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A Retrorectal Cystic Hamartoma (Tail Gut Cyst)

KEYWORDS: RETRO RECTAL, HAMARTOMA, TAILGUT, CYST.

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ABSTRACT *Retrorectal cystic hamartomas are rare congenital anomalies most commonly seen in a retrorectal area; most common in middle aged women. This article reviews about the radiological appearances of tail gut cyst in a 65-year-old female patient with post operative history for breast cancer. The patient had no fresh complaints and came for follow up. Tailgut cyst was found as an incidental finding in this patient.*

Introduction:

Tailgut cysts, also known as retrorectal cystic hamartomas, are rare congenital lesions thought to arise from the remnants of the embryonic postanal gut. They predominantly occur as retrorectal multicystic masses in women. Nearly half of the cases are asymptomatic and are frequently found on routine physical exam. When present, reported complaints include chronic perirectal pain and symptoms secondary to mass effect.

Case report:

65y old female with previous diagnosis of right sided breast cancer, the patient underwent mastectomy for the cancer one year back. The patient came to the radiology department for followup. Patient had no other fresh complaints.

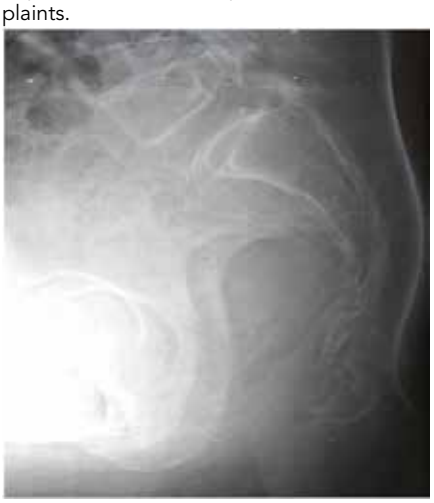

Fig A :X ray lateral view of the sacrum and coccyx showed posterior displacement of the lower coccygeal segments without any soft tissue swelling

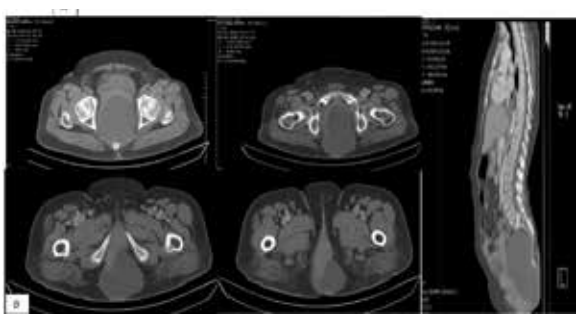

Fig B: Axial and sagittal CT images of a 65yrs old asymptomatic patient showing a well defined homogenous cystic density lesion in the retrorectal space.

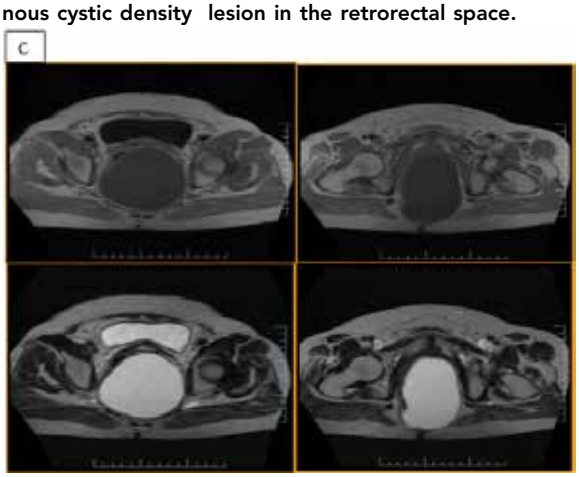

Fig C:AxialT1 and T2-weighted images of a 65yrs old asymptomatic patient showing a well defined homoge-

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nous cystic density lesion in the retrorectal space.

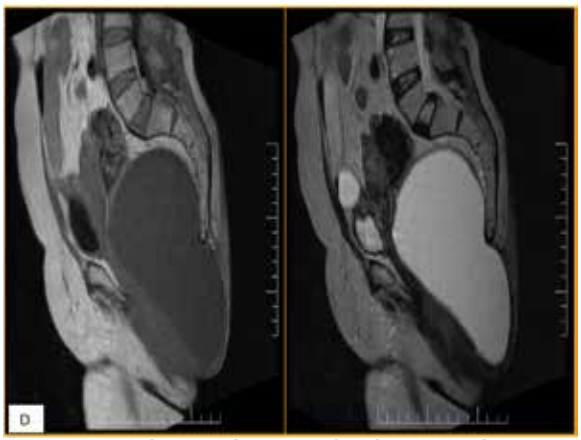

Fig D: Sagittal T1 and T2 weighted images showing a well defined homogenous cystic lesion in the presacral space.

Discussion :-

RETRORECTAL CYTS :-

Retrorectal cystic lesions in adults are rare, and most cases are congenital[1].Developmental cysts are the most common congenital entity encountered in the retrorectal space[2,3]. These include epidermoid cysts, dermoid cysts, and enteric cysts. There are two types of enteric cysts[4]:

- Tailgut cysts (retrorectal cyst-hamartomas)
- Cystic rectal duplication

Developmental cysts are defined by their Histologic components ,Retrorectal location, lying anterior to the sacrum and posterior to the rectum. They occur mostly in middle-aged women and in a 3:1 female-to-male ratioThe most important complications of these cysts are infection with a secondary fistula and malignant degeneration of enteric cysts[5,6].

TAILGUT CYST :-

Tailgut cyst or retrorectal cystic hamartoma is a rare congenital lesion thought to arise from vestiges of an embryonic hindgut and is found in the retrorectal or presacral space[7,8].The tailgut/postnatal gut normally involutes by the 8th week of embryonic development. If a tailgut remnant persists, it may give rise to a tailgut cyst.Tailgut cyst is more common in women and usually presents in middle age, but can present at any age.It is usually detected as an asymptomatic mass but may present with abdominal pain or constipation.

Grossly , tailgut cyst is a multiloculated, cystic mass with a thin wall and glistening lining , filled with a mucoid material.

Microscopy , shows a variety of epithelia without the presence of villi and crypts. An incomplete muscle layer is often associated with the cyst wall. Infection or inflammation may cause fibrosis of the cyst wall and breakdown of the cyst lining

The clinical presentation is variable, depending on the size and therefore mass effect of the developmental cyst and the presence of infection in the cyst [5,9]. It has been estimated that 50% of developmental cysts are asymptomatic [10], being discovered during routine physical examination or ultrasonography (US). Physical examination can demonstrate a funnel-shaped dimple in the postanal midline or a chronic fistula that may communicate with the cyst, involving the perianal skin posterior to the anus or the anal canal in the midline. Digital rectal examination may reveal a smooth, firm mass in the retrorectal space, bulging into the rectal lumen[4,5,9].

Symptoms are often related to local compression on the rectum, which causes constipation, rectal fullness, painful defecation, and lower abdominal pain, and to local compression on the lower urinary tract, which causes dysuria and urinary frequency [1,4,9]. Dystocia and sciatic pain have been reported (14). Developmental cysts are commonly revealed by complications such as local infection with a chronic perianal fistula or rectal bleeding [1,9].

On imaging , ultrasound is of limited value, showing only a multilocular cyst with debris and gelatinous material seen as internal echoes. On CT it appears as a discrete, well-marginated mass of water or soft-tissue density. Calcification of the thin wall may rarely be seen[10] . In case of infection or rarely malignant transformation, loss of discrete margins and involvement of adjacent structures may be seen. On MRI tailgut cyst is typically hypo intense on T1W and hyper intense on T2W images, a heterogeneous appearance on MRI may be seen owing to mucin, proteinaceous material, or hemorrhage within the cyst.

Complications

Infection :- the most frequent complication. The infection may manifest as pelvic pain, a local abscess, and a secondary perianal or anorectal fistula with discharge of pus. A communication or primary fistula may exist between the cyst and the anorectal lumen without infection[1,5,9].

Malignant degeneration :- Malignant degeneration has been reported in enteric cysts[6,7,11,12,] and results in adenocarcinoma or squamous carcinoma. It has been estimated that malignant degeneration occurs in 7% of enteric cysts[13].

DD's :-

Sacrococcygeal Teratoma :

Sacrococcygeal teratomas are germ cell tumors containing elements derived from all three germ layers. Sacrococcygeal teratoma is the most frequently encountered presacral lesion in the pediatric age group, and most (90%) are diagnosed in the newborn period and are benign. The prevalence of malignancy increases with age; however, they are rarely discovered in adult life. They are found more frequently in females [4,14,15]. At CT or MR imaging, sacrococcygeal teratomas appear as heterogeneous, well-defined lesions with mixed cystic and solid components. Uncommonly, sacrococcygeal teratomas are entirely cystic, and these are more likely to be benign. Sacrococcygeal teratomas contain fat or calcifications in 50% of cases.

Anterior Sacral Meningocele:

Anterior sacral meningocele is a rare congenital disorder. Anterior sacral meningocele is defined as a meningeal cyst that occurs in the presacral space secondary to agenesis of a portion of the anterior sacrum. In approximately 50% of cases, associated malformations are found, such as spina bifida, spinal dysraphism, bicornuate uterus, and imperforate anus. Plain radiography shows a typical scimitar-shaped sacral bone defect in 50% of cases[16,17]. CT and MR imaging demonstrate the sacral defect in association with a well-defined, unilocular, fluid-filled lesion in the retrorectal space. MR imaging may demonstrate the stalk of the

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meningocele, which communicates with the thecal sac.

Anal Duct or Gland Cyst:
At histopathologic analysis, anal duct or gland cysts are mucus-secreting cysts lined with a combination of stratified squamous epithelium, columnar epithelium, and transitional epithelium. They often communicate with an anal duct or crypt. CT and MR imaging demonstrate a uni- or multilocular cystic lesion near the anal sphincter in the retrorectal space. Anal duct or gland cysts may involve the coccyx and the ischioanal space[18].

Rectal Leiomyosarcoma:
Rectal leiomyosarcomas (malignant rectal stromal tumors) are rare, accounting for less than 0.1% of all malignant tumors of the rectum. Rectal leiomyosarcomas are malignant tumors of smooth muscle. Most rectal leiomyosarcomas occur in the lower third of the rectum, and they are more common in men.

The spread of these tumors is principally local and hematogenous. They may involve the rectal mucosa, leading to surface ulceration and bleeding[19]. CT shows a well-circumscribed, heterogeneously enhanced, multilobulated tumor with cystic necrotic components

Treatment :-
The recommended treatment for developmental cysts is complete surgical excision of the epithelial lining of the cyst because of the risks of recurrence, malignant degeneration, and chronic infection [5]. If malignant degeneration is suspected, total excision including the normal rectum may be necessary.

Conclusion :-
A wide variety of cystic lesions occur in the retrorectal space, and most are congenital. Developmental cysts are the most common lesions that represent a real radiologic-histopathologic entity, resulting in a diagnostic dilemma. Imaging may show specific signs, but the diagnosis remains histopathologic. The surgical approach is the rule to accurately establish the diagnosis and avoid complications.

References :-

1. Leborgne J, Guiberteau B, Lehur PA, Le Goff M, Le Néel JC, Nomballais MF. *Les tumeurs kystiques vestigiales rétrorectales de l'adulte. Chirurgie* 1989; 115:565-571.
2. Jao SW, Beart RW, Spencer RJ, Reiman HM, Ilstrup DM. *Retro-rectal tumors: Mayo Clinic experience 1960–1979. Dis Colon Rectum* 1985; 28:644-652.
3. Uhlig BE, Johnson RL. *Presacral tumors and cysts in adults. Dis Colon Rectum* 1975; 18:581-596.
4. Levine E, Batnitzky S. *Computed tomography of sacral and perisacral lesions. Crit Rev Diagn Imaging* 1984; 21:307-374.
5. Williams LS, Rojiani AM, Quisling RG, Mickle JP. Retrorectal cyst-hamartomas and sacral dysplasia: MR appearance. AJNR Am J Neuroradiol 1998; 19:1043-1045.
6. Lim KE, Hsu WC, Wang CR. Tailgut cyst with malignancy: MR imaging findings. AJR Am J Roentgenol 1998; 170:1488-1490.
7. Marco V, Fernandez- Autonell J, Doncel F, Farre J. Retrorectalcyst-hamartomas: report of two cases with adenocarcinomas developing in one. *Am J Surg Pathol* 1982;6:707–714
8. Mills SE, Walker AN, Stallings RG, Allen MS. Retrorectal cystic hamartoma:report of three cases, including one with a perirectal component.
9. La Quaglia MP, Feins N, Eraklis A, Hendren WH. *Rectal duplications. J Pediatr Surg* 1990; 25:980-984.
10. Hjermstad BM, Helwig EB. *Tailgut cysts: report of 53 cases. Am J Clin Pathol* 1988; 89:139-147.
11. Johnson AL, Ros PR, Hjermstad BM. *Tailgut cysts: diagnosis with CT and sonography. AJR Am J Roentgenol* 1986; 147:1309-1311.
12. Crowley LV, Page HG. *Adenocarcinoma arising in presacral enterogenouscyst. Arch Pathol* 1960; 69:64-66.
13. Abel ME, Nelson R, Prasad ML, Pearl RK, Orsay CP, Abcarian H. Parasacrococcygeal approach for the resection of retrorectal developmental cysts. Dis Colon Rectum 1985; 28:855-858.
14. Winderl LM, Silverman RK. Prenatal identification of a completely cystic internal sacrococcygeal teratoma (type IV). Ultrasound Obstet Gynecol 1997; 9:425-428.
15. Wetzel LH, Levine E. MR imaging of sacral and presacral lesions. AJR Am J Roentgenol 1990; 154:771-775.
16. O'Riordain DS, O'Connell PR, Kirwan WO. Hereditary sacral agenesiswith presacral mass and anorectal stenosis: the Currarino triad. Br J Surg 1991; 78:536-538.
17. Thomas M, Halaby FA, Hirschauer JS. Hereditary occurrence of anteriorHalaby FA, Hirschauer JS. Hereditary occurrence of anterior sacral meningocele: report of ten cases. Spine 1987; 12:351-354.
18. Kulaylat MN, Doerr RJ, Neuwirth M, Satchidanand SK. Anal duct/glandcyst: report of a case and review of the literature. Dis Colon Rectum 1998; 41:103-110.
19. Wang TK, Chung MT. Anorectal leiomyosarcomas. J Gastroenterol 1998; 33:402-407.