TAILGUT CYST PRESENTING AS GLUTEAL SWELLING: EXPLORING THE TIP OF AN ICEBERG

Sajid Ansari, Kaleem Ahmad, Mukesh Kumar Gupta, Ashok Raj Pant, Pranav Kumar Santhalia, Raj Kumar Rauniyar

Department of Radiodiagnosis and Imaging, B.P. Koirala Institute of Health Sciences, Dharan, Nepal.

ABSTRACT

Tail gut cysts or retorectal cystic hamartomas are rare developmental anomalies commonly located in the retrorectal space. Age of presentation ranged from 4 to 73 years with an average age of 35 years; with female predominance. Complete surgical excision is the treatment of choice. Hereby we report a case of 34 years old Asian female presented with swelling and pain in the left gluteal region since 7 months. Digital rectal examination revealed mild tenderness with palpable and fluctuant mass in the posterior aspect and local examination showed a soft swelling in the left gluteal region. Magnetic resonance imaging pelvis revealed a large well defined cystic lesion posterior to rectum and anterior to sacrum and coccyx, extending in the subcutaneous plane behind the coccyx; suggestive of retorectal cyst. Complete excision of the cyst and histopathological evaluation revealed tail gut cyst.

Keywords: Tail gut cyst, Retorectal cystic hamartomas, Developmental cyst, Radiological evaluation.

Introduction

Tail gut cysts or retorectal cystic hamartomas are rare congenital lesion derived from the remnants of the embryonic hindgut i.e. post-anal gut. They are located mostly in the retorectal space. Tail gut cysts are more common in females with female:male ratio of 3:1 and usually present in middle age; however it can be discovered at any age. Age of presentation ranged from 4 to 73 years with an average of 35 years. Magnetic resonance imaging (MRI) is the best imaging modality for diagnosis of tail gut cyst.

Case Presentation

A 34 years old Asian female presented with swelling and pain in the left gluteal region since 7 months.

Her bowel-bladder habits were normal. No history of rectal bleeding or change in stool color. There was no history of fever and her vital parameters were normal. On digital rectal examination, there was mild tenderness with palpable and fluctuant mass in the posterior aspect causing extrinsic compression of the rectum without any mucosal involvement. Local examination showed a soft swelling in the left gluteal region. Routine laboratory investigations and tumor markers were within normal limits. Ultrasonography of the left gluteal swelling (Fig.1) revealed a well defined cystic lesion with internal echoes which was extending into the pelvis. Pelvic ultrasonography revealed a large cystic lesion posterior to the rectum. Plain radiograph of lumbosacral spine showed deviation of coccyx towards right side.
Computed tomography (CT) of pelvis (Figure 3 and 4) also revealed the same findings without any calcification within the cyst or bone destruction. Diagnosis of retrorectal cyst was made on imaging. Complete surgical excision was done. Histopathology showed cyst wall lined with stratified squamous and cuboidal epithelium and scattered smooth muscle cells, suggestive of tailgut cyst. Post-operative period was uneventful. On follow-up after 3 months, there was no recurrence of the cyst. Post-operative period was uneventful and the patient was symptom free on three months of follow-up.
Embryologically, tail gut cysts are believed to arise from vestigial remnants of the embryonic hindgut. The tail gut or postanal gut is the most caudal part of the hindgut, distal to the future anus. It normally involutes by the 8th week of embryonic development and if a tail gut remnant persists, it may give rise to a tail gut cyst. The differential diagnoses of lesions in the presacral space are divided into congenital, neurogenic, osseous, inflammatory and miscellaneous. Congenital lesions account for nearly two-thirds of retrorectal lesions. Congenital lesions include tail gut cysts, enteric duplication cysts, dermoid cysts, epidermoid cysts, teratomas, chordomas and anterior sacral meningoceles. Retrorectal developmental cysts are distinguished by their characteristic features on histopathology. Both dermoid and epidermoid cysts are lined by stratified squamous epithelium; however, only dermoid cysts contain dermal appendages (like hair follicles, sweat glands, tooth, etc). Rectal duplication cysts are lined by typical gastrointestinal epithelium often with crypts, villi and glands and they are surrounded by two well-formed layers of smooth muscle with nerve plexuses.

Grossly, tail gut cyst is a multiloculated, cystic mass with a thin wall and is filled with a mucoid material. It usually measures several centimeters in diameter. Infection or inflammation may cause fibrosis of the cyst wall and breakdown of the cyst lining. Microscopically, it is characterized by the presence of a cyst lined with multiple, varying types of epithelium: columnar, cuboidal, squamous, mucin-secreting and transitional epithelium. In contrast to enteric duplication cysts, tail gut cysts have disorganized smooth muscle fibers within the cyst wall and do not contain neural plexuses.

Clinically, the patients present with low back pain or rectal pain, pain during defecation, rectal fullness, painless rectal bleeding, change in caliber of stool, urinary frequency, etc. In asymptomatic patients most lesions are detected incidentally. Almost all are palpable on rectal examination as extrinsic, contained fluctuant masses. The diagnosis is often delayed due to the rarity and unfamiliarity of this entity and because the symptoms often mimic other more commonly occurring pathologies at this site. Plain radiography, colonoscopy, barium enema, transrectal ultrasonography and CT may
be useful; but MRI has become the modality of choice. MRI typically demonstrates a rectal lesion appearing hypointense on T1-weighted and hyperintense on T2-weighted images; although it may vary according to the content of the lesion.\textsuperscript{8,12-14} Heterogeneous appearance of the cyst on MRI may be seen owing to mucin, proteinaceous material or hemorrhage within the cyst.\textsuperscript{13,14} Kim et al has described the multilocular appearance with internal septae on T2-weighted images as being specific for tailgut cysts.\textsuperscript{13} Yang et al\textsuperscript{8} has described the presence of a small peripheral cyst accompanying the larger cyst. A mass effect in large lesions has been described, with displacement of the adjacent rectum, as was observed in our patient. Malignant transformation has rarely been reported, in which case adenocarcinoma, carcinoma, neuroendocrine carcinoma and sarcoma arise within the cyst.\textsuperscript{11,14} Focal irregular wall thickening and intermediate signal intensity on pre-contrast T1 and T2 weighted images with enhancement on contrast study, suggests malignancy.\textsuperscript{2,12} In suspected malignancy, preoperative biopsy is indicated. Complete surgical excision is recommended to avoid recurrence, alleviate symptoms, prevent infection and potential local dissemination of malignant cells. Different surgical approaches are there like posterior approach, abdominal approach or a combination of both. In recent studies, laparoscopic excision of the cyst is considered to be safe and effective approach.

Conclusion

Tailgut cyst is a rare developmental lesion and should be considered in the differential diagnosis in any case of rectal cyst, irrespective of age and sex. Radiological evaluation should be done in all patients with rectal cysts. MRI is the modality of choice for characterizing the lesion and assessing the extent of the lesion prior to surgery.

References


