RETRORECTAL TUMOR

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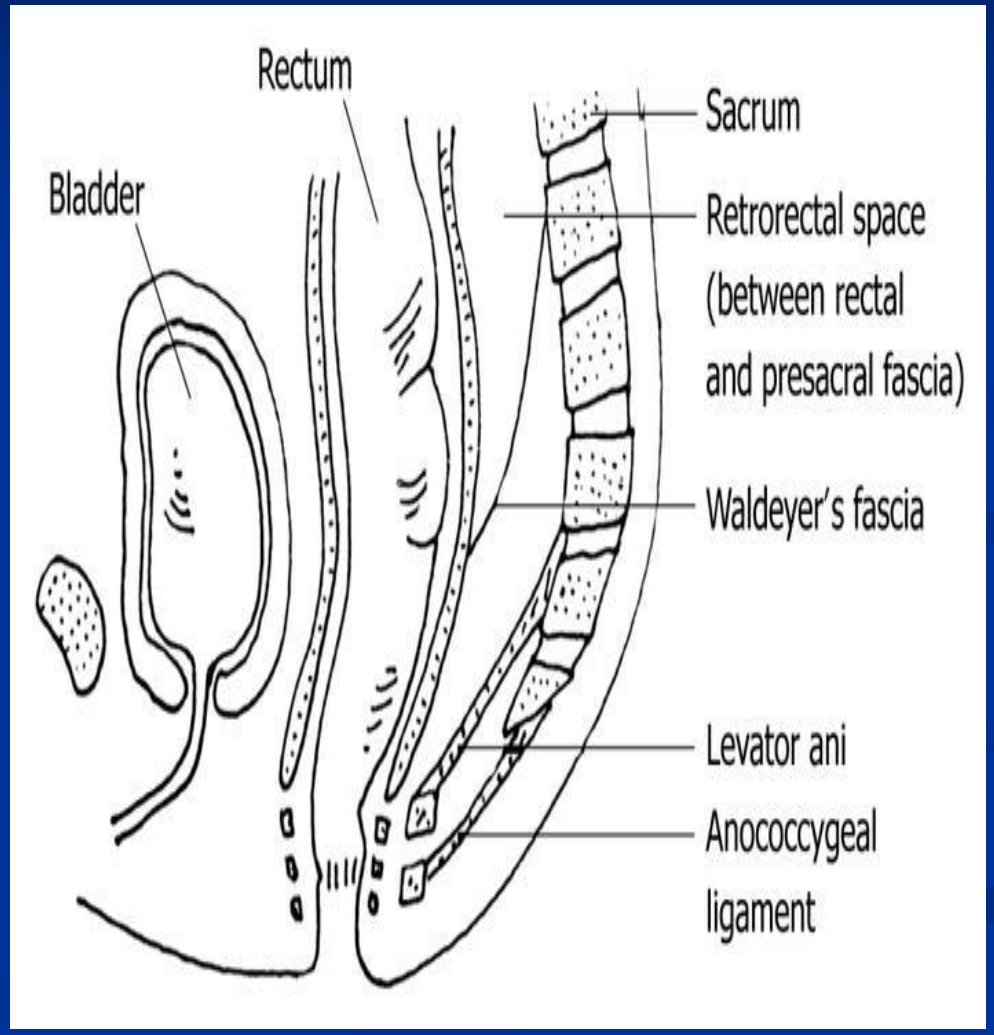
The retrorectal space

- Lies between the upper two-thirds of the rectum and the sacrum, above the rectosacral fascia.

- It is limited--
  - anteriorly ---fascia propria ,
  - posteriorly ---presacral fascia,
  - laterally --- lateral ligaments ureters, and the iliac vessels.
  - Superiorly---by peritoneal reflection of the rectum and communicates with the retroperitoneal space.
  - Inferiorly ----rectosacral fascia, from S4 to the rectum 3–5cm proximal to the anorectal junction.



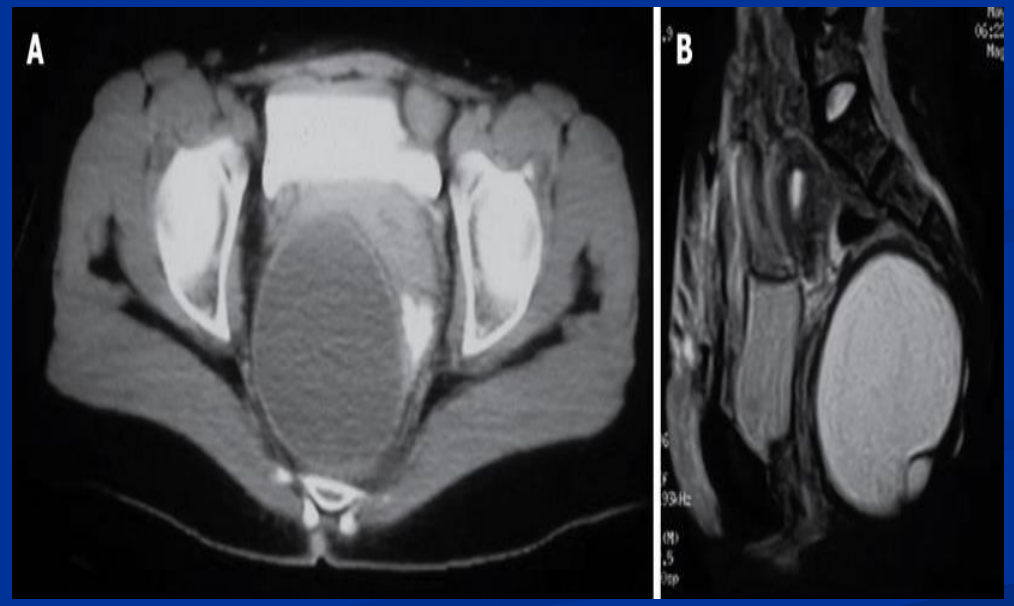
Clinical importance:

- Site for rare presacral tumors.

Retrorectal tumor

INCIDENCE---

- rare.
- congenital or acquired, benign or malignant.
- 2/3rd are congenital, 2/3rd are benign and 1/3rd neoplastic.
- Most are teratomas or chordomas,
- Sacrococcygeal teratomas---most common retrorectal tumors in the pediatric group.



PATHOLOGY---

- Congenital lesions----->more ½ of all presacral tumors, and 2/3rd of these are developmental cysts.

TERATOMA AND TERATOCARCINOMA

Congenital but may present later.

Origin:

- From totipotential cells.
- Classically, they have representative tissue from each germ cell layer.
- Malignancy tends to arise from one of the germ cells.

Malignant potential:

- approximately 30%.
- The more mature the tissue, the more benign the neoplasm ,
- All neoplasms should be viewed as potentially malignant.

Time of Malignant potential:

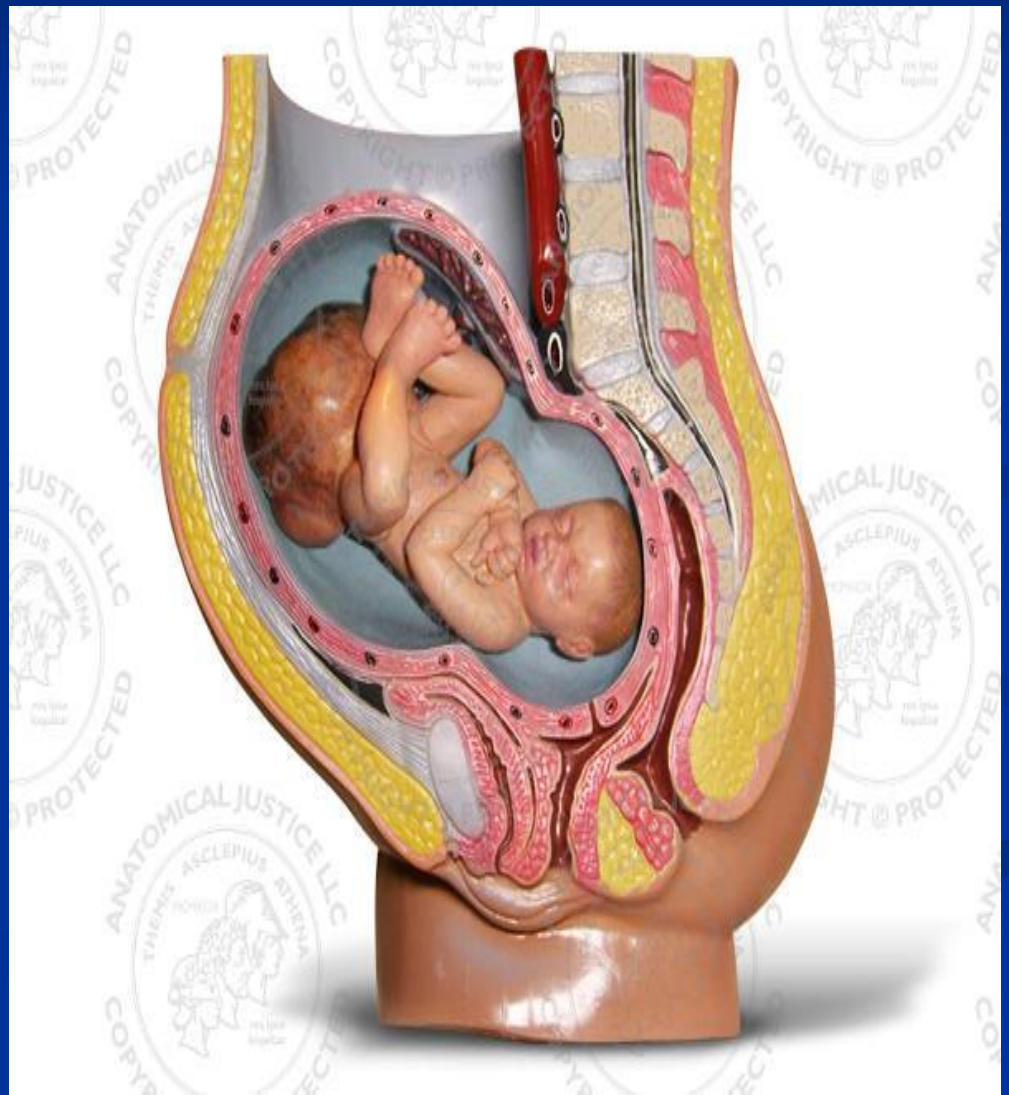
- Tumors of infancy and childhood.
- >20 year, malignancy < common.
- greatest during the period of growth, in early years of life. But still present in adults.

Sex variation:

- Mature teratoma --typically benign & more common in woman,
- Immature teratoma --typically malignant & more in male.

Association: Often associated with anomalies of the-----

- vertebrae,
- urinary tract, or
- anorectum .



Characteristics:

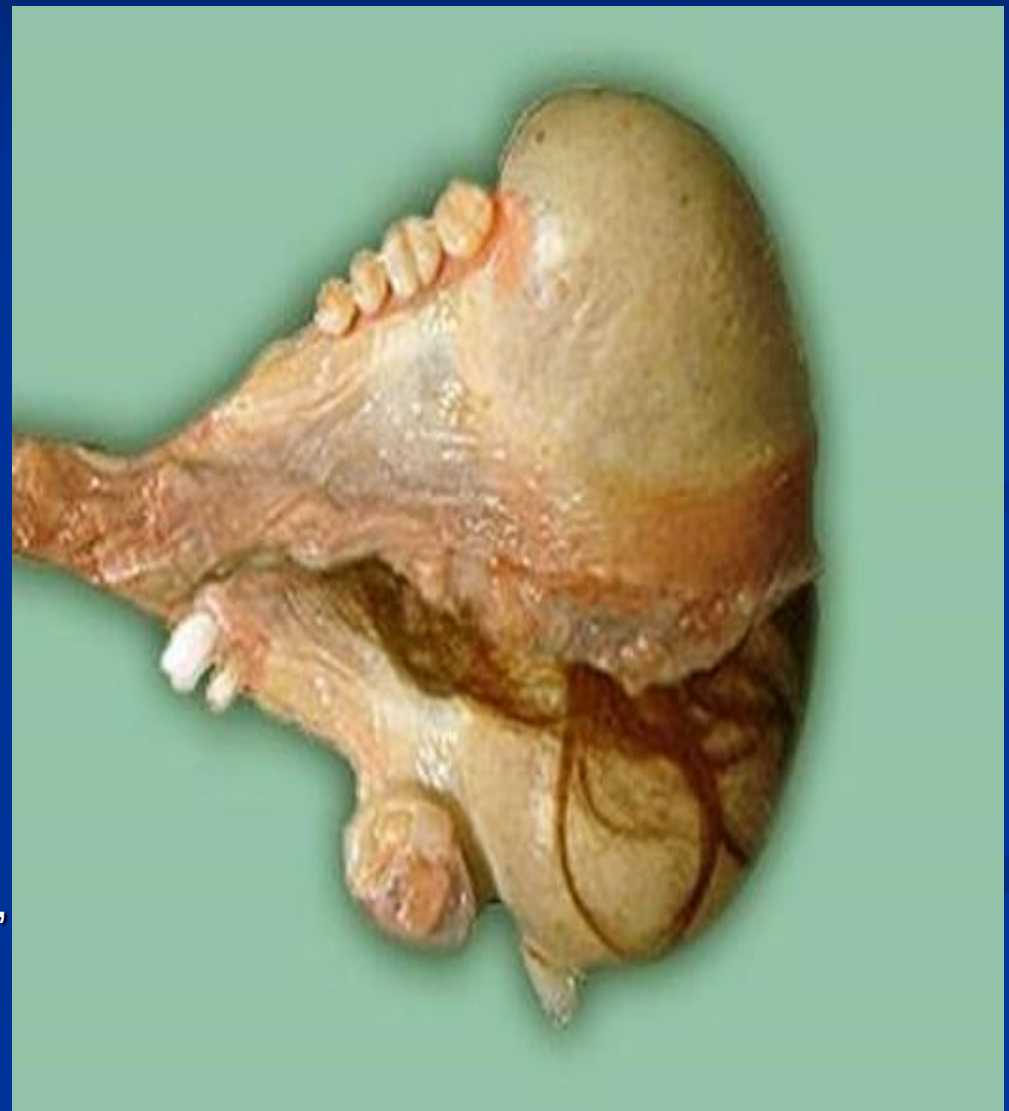
- well encapsulated
- Solid or cystic.
- Strong attachment to the coccyx and sometimes the sacrum, but rarely to pelvic viscera unless previous inflammation.

Composition:

- may contain all kinds of tissues.

Blood supply:

- mainly from the mid-sacral vessels,
- hypogastric vessels may contribute.



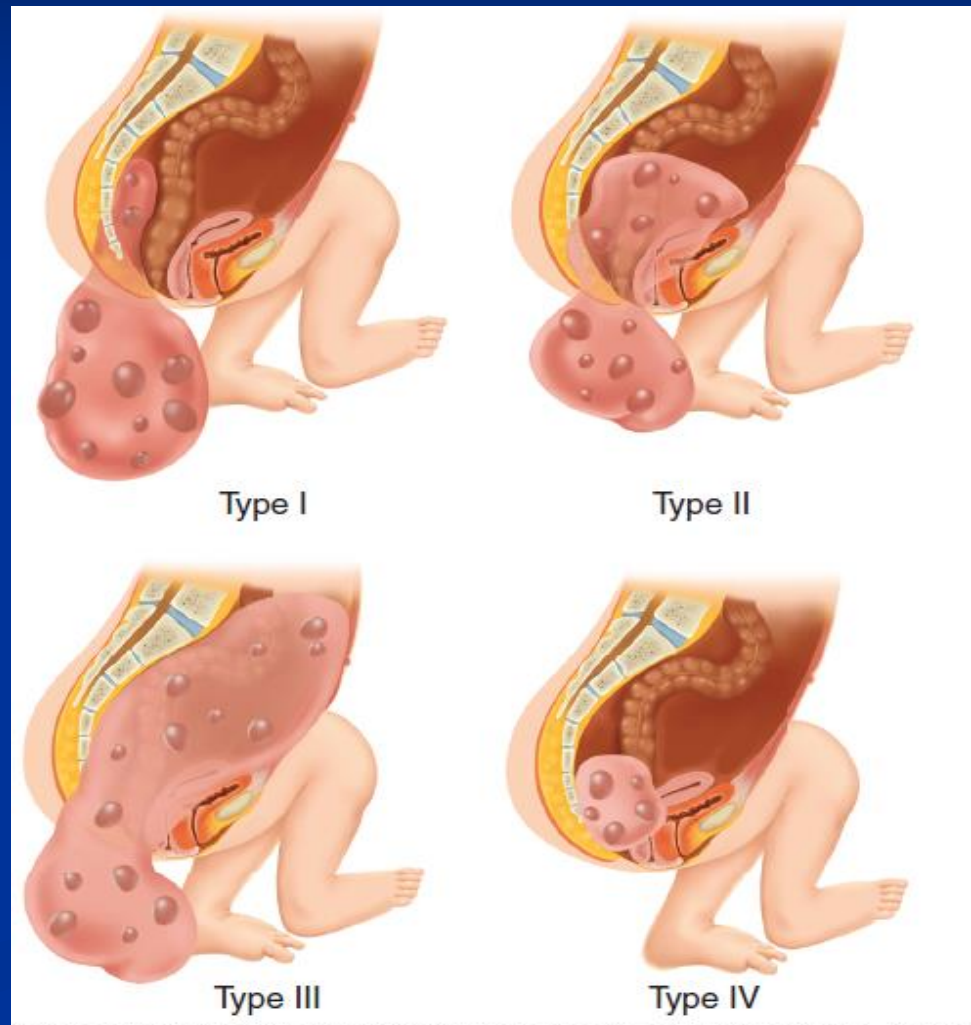
Site:

- may remain confined to the pelvis
- may extend upward into the retrorectal space or downward, distending the perineum and displacing anus and the external genitalia.
- may extend into the spinal canal or through the sacrosciatic notch to emerge in the buttocks region.

Incidence: 1 in 35000 live birth.

Classified according to relative extent-

- Altman type -1-entirely outside.
- Altman type-2-mostly outside.
- Altman type-3- mostly inside.
- Altman type-4-entirely inside; also known as presacral teratoma or retrorectal teratoma.



Source: Diana W. Bianchi, Timothy M. Crombleholme, Mary E. D'Alton, Fergal D. Malone: *Fetology: Diagnosis and Management of the Fetal Patient*, 2nd Edition:
www.obgyn.mhmedical.com

Presentation:

- skin discoloration or a dimple. Occasionally, the dominant mass is toward the peritoneal cavity.
  - pelvic obstruction during labor.
  - ulceration,
  - infection,
  - bleeding, or
  - urinary retention.
  - associated with infection, and may be treated for persistent anorectal fistulas
  - may rupture into the rectum.

Size:

  - Vary
  - from a small retrorectal mass to one that weighs more than the infant.

Prognosis:

- The 5-year survival for children with malignant germ cell tumors is approximately 80% after complete resection.
- Germinomas tend to be radiosensitive, but
- overall, only approximately 50% of patients can be expected to survive 2 years.

OPERATIVE APPROACHES

- Once diagnosed, resection, even in asymptomatic patients, is recommended---
  - lesion may be malignant.
  - With time, a teratoma has a greater chance to become malignant.
  - Cystic lesions may become infected. Once infection occurs, the postoperative recurrence rate is 30%, and repeated operations in the infected area can cause FI.
  - Mortality with untreated anterior sacral meningocele is 30%, chiefly because of infection and meningitis.
  - Young female patients may have dystocia, which increases the risk to mother and fetus.
- A multidisciplinary an orthopedic surgeon or a neurosurgeon might be included.
- Complete mechanical and antibiotic bowel preparation

ABDOMINAL APPROACH

- High retrorectal tumors where safe access is not possible from below.
- for extraspinal neurogenic neoplasms

CHORDOMA

Most common retrorectal malignancy.

Origin:

- From remnants of the fetal notochord (primitive flexible vertebral column).
- Not from nucleus pulposus but from the vertebral bodies.

Site:

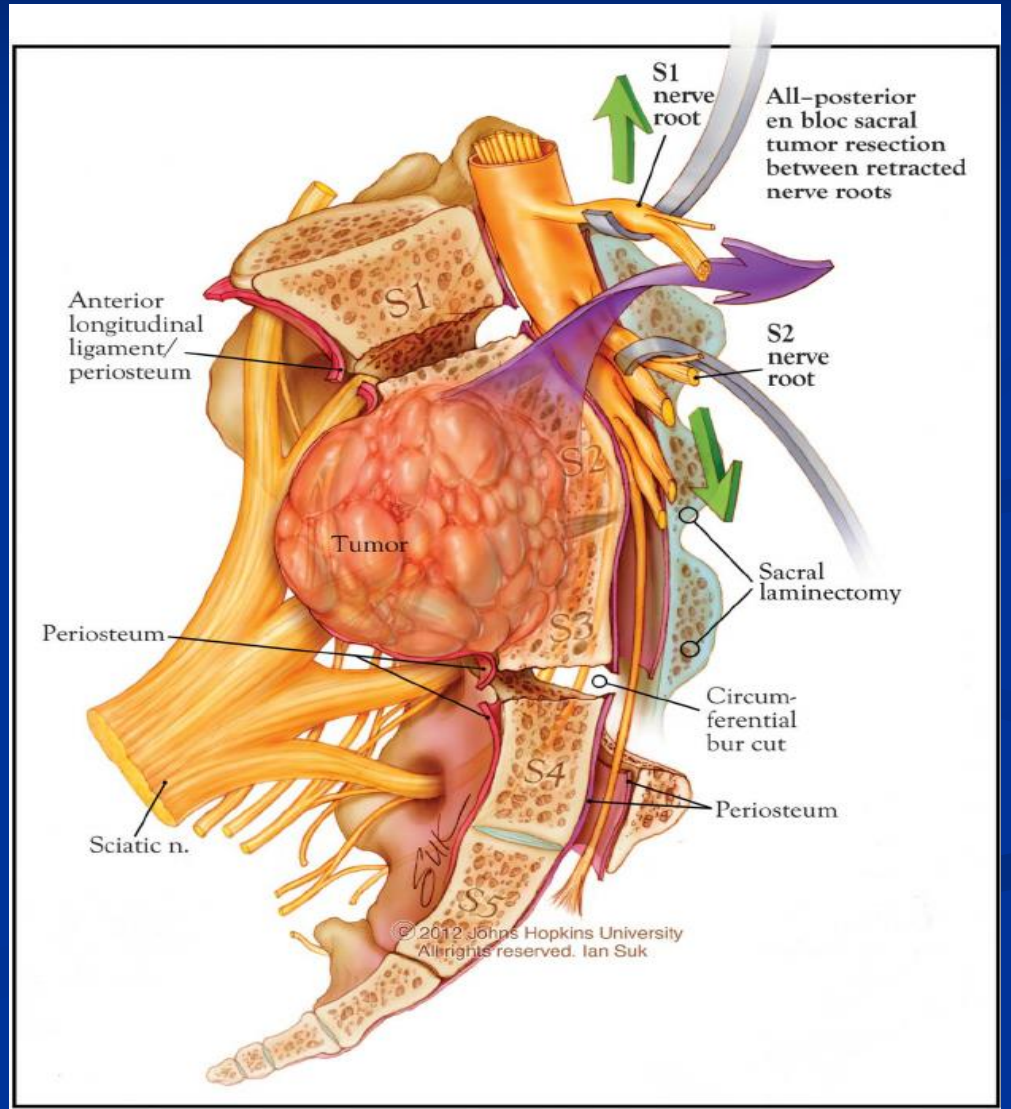
- Anywhere from the hypophysis cerebri to the coccyx,
- Sacrococcygeal area--in approx. 50% of cases.

Sex variation:

- More frequent in men (2:1 to 5:1),

Age:

- 40---70 years. rarely <30 years of age.



Incidence: 3–4% of primary bone tumor.

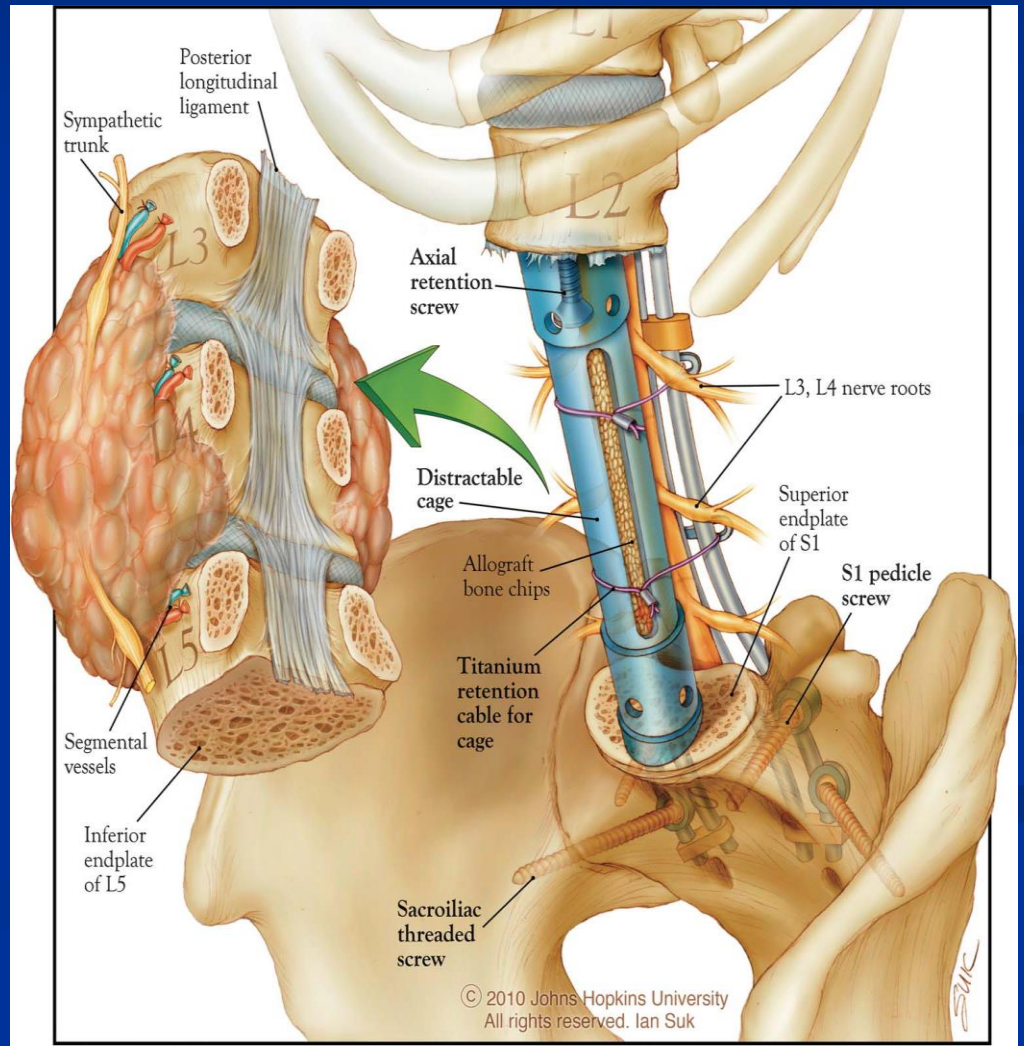
Presentation:

- pelvic, buttock, and LBP ,
- rectal or perineal pain often aggravated by sitting and alleviated by standing or walking.
- constipation, FI, UI, or impotence.

Diagnosis ---often delayed.

Morphology:

- slow growing, lobulated, well-defined structure composed of soft gelatinous tissue, often with areas of hemorrhage.
- invades, distends, and destroys neighboring bone and extends into adjacent regions.



Physical examination—

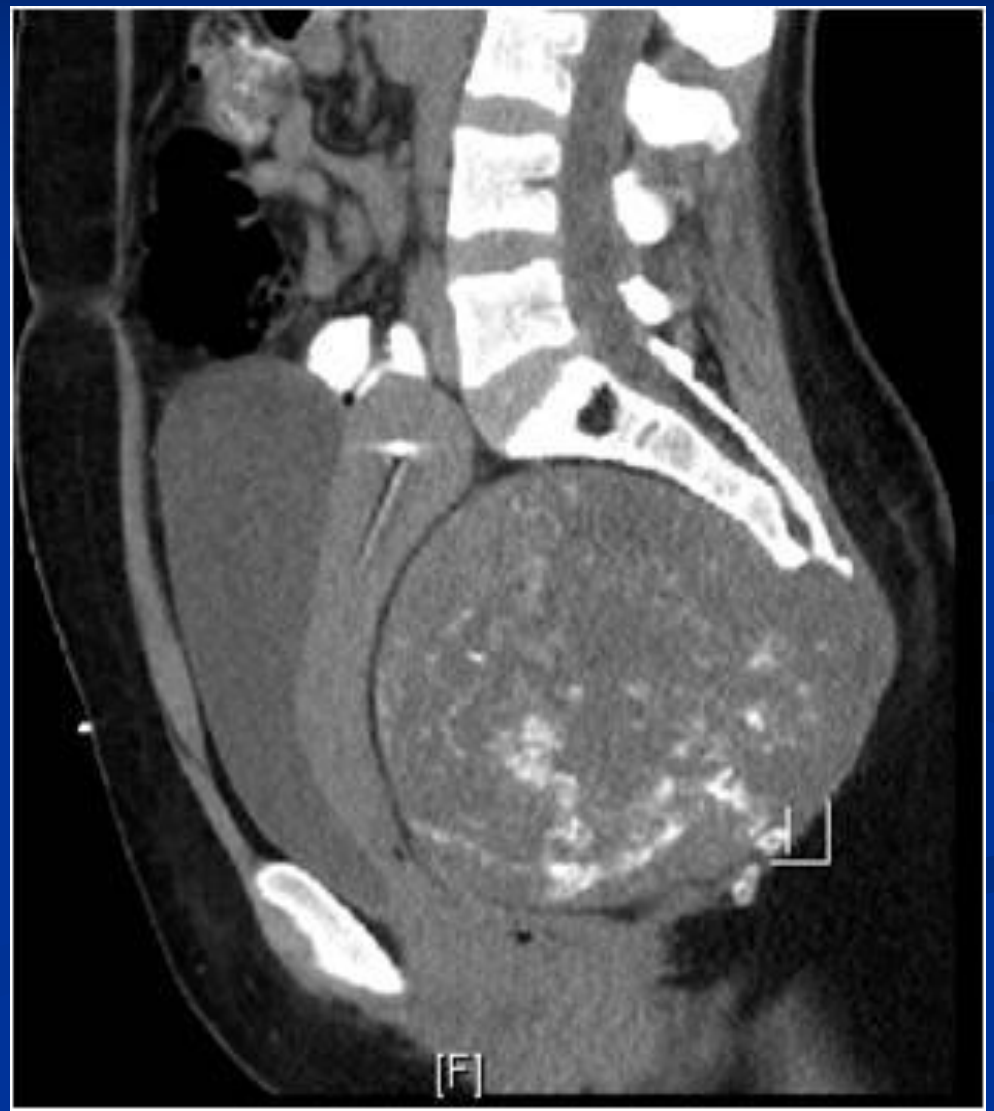
- smooth extrarectal mass with intact overlying mucosa.

Investigation:

Diagnosis usually made with plain x-ray using 4 criteria:

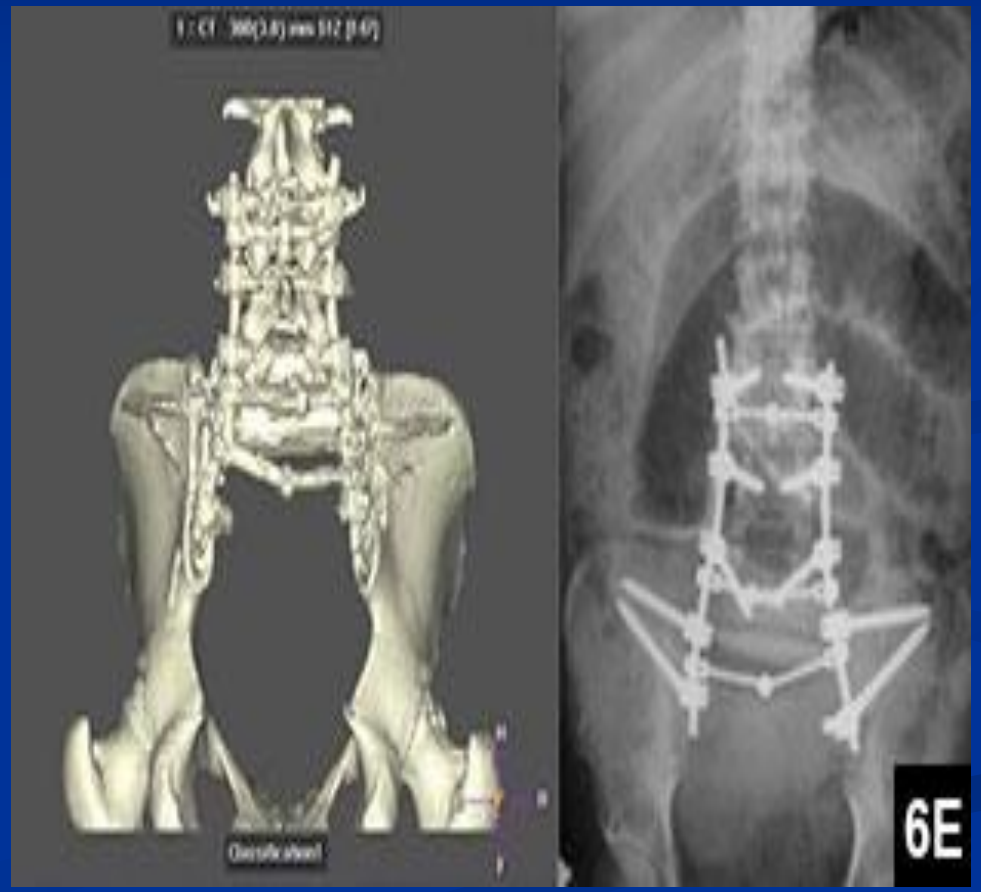
- expansion of bone,
- rarefaction or destruction,
- trabeculation, and
- calcification (occasional) .

- CTscan usually confirms of those not seen on plain films



Treatment:

- En bloc resection of portions of the sacrum, neural sacrifice, and even loss of the rectum.
- Preservation of the upper ½ of the S1 is necessary for stability of the spine and pelvis.
    - Below S3---posterior approach,
    - extending above S3---- anteroposterior approach.
    - If both S3 roots must be sacrificed, the rectum is transected and an end colostomy is fashioned.
    - If 1 S3 root preserved, the rectum is left in place.
    - No patients with bilateral preservation of S3 experienced urinary or bowel symptoms.
- complete excision including preoperative biopsy tracks.
- High dose RT--palliative for incompletely removed lesions.



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