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Original Research Paper

General Surgery

RETRORECTAL TUMOR : A RARE ENTITY

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ABSTRACT

INTRODUCTION: Primary neoplasms of the retrorectal (presacral) space are very rare. These lesions may be congenital or acquired, benign or malignant. They often arise with subtle clinical symptoms, or they may be found incidentally during evaluation for other conditions. Preoperative imaging can provide useful information for operative planning, however, it does not replace findings for surgery. Approaches for resection include posterior only and combined abdominoperineal, depending on the characteristics of the lesion.

CASE PRESENTATION: A case of 20 years old female, presented in outpatient clinic with bowel disturbance and lower abdominal pain. Investigations were done including contrast enhanced computed tomography (CT)and magnetic resonance imaging (MRI) of whole abdomen and pelvis which revealed ill defined heterogenously enhancing mass in retrorectal space; followed by which patient was taken for surgery and biopsy taken.

CONCLUSION: Retrorectal tumors remain a difficult diagnostic and therapeutic challenge despite the use of newer imaging modalities and improvements in perioperative care. Complete resection with negative margins is the standard for benign retrorectal tumors.

KEYWORDS : Retrorectal Tumor, Presacral Space.

INTRODUCTION

Lesions within the retrorectal, or presacral, space are uncommon and therefore present a challenge to diagnose and manage. Although retrorectal tumors are estimated to account for approximately 1 in 40,000 hospital admissions, the true incidence in the general population is unknown. The retrorectal space contains embryologic remnants that have pluripotent capability; hence, a heterogeneous histologic group of tumors arise in this region.[1] Lesions may be solid or cystic, the solid variety having a higher predilection for malignancy. Surgical resection is the principal therapy for retrorectal tumors. Even benign cystic lesions are excised because doing so eliminates or prevents symptoms and addresses the risks of subsequent infection or malignant transformation. CT and MRI have been found effective for differentiating benign from malignant entities. CT or MRI can be used for surgical planning because the size and extent of the tumor can be delineated, and the presence of local invasion can be assessed with either modality. Retrorectal lesions can be resected via one of three approaches: anterior (abdominal), posterior (perineal), or combined.

ANATOMY:[2]

Anatomically, the retrorectal space is the pelvic continuation of the retroperitoneum. In adults, this space is filled with loose areolar tissue, fat, and connective tissues. Its boundaries are the rectum anteriorly and the sacrum and investing sacral fascia posteriorly. Although the rectosacral (Waldeyer's) fascia separates the true retrorectal space from the more inferior/caudal horseshoe-shaped supralevator space, lesions in both regions are typically considered together, with the levator and coccygeal musculature constituting the inferior boundary. The ureters, internal iliac arteries, and lateral rectal stalks mark the lateral extent of the retrorectal space, and it extends cranially to the peritoneal reflection.

Various classification schemes have been reported for retrorectal tumors. Most commonly, these tumors are broadly categorized as inflammatory, congenital, neurogenic, osseous, and miscellaneous (Table 1).

Table 1. Classification of retrorectal tumors.[3]

Benign	Malignant
Congenital	
Developmental cysts (Tailgut cyst, rectal duplication cyst, epidermoid, dermoid, and teratoma)	Chordoma Teratocarcinoma
Anterior sacral meningocele	
Adrenal rest tumors	
Neurogenic	
Schwannoma	Ependymoma
Neurofibroma	Malignant nerve sheath tumors
Ganglioneuroma	Neuroblastoma Ganglioneuroblastoma
Osseous	
Giant-cell tumor	Osteogenic sarcoma
Aneurysmal bone cyst	Ewing's sarcoma
Osteoblastoma	Myeloma Chondrosarcoma
Miscellaneous	
Lipoma	Liposarcoma
Myelolipoma	Mucinous cystadenocarcinoma
Myxoma	Fibrosarcoma
Extramedullary hematopoiesis	Malignant fibrous histiocytoma
Fibroma	Hemangiopericytoma
Leiomyoma	Metastatic carcinoma
Desmoid tumor	Lymphoma
Abscess/hematoma	

CASE PRESENTATION:

A 20year old female presented at outpatient clinic with complaint of bowel disturbance and lower abdominal pain since 5 days. Per abdomen examination was normal, digital rectal examination did not reveal any enlarged hemorrhoids or mass.

USG abdomen was done showed 37*34mm sized hypoechoic lesion without significant vascularity in pelvis displacing the rectum right laterally. All routine blood investigations were within normal limits and stool routine and culture examination were also normal.

CECT abdomen and pelvis was done which revealed approx 41*50*80 mm sized lobulated soft tissue density lesion showing homogenous enhancement deep to the natal cleft anterior to coccyx in midline reaching upto left mesorectum

without any evidence of calcification,necrosis or fat density within it. Lesion displaces anus anteriorly; laterally on the right side it compresses and displaces rectum and shows focal obliteration of fat plane with the distal rectum.

Exploratory laparotomy was done with lower midline incision and on entering the peritoneal cavity, hard impacted mass was palpable over the pre-coccygeal region and so small part was resected and sent for frozen section histopathology. Hard mass was adherent to coccyx and hence was difficult to separate.

HPE showed SPINDLE CELL TUMOR; possibility of
(1) LOW GRADE FIBROMYXOID SARCOMA
(2) SPINDLE CELL TUMOR OF NEUROGENIC ORIGIN

Later MRI abdomen was done which showed approx 41*45*32mm sized ill defined hetergenously enhancing altered signal intensity lesion noted involving retrorectal space (left>right). The lesion invades both levator ani/puborectalis muscle; infiltrates in fat of both ischiorectal fossa, displaces rectum anteriorly with loss of fat plane and possible invasion into postero-lateral wall of rectum. The lesion extends inferiorly upto natal cleft.

Patient was then again taken for surgery, and jack-knife position was given. Vertical incision put on gluteal cleft, approx 6*4*3cm sized firm-hard consistency mass visualised and removed from left ischio-rectal space. Another 2*2*3cm sized mass with similar consistency removed from right ischiorectal space (Figure 2 &3) and sigmoidostomy done. Both the tissues were sent for HPE.

Biopsy from the mass revealed LOW GRADE SPINDLE CELL SARCOMA and immunohistochemical staining was advised which confirmed the diagnosis of spindle cell sarcoma but sensitive to VIMENTIN.

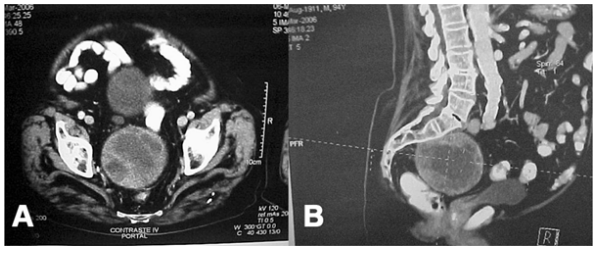

Fig 1. MRI showing heterogenous mass in retrorectal space.

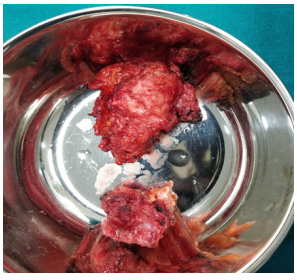

Fig 2. Two masses removed from both ischiorectal fossae.

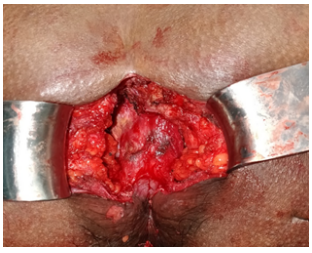

Fig 3. Photograph showing bilateral ishiorectal fossae after removal of mass.

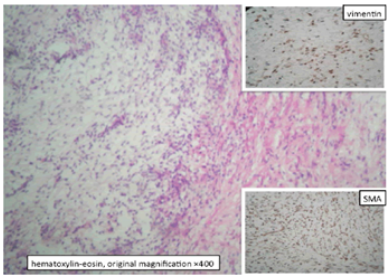

Fig 4. Histopathology and immunohistochemical staining showing fibroblasts and myxoid stroma.

DISCUSSION

The true incidence of tumors occurring in the retrorectal (presacral) space is unknown, yet several retrospective series suggest that between one and six patients will be diagnosed annually in major referral centers [4]. The retrorectal space contains multiple embryologic remnants derived from a variety of tissues. Tumors that develop in this space are both macroscopically and histologically heterogeneous. Most lesions are benign, but malignant neoplasms are not uncommon. Solid lesions are more likely to be malignant than are cystic lesions. Neurogenic lesions typically arise from peripheral nerves and represent about 10% of retrorectal tumors [5]. These tumors include neurofibromas and sarcomas, neurilemomas, ependymomas, and ganglioneuromas. Fibromyxoid sarcoma is a rare soft tissue sarcoma usually located in the deep soft tissue in the groin or lower extremities. No case located in retrorectal space has been reported in literature review[4]. Thus, it is important to differentiate this tumor from other soft tissue tumors [6]. Symptoms of retrorectal tumors are often nonspecific and are related to the location and to the size of the lesion. The majority of benign cystic lesions are asymptomatic and usually discovered on routine rectal examination. Pelvic MRI is emerging as the most sensitive and specific imaging study of these tumors. Non-surgical treatment, such as radiation treatment and chemotherapy, might improve local control making the appearance of clinically evident metastatic disease less likely. Although adjuvant therapy has enhanced the chance of cure for retroperitoneal sarcomas, there are no studies for tumors of retrorectal space. In fact, chemotherapy for retrorectal sarcomas seems to be ineffective. Thus, further studies are necessary to clarify the role of adjuvant treatment for local control of these tumors [6]. Overall survival appears to be good if the resection is complete [7].

CONCLUSION

Surgery followed by adjuvant chemotherapy depending upon histological grade and immuno-histo-chemical staining remains the mainstay of treatment. For larger tumors, aggressive resection may be required to achieve N0 margin.

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