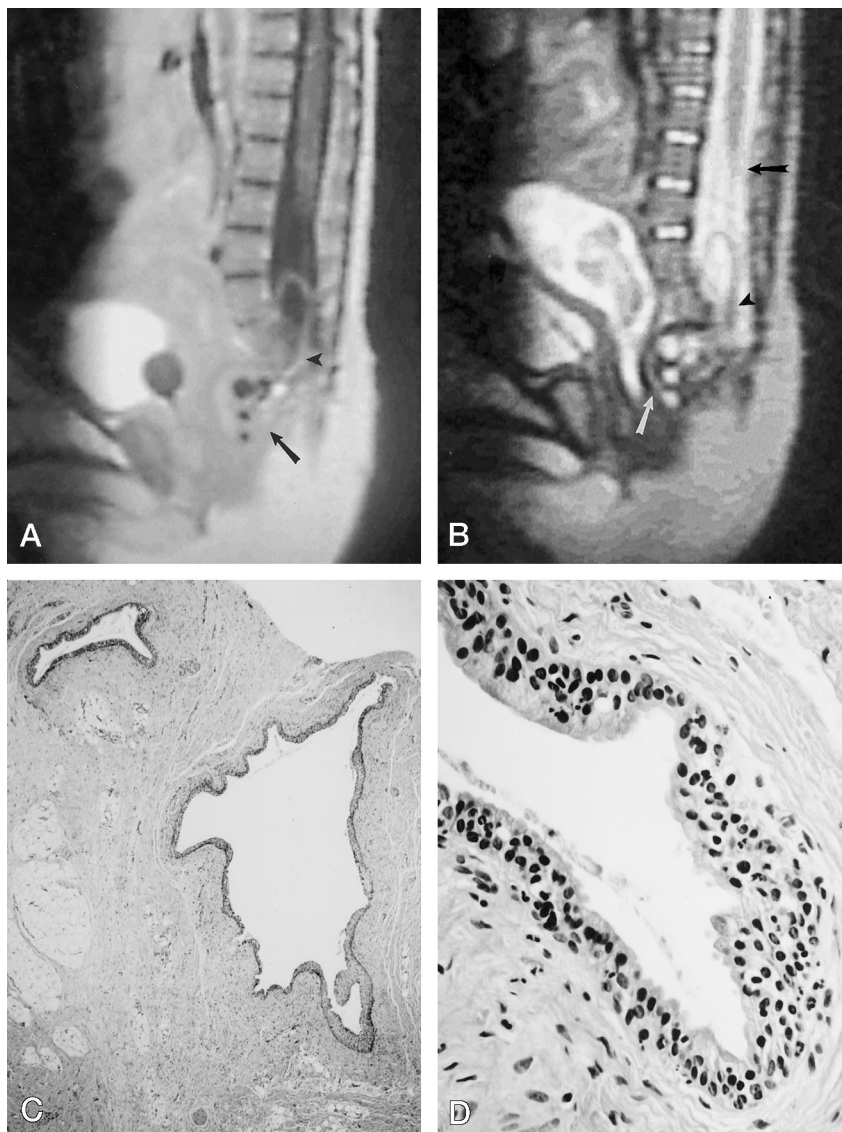
FIG 1. 4-month-old boy with retrorectal cyst-hamartoma, which lies in the presacral space and is associated with sacral dysplasia.

A, Postcontrast sagittal T1-weighted MR image (600/15) shows T1 shortening within the mass (arrow) corresponding to fatty elements adherent to the cauda equina (arrowhead). The mass does not enhance.

B, Sagittal T2-weighted fast spin-echo MR image (4200/91) shows a tethered cord at mid L3 (black arrow) within an abnormally dilated thecal sac. The anterior portion of the mass is thick-walled and contiguous with the posterior rectal wall (white arrow). Segments of the cauda equina appear adherent to the mass (arrowhead).

C, Low-magnification view of the resected mass shows multiple cysts lined primarily by stratified epithelium. Note the adjacent smooth muscle bundles, some of which follow the contours of the cyst. Hematoxylin-eosin, original magnification $\times 4$.

D, Higher magnification view shows presence of stratified squamous and cloacogenic epithelium. Hematoxylin-eosin, original magnification $\times 20$.



cyst-hamartomas, or ventral cysts, lie in the presacral space, anterior to the sacrum and posterior to the rectum.

Various names have been used to describe this entity, such as cyst of the postanal intestine, myoepithelial hamartoma of the rectum, mucin-secreting developmental cyst, tailgut vestige, and rectal cyst (3). Recently, Hjermsstad and Helwig (1) have suggested the term tailgut cyst because of its unambiguity.

The gross appearance of a retrorectal cyst-hamartoma is that of a soft, thick-walled, mucoid-filled mass (1). If secondarily infected, the lesion may be thick-walled with surrounding inflammatory changes and fibrosis (5). Histologically, various epithelial cell types line the cyst, but identification of transitional or glandular-type epithelium with or without stratified squamous components is essential to make the diagnosis. The other histologic requisites for the diagnosis are absence of a well-defined muscular coat containing a myenteric plexus, and serosa. These criteria differentiate retrorectal cysts from epidermoid (stratified squamous epithelium only), dermoid (stratified squa-

mous epithelium with skin adnexal structures), and duplication cysts (well-defined intestinal mucosa with all components including villi and crypts).

In their review of 53 cases, Hjermsstad and Helwig (1) found the average diameter of the cysts to be 3.9 cm, with half multicystic and the other half unilocular. In these patients, almost 50% were symptomatic, with an average duration of symptoms of 7.5 months. In the asymptomatic patients, the lesions were discovered during routine physical or pelvic examinations (4). These lesions have been reported in infants and adults with a 3:1 female to male ratio (4, 5). Most of the lesions have occurred in adult women (4).

Sacral abnormalities reported in association with retrorectal cyst-hamartomas include sacral or coccygeal agenesis and various sacrococcygeal defects (1, 6). Symptoms often result from infection; less often, mass effect causes rectal pain or pressure (5). A draining sinus to the perianal skin or rectal wall is a reported complication, and sinography (injection of contrast agent into the sinus tract) reportedly was used as a means of diagnosis in some earlier cases (7).

Current methods of detection and surgical planning rely primarily on cross-sectional imaging (1).

The primary differential diagnosis of these presacral cystic masses is teratoma, dermoid cyst, duplication cyst of the rectum, anal gland cyst, and neurenteric cyst (2, 8). Teratomas, which have derivatives of all three germ layers, may also contain fat or calcification visible on cross-sectional imaging (3, 4). Dermoid cysts, formed by ectodermal invagination, may contain skin adnexa and hair follicles, and can be grossly similar to retrorectal cyst-hamartomas (3). Differentiating among these possibilities may only be possible by histologic evidence of a lining containing solely stratified squamous epithelium (3). Rectal duplication cysts are lined by squamous or intestinal-type epithelium with a well-developed smooth muscle wall, myenteric plexus, and serosa; they are typically unilocular, whereas retrorectal cyst-hamartomas are multilocular about half the time (1, 2). Anal gland cysts are typically located near the anal sphincter, more cephalad and posterior to the expected location of retrorectal cyst-hamartomas, and have a stratified squamous epithelial lining (2). Other entities to consider are anterior meningocele, rectal leiomyosarcoma, and chordoma. An anterior meningocele would be seen in association with a dystrophic spine and tethered cord, and a myelogram would be diagnostic. A rectal leiomyosarcoma is a more aggressive lesion with rectal invasion and mucosal involvement (7). A chordoma would also be expected to show evidence of local bone destruction or invasion (4). A neurenteric cyst may communicate with the subarachnoid space; it is typically unilocular and has a thin wall, unlike the retrorectal cyst-hamartoma (1, 8). Neurenteric cysts also differ histopathologically from retrorectal cyst-hamartomas in that they contain villi, a well-defined lamina propria, and more mature mucosa (1).

The recommended surgical management of these benign masses is complete excision with removal of the coccyx to reduce the possibility of any residual

lesion or recurrence (1, 2, 8). Complete excision is stressed for two reasons. First, recurrent retrorectal cyst-hamartomas may be complicated by chronic infection with possible abscess and fistula formation (2). Second, these entities have been known to undergo malignant transformation into adenocarcinoma or squamous carcinoma (1, 2, 3, 8). Incisional biopsy is not encouraged owing to the limited amount of diagnostic tissue available and because a follow-up excision is necessary (1). A planned total excision based on preoperative imaging findings is advocated (1). Digital rectal examination every 3 months during the first postoperative year and then yearly is recommended (3). The postoperative imaging recommendations are a baseline CT study at 1 month and then additional imaging if the physical examination so indicates (3).

Conclusion

We have outlined the clinical, pathologic, and neuroimaging features of a case of retrorectal cyst-hamartoma in an infant, highlighting the MR imaging characteristics.

References

1. Hjerstad B, Helwig E. **Tailgut cyst: a report of 53 cases.** *Am J Clin Pathol* 1988;89:139-147
2. Marco V, Autonell J, Farre J, Fernandez-Layos M, Doncel F. **Retrorectal cyst-hamartomas.** *Am J Surg Pathol* 1982;6:707-714
3. Ottery FD, Carlson RA, Gould H, Weese JL. **Retrorectal cyst-hamartomas: CT diagnosis.** *J Comput Assist Tomogr* 1986;10:260-263
4. Johnson A, Ros P, Hjerstad B. **Tailgut cyst: diagnosis with CT and sonography.** *AJR Am J Roentgenol* 1986;147:1309-1311
5. Pyo D. **Tailgut cyst (retrorectal cyst-hamartoma): report of a case and review.** *Mt Sinai J Med* 1990;57:249-252
6. Gerwig W. **Presacral cystic tumors.** *Ann Surg* 1954;140:81-85
7. Campbell W, Wolff M. **Retrorectal cysts of developmental origin.** *AJR Am J Roentgenol* 1977;117:307-312
8. Brooks BS, Duvall ER, El Gammal T, Gupta KL, Kapila A. **Neuroimaging features of neurenteric cysts: analysis of nine cases and review of the literature.** *AJNR Am J Neuroradiol* 1993;14:735-746