Doc, I've got a mass behind my rectum

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Introduction

- Presacral – retrorectal space is a potential space and the location of wide range of rare tumors
- Incidence as low as 1 in 40,000-60,000 hospital admissions
- Usually asymptomatic and diagnosis is frequently delayed

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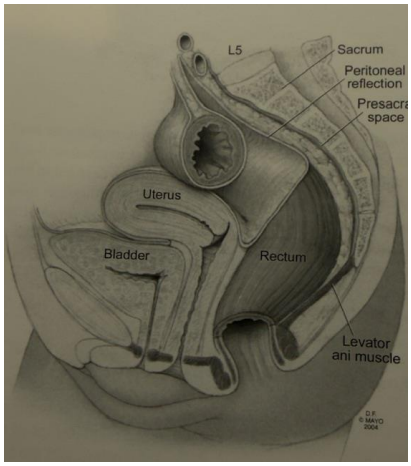
Anatomy


L5
Sacrum
Peritoneal reflection
Presacral space
Uterus
Bladder
Rectum
Levator ani muscle

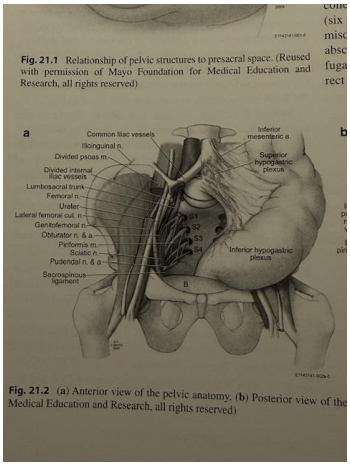


Fig. 21.1 Relationship of pelvic structures to presacral space. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)
a
b
Common iliac vessels
Ilioinguinal n.
Divided psoas m.
Divided internal iliac vessels
Lumbosacral trunk
Femoral n.
Ureter
Lateral femoral cut. n.
Genitofemoral n.
Obturator n. & a.
Piriformis m.
Sciatic n.
Pudendal n. & a.
Sacrospinous ligament
Inferior mesenteric a.
Superior hypogastric plexus
Inferior hypogastric plexus
S1
S2
S3
S4
Fig. 21.2 (a) Anterior view of the pelvic anatomy. (b) Posterior view of the
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Clinical Presentation

- Most discovered incidentally during routine pelvic/rectal exam
- Pain is the most common symptom- vague pain in pelvis, perineum and or low back
- Pain worse with sitting and improved by standing or walking
- Pain is more associated with malignant lesions
- Constipation, urinary symptom, fecal incontinence, and sexual dysfunction are associated with sacral nerve involvement with advanced tumors
- Some patients present with perineal discharge – misdiagnosed as anal fistula

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Physical Examination

- DRE – presence of an extra-rectal mass displacing the rectum anteriorly
  - Determine fixation to the surrounding structures
  - Extent of the mass
- Check for post anal dimple
- Proctoscopy or flexible sigmoidoscopy
  - Evaluate the upper and lower extent of the tumor
  - Common finding is normal mucosa with extrarectal mass effect
  - Inflamed mucosa suggestive of infection

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Imaging Studies

- MRI of the pelvis is the gold standard imaging modality
- Scimitar sign – Pelvic Xray – Meningocele
- Transrectal ultrasound
- CT scan

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Preoperative Biopsy

- Controversial
- Advances in imaging has increased ability to accurately diagnosis
- Biopsy if it changes pre or post operative management
- If biopsy is necessary, it should be performed within the field of proposed area of resection
- Avoid transrectal, transvaginal or transperitoneal approach
- Biopsy of cystic lesions increases risk of secondary infection and recurrence after resection
- Biopsy of meningocele could lead to meningitis and death

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Classification

- Congenital
- Acquired
- Benign
- Malignant

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Epidermoid and Dermoid Cysts

- More common in females
- Well circumscribed, defects during closure of the ectodermal layer
- Typically, benign
- Occasionally communicate with skin creating a postanal dimple
- Misdiagnosed as perirectal abscess
- Dermoid cyst may contain sweat glands, hair follicle, sebaceous cyst but epidermoid cysts do not

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Tailgut Cysts

- More common in females and arise from post anal primitive hindgut
  - Cystic hamartomas/mucus secreting cysts
- Can resemble the adult or fetal intestinal tract
- Malignant transformation has been reported

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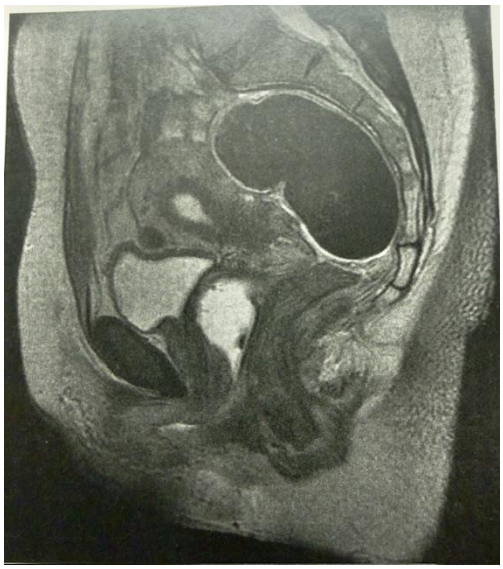

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Enterogenous Cyst

- Rectal duplication cysts are more common in women
- Communicates with the rectum – derive from developing hindgut
- Multilobular with satellite cysts
- Mucus membrane, smooth muscular coat and attached to the alimentary tract
- Generally benign but could lead to malignant transormation

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Teratomas

- Sacrococcygeal teratomas are neoplasms that contain all 3 germ layers – totipotential cells, contain both solid and cystic components
- More common in pediatric age group and females
- May contain tissues from almost any organ system – including digestive, nervous respiratory and skeletal system – contain hair, bone and teeth
- Cystic components are typically benign and whereas solid components are more associated with malignant degeneration
- Reach considerable size and in infants only 7% of girls and 10% of boys present with malignancy
- Malignant degeneration can occur in adults

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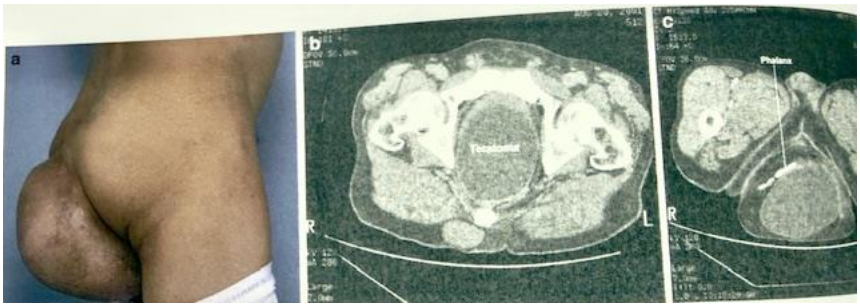

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Chrodomas

- Sacrococcygeal chordomas are the most common malignant tumor of the presacral space
- More common in male
- Arise from vestigial notochord tissue, and can occur along the spinal tract but most commonly noted in the pheno-occipital region and sacrococcyeal region
- Symptoms are often vague – pain with sitting and alleviated by standing

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Chordomas

- Due to vague symptoms diagnosis is often delayed and tumors can reach considerable size
- Resulting in constipation, fecal, urinary incontinence and sexual dysfunction
- Local and distant recurrence rate – 43 % and 22% with 5 year survival rate of 67% and 40%

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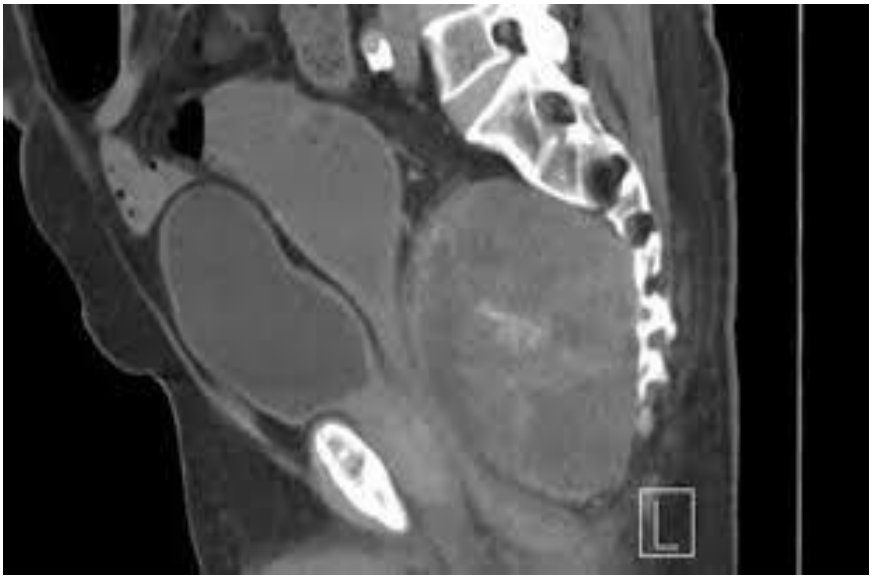

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Meningoceles

- Anterior sacral meningocele arise from protrusion of the thecal sac through a defect in the sacrum contain cerebrospinal fluid
- Scimitar sign- sickle shaped sacrum – unilateral well-marginated, cresecent shaped defect in the lateral sacrum
- Head ache related to postural changes and Valsalva maneuver, low back, pelvic pain, constipation, urinary dysfunction, incontinence
- Associated with other congenital abnormalities
- Should not be biopsied
- Surgical treatment consists of obliterating the communications between the subarachanoid space and herniated sac, detethering spinal chord and resection

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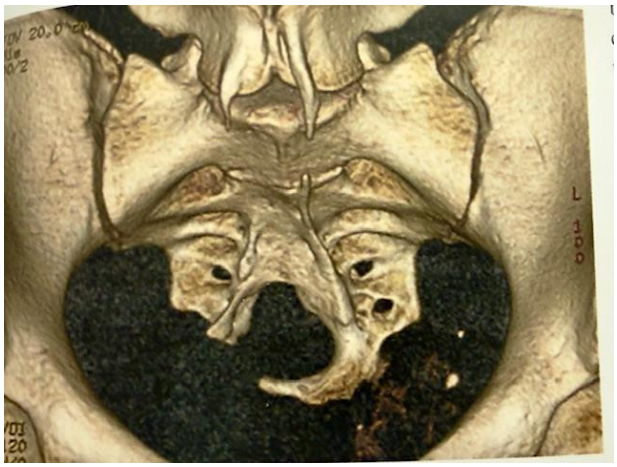

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Neurogenic Tumors

- Arise from the peripheral pelvic nerve plexus
- Make up 10-15% of all presacral tumors
- Most >90% are benign but differentiating benign from malignant tumor needs tissue biopsy
- Schwannomas and ependymomas are 2 most commonly seen tumors
- Symptoms – neuropathy, low back and pelvic pain
- Benign and malignant tumors have a high local recurrence rate and survival for malignant tumors is poor
- Early detection and aggressive surgical intervention

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Osseous Tumors

- Presacral osseous tumors make up less than 10% and arise from bone, cartilage, fibrous tissue and marrow
- More common in males
- Half are malignant at the time of diagnosis
- Can reach considerable size and cause local destruction
- Pronounced metastatic potential – pulmonary being the most common

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Currarino Syndrome

- Rare congenital malformation associated with 3 main features
  - Sacral malformation – agenesis or sickle shaped
  - Hind gut anomaly
  - Presacral tumors
- Autosomal dominant – mutations in HLXB9 gene
- Most common presacral tumor is dermoid cyst or teratoma
- Malignancy is rare

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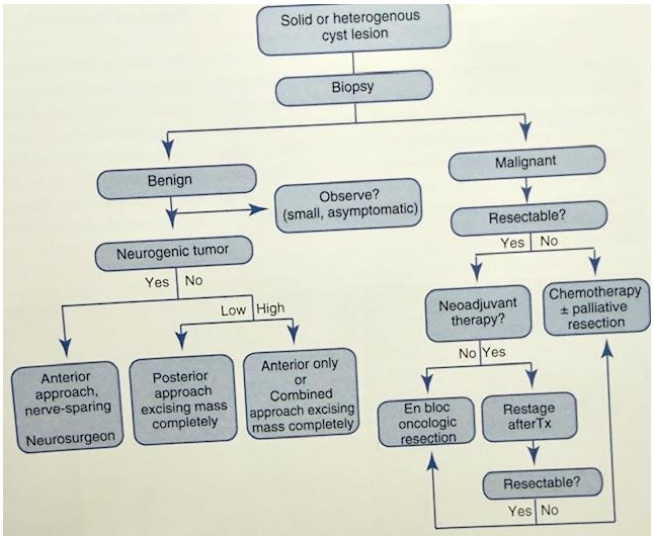
Solid or heterogenous cyst lesion
Biopsy
Benign
Observe? (small, asymptomatic)
Neurogenic tumor
Yes
No
Low
High
Anterior approach, nerve-sparing
Neurosurgeon
Posterior approach excising mass completely
Anterior only or Combined approach excising mass completely
Malignant
Resectable?
Yes
No
Neoadjuvant therapy?
Chemotherapy ± palliative resection
No
Yes
En bloc oncologic resection
Restage afterTx
Resectable?
Yes
No

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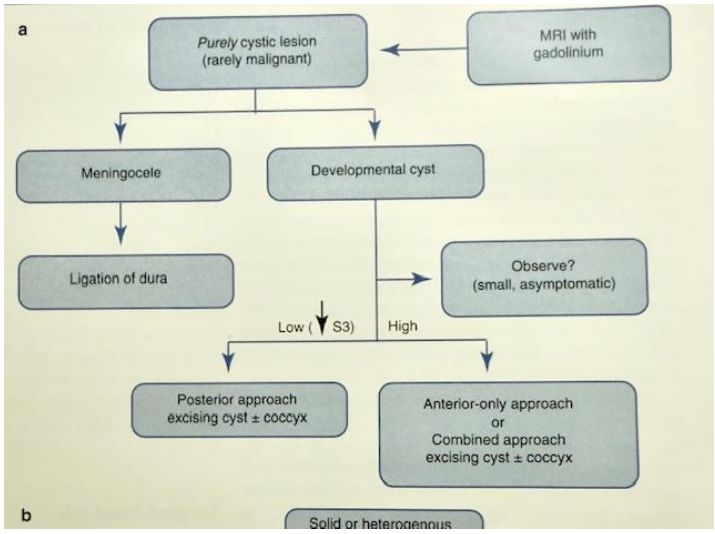
a
Purely cystic lesion (rarely malignant)
MRI with gadolinium
Meningocele
Developmental cyst
Ligation of dura
Observe? (small, asymptomatic)
Low (
S3)
High
Posterior approach excising cyst ± coccyx
Anterior-only approach or Combined approach excising cyst ± coccyx
b
Solid or heterogenous

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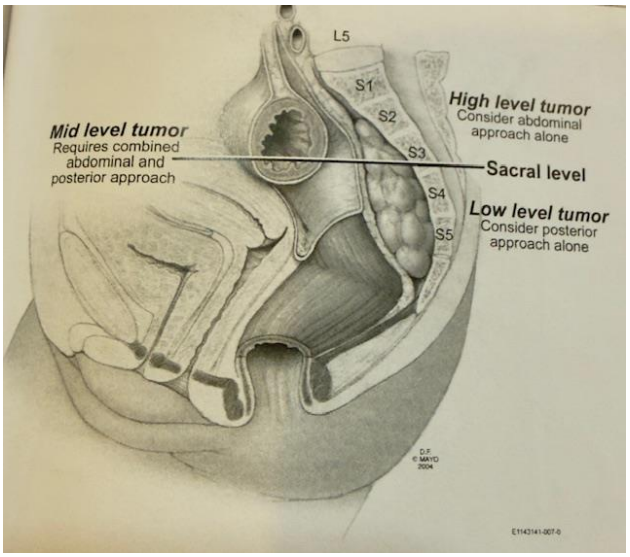

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Multidisciplinary Team

- Colorectal
- Orthopedic
- Spine/neurosurgeon
- Urology
- Vascular surgery
- Plastic surgery
- Medical oncology, musculoskeletal radiology, and anesthesiology

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Management of Presacral Tumors

- Historically has been surgical resection
- Indications for surgery – known malignancy, future malignant transformation, alleviation of symptoms, consistent increase in size
- Small lesions – excision by colorectal surgeon
- Large lesions need multidisciplinary team

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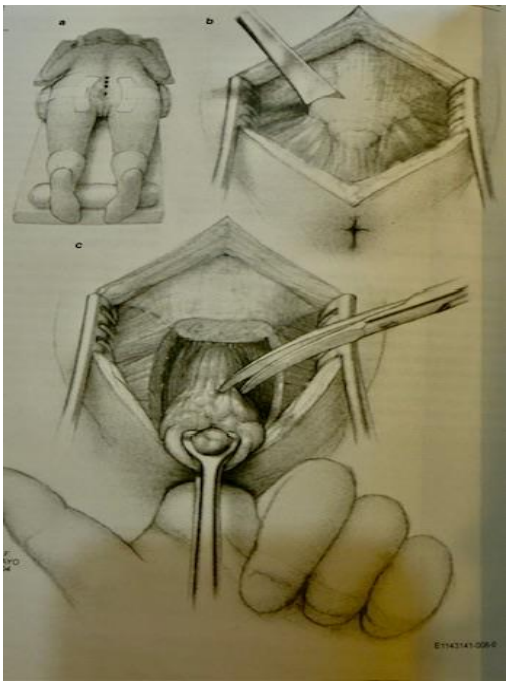

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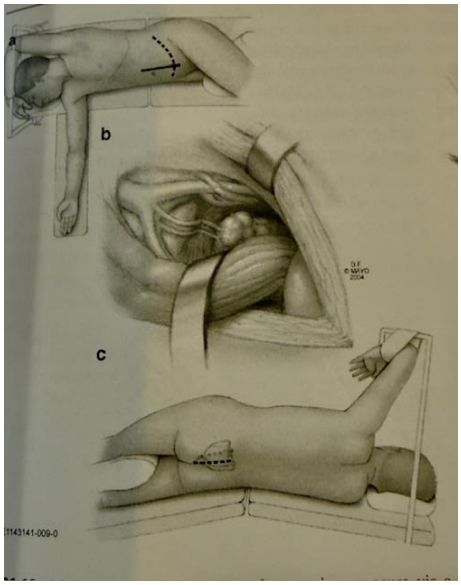
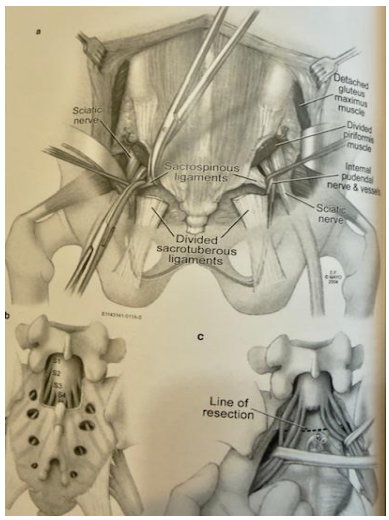

a
b
c
31143141-009-0

31


a
Sciatic nerve
Detached gluteus maximus muscle
Divided piriformis muscle
Sacrospinous ligaments
Internal pudendal nerve & vessels
Sciatic nerve
Divided sacrotuberous ligaments
b
c
S1
S2
S3
S4
Line of resection

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Neoadjuvant Therapy

- Addition of neoadjuvant therapy can decrease tumor size, increase resect ability and decrease rate of recurrence
- Ewings and osteogenic sarcomas respond well to Neoadjuvant therapy
- Chondrosarcomas and chordoma do no respond well
- Squamous cell or adenocarcinoma in malignant cysts respond well

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- Bilateral S2-S5 nerve root removal leads to complete loss of bowel and bladder function
- Bilateral S3-S5 nerve root removal – 40% had normal bowel function and 25% had normal bladder function
- Bilateral S4-S5 nerve root removal – 100% normal bowel function and 69% normal bladder function
- Unilateral S1-S5 nerve root removal – 87% had normal bowel function and 89% normal bladder function
- Resection of S1 or sciatic nerve could lead to drop foot
- When high sacrectomy is performed and with removal of half of the S1 vertebral body pelvic stability can be compromised
- Preoperative radiotherapy increases risk of stress fractures
- May need sacropelvic fixation with metal fixation, bone graft or 3-D printed prosthesis

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- Posterior approach
- Combined approach
- Anterior approach – open or robotic

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Outcomes

- Recurrence rate for benign tumors range from 0 -35%
- Recurrence rate for malignant tumors range from 0 – 48%

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Conclusion

- Rare group of both benign and malignant tumors
- Most benign tumors have malignant potential and if non operative treatment is chosen careful follow up is necessary
- MRI is best imaging study
- Biopsy of the lesion should be performed selectively in lesions that are solid or heterogenous cystic lesion
- Function sparing surgery

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