

Retrorectal epidermoid cyst – experience of four cases.

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Abstract: Four cases of retrorectal cysts are presented. Two cases presented as bulge perineum, one obstructive rectal symptoms and one as a natal cleft sinus. Diagnosis was established by M R I Scan. Surgical treatment offered was simple excision in first case, while three cases required en-bloc coccygectomy, all the cases were done by posterior approach.

The last case which presented with natal cleft sinus, in view of recurrence after two operations at primary care center, M R I was performed registering well defined cystic lesion in left perianal region situated immediately beneath tip of coccyx. At operation methylene blue injected in natal cleft sinus went in easily without any resistance with no escape of dye from any area. Complete excision of retrorectal cyst with communicating track done by posterior approach en-bloc coccygectomy.

Key words: developmental cyst, retrorectal epidermoid cyst, retrorectal space, retrorectal lesions, natal cleft sinus

I. Introduction

Retrorectal epidermoid cyst is one of the developmental cysts which arise from remnants of embryonic tissue. Because of rarity of these tumors in adults, diagnosis is usually difficult before surgery¹. Further hidden in the hollow of the sacrum, it is unlikely to be discovered unless a sinus communicating with the exterior is present or it develops as a result of inflammation².

II. Clinical Material

Four cases of retrorectal cysts were operated by the author, all four cases were diagnosed by M R I scan. In three cases symptoms were due to large size of the lesion – swelling perineum in two and pressure on rectum leading to constipation in one. In both perineal swelling presentation cases per-rectal examination was unremarkable, while in pressure on rectum presentation case per-rectal examination was feeling a smooth soft extraneous swelling bulge in posterior wall.

The fourth case a 22year old woman presented with a natal cleft sinus discharging purulent material off and on for the past one year. Six months back she was diagnosed as a case of pilonidal sinus at primary care center and operated twice but resulted in recurrence of sinus. When presented to us there was a 3mmx3mm sinus in middle natal cleft area. A 3.5cm long vertical scar was appreciated separately just cranial to the sinus opening suggestive of prior intervention (Photo 1). Per-rectal examination was unremarkable.

In view of recurrence in fourth case, M R I scan done, which revealed well defined cystic lesion in left perineal region measuring 27mmx22mm, situated immediately beneath tip of coccyx –suggestive of benign lesion (fig 1). The patient was operated in the prone jack-knife position. At start of operation methylene blue dye 10cc injected in sinus opening, it went easily without any resistance or leakage of dye anywhere. Sinus track lifted off from sacral fascia, it was going down so decision taken for doing coccygectomy (Photo 2&3) and then further dissection of swelling was done from behind forward and the whole track was excised in continuum with the cyst (Photo 4). Wound closure was done by double Z-plasty (Photo 5).

In all the four cases post operative period was uneventful and histopathology reported as epidermoid cyst. In fourth case sections from sinus track showed hyperkeratotic stratified epithelium showing pseudoepitheliomatous hyperplasia and ulceration.

III. Comments

Retrorectal tumors are the heterogenous lesions in the space in front of the lower part of the sacrum and coccyx. The world wide accepted classification of these lesions is that proposed by Uhlig and Johnson³ stating major groups as congenital, inflammatory, osseous and miscellaneous.

Incidence : The Mayo clinic estimates the incidence of retrorectal tumors to be about 1 in 40,000 hospital admissions. They reported 120 cases of which 66% congenital, 12% neurogenic, 11% osseous and 11% were miscellaneous. Metastatic and inflammatory masses were excluded from this report⁴. Sacrococcygeal teratoma is the most common retrorectal tumor in the pediatric population and occurs in 1 in 40,000 births⁵.

Congenital lesions accounts for more than 50% of all retrorectal tumors and about two thirds of them are developmental cysts. The majority of developmental cysts are asymptomatic as they remain hidden in the hollow of the sacrum and are missed even on rectal examination due to low tension in the cyst. In 3 of our four cases rectal examination was unremarkable. However in situation of becoming infected cyst may present with a sinus communicating with the exterior. When a discharging sinus is present , a postanal dermoid will probably be mistaken for a pilonidal sinus or even anal fistula². One of our patient presented as discharging natal cleft sinus and was operated twice at primary care center suspecting it to be pilonidal sinus.

Epidermoid and dermoid cysts results from defective closure of the ectodermal tube, which results in inclusion of skin with or without accessory appendages. Both are lined with stratified squamous epithelium, well circumscribed with thin layer of connective tissue and filled with thick yellow green fluid. The difference is that epidermoid cysts have no skin appendages whereas dermoid cysts contain them¹.

Clinical manifestations : clinical symptoms are variable and are determined by the volume of the cyst, its mass effect or the presence or absence of infection. It is estimated that about 50% of these cysts are completely asymptomatic⁶.

Imaging : simple abdominal radiography may reveal various sacrococcygians bone lesions that often evolve together with developing cysts. Retrorectal space widening in lateral barium enema study film may be suggestive of diagnosis.

Ultrasound examination shows uni- or multi-locular cystic lesion located retrorectal, sometimes with internal echoes caused by mucoid material or present with infection.

Cross sectional imaging (CT and MRI) provide excellent direct anatomic description view⁷ and thus helps not only in making the diagnosis in asymptomatic retrorectal lesions but also in planning correct surgical approach by determining the extent and nature of the lesion. In cystic lesions especially epidermoid and dermoid cysts CT scan shows a well defined oval, thin walled, unilocular formation, may be associated with fine calcifications and CT signal attenuation. If the cyst is infected , the walls may appear thickened with pericystic inflammatory changes.

Endorectal ultrasonography is a very sensitive method in assessing the rectal wall involvement and pelvic floor muscle invasion. MRI for some has become the preferred imaging modality to CT because of its superior characteristic in delineating the structures in this area⁸.

MR angiogram or venogram may add additional information regarding vascular involvement and indicate the need for a vascular surgeon to be a member of multidisciplinary surgical team.

Biopsy : Preoperative tissue biopsy is controversial^{9,10}. If tissue diagnosis of a retrorectal mass is required, a needle biopsy should be performed by an experienced radiologist and within the field of the proposed area of resection so that the needle tract may be excised en-bloc with the specimen at the time of operation, if turns out to be malignant on histopathological report.

However, preoperative tissue diagnosis with image guided biopsy is considered essential for the management of solid and heterogenous cystic tumors. Purely cystic lesion should not be biopsied unless there is a high suspicion of malignancy.

Treatment : Treatment involves complete excision of the cyst and , if present, the sinus². Posterior approach through parasacro-coccygeal midline, curvilinear or horizontal incision with the patient in prone jack knife position is indicated for low lesions or infected cysts¹¹. We in our 3 cases used upward arched incision¹², and it gives not only good exposure but good outcome also as none of our 3 cases developed ischemia of skin flap.

Further these lesions tend to adhere to the coccyx, so in the case of large cyst to gain access, surgical approach requires en-bloc coccygectomy¹³. The coccyx should also be removed en-bloc in any child with a presacral dermoid because of the risk of sacrococcygeal teratoma².

IV. Conclusion

Retrorectal cysts are difficult for treatment as well as for diagnosis, where even puncture biopsy is not recommended, and along with the fact that they may be complicated by recurrence as in one of our case, they should be treated in a specialized institutions by experienced surgeons from the moment of diagnosis to the definitive surgical treatment. Surgical treatment requires en-block coccygectomy, which helps in deeper dissection

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Photo 1: Photograph showing natal cleft sinus



Fig 1: M.R.I scan showing well defined cystic lesion in left perineal region



Photo 2: Intra-operative picture showing sinus tract being lifted from sacral fascia



Photo 3: Coccygectomy in progress



Photo 4: Dissection of retro-rectal cyst being performed from behind forward



Photo 5: Wound closure done by double Z-plasty