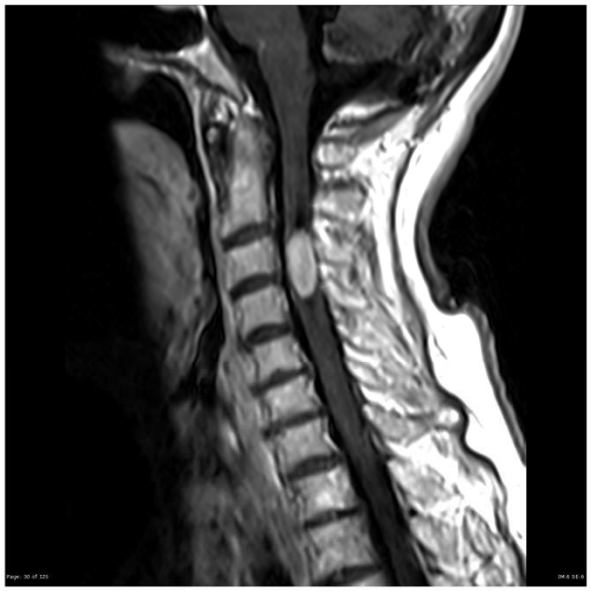
SHORT OVERVIEW OF SPINAL CORD TUMORS

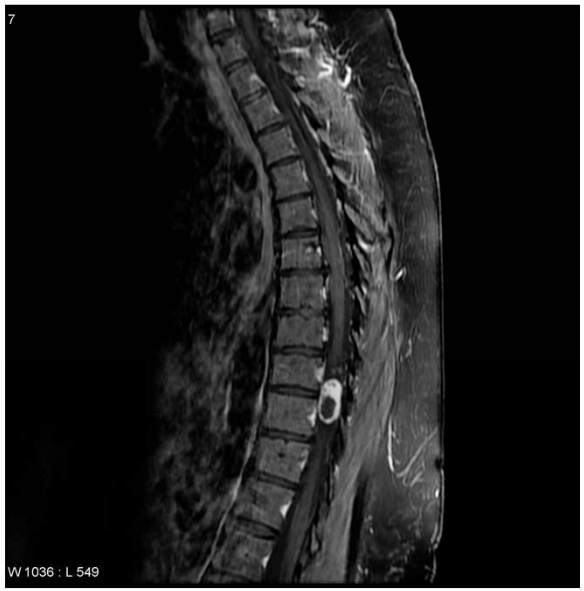
INTRODUCTION

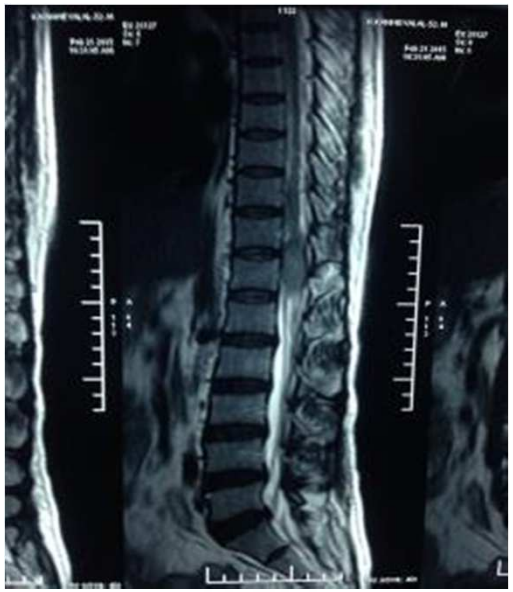
- RARE HETEROGENEOUS GROUP OF TUMORS.
- 15% OF ALL PRIMARY CNS NEOPLASMS ARISE IN THE SC.
- INCIDENCE HIGHER IN MALES THAN FEMALES
- AGE – 10 TO 40 YRS
- MOST PRIMARIES ARE INTRA-DUCTAL
- EXTRAMEDULLARY TUMORS COMPRISES AROUND 2/3 & INTRAMEDULLARY ~ 10%.
- CERVICAL SPINE INVOLVEMENT IS SEEN IN 15-20%, THORACIC IN 50-55%, LUMBAR IN 25-30% CASES.

INCIDENCE

CERVICAL :- 15-20% **THORACIC:-50-55%** **LUMBAR:-25-30%**

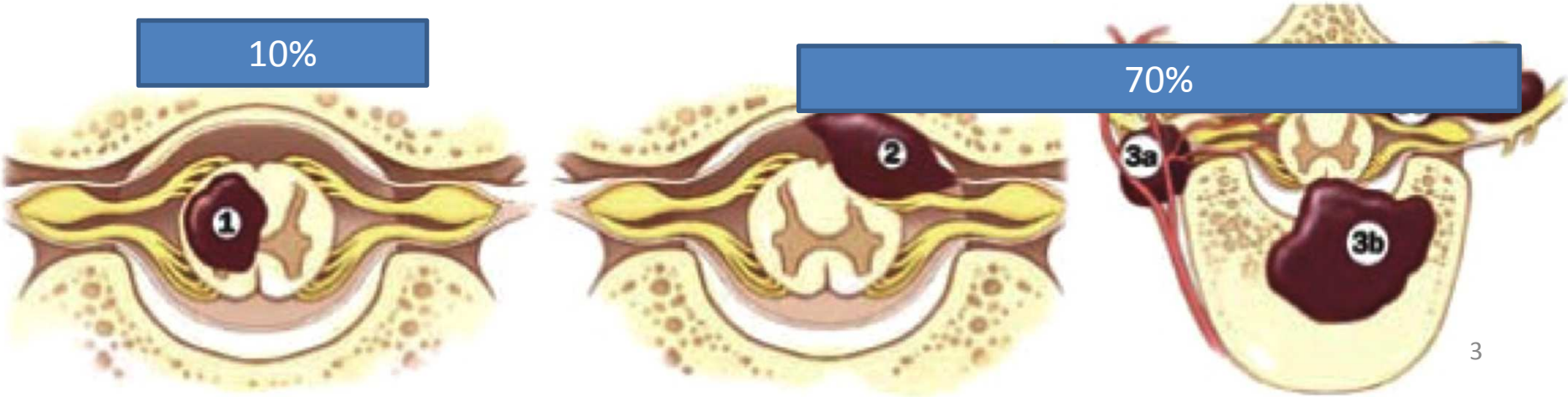






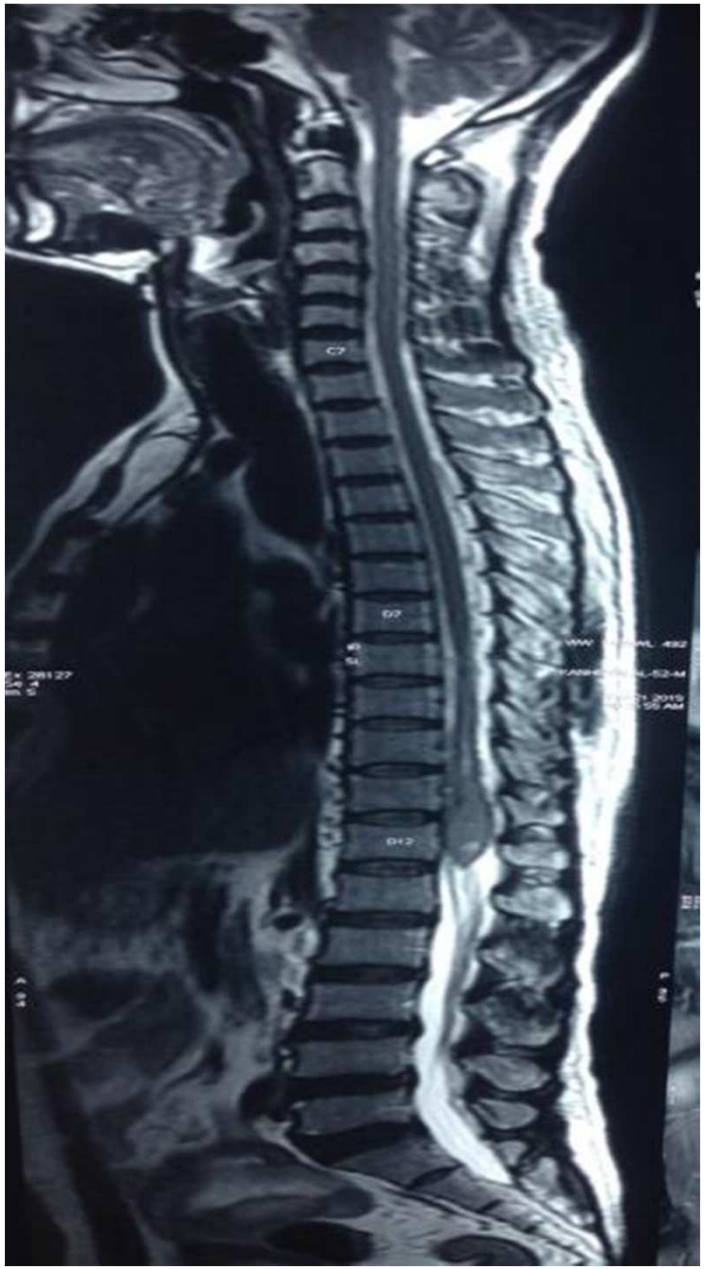
10%

70%



ANATOMY

- SPINAL CORD → EXTENDS FROM MEDULLA OBLONGATA TO L1 / L2 IN ADULTS, NEW BORN L3/L4.
- SIZE 25CM
- WHITE MATTER IN PERIPHERY AND GRAY MATTER IN THE CENTRE (OPPOSITE TO BRAIN)
- 3 LAYERS OF MENINGES
- TRANSMITS SENSORY & MOTOR NERVES.
- IN CERVICAL REGION THE SPINAL NERVES EXIT ABOVE THE CORRESPONDING VERTEBRAE & THORACIC REGION DOWNWARDS EXIT BELOW THE EQUIVALENT NAMED VERTEBRAE.
- BLOOD SUPPLY– 1 ANTERIOR SPINAL & 2 POSTERIOR SPINAL. ARTERY



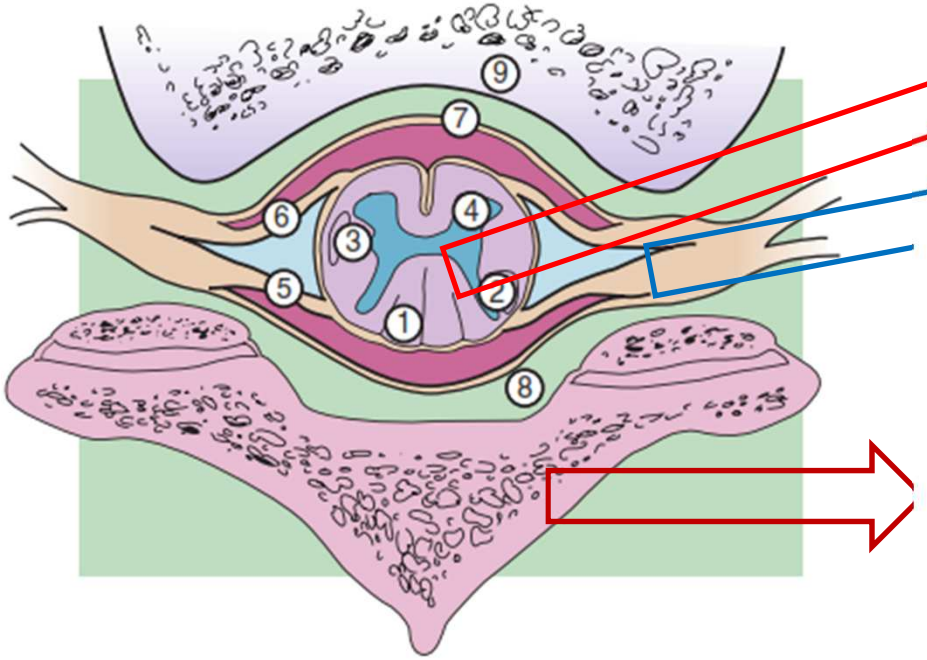
BIOLOGIC CHARACTERISTICS & MOLECULLAR BIOLOGY

1. DIVERSITY OF SPINAL AXIS TUMORS DUE TO LARGE SPECTRUM OF PHENOTYPICALLY DISTINCT CELLS IN THE AXIS.
2. MOST ARE BENIGN.
3. SIGNIFICANT MORBIDITY DUE TO DIRECT COMPRESSION OF IMPORTANT NEURAL STRUCTURES.
4. HISTOLOGY IS AN IMPORTANT PROGNOSTIC FACTOR

    EPENDYMOMA BETTER PROGNOSIS THAN > ASTROCYTOMA

    LOW GRADE & PILOCYTIC ASTROCYTOMA BETTER PROGNOSIS.

SPINAL TUMORS
ANATOMIC DISTRIBUTION ACCORDING TO HISTOLOGY (CBTRUS2010)



Intradural Intramedullary Tumors

- Astrocytoma
- Ependymoma
- Ganglioglioma
- Hemangioblastoma
- Hemangioma
- Lipoma
- Medulloblastoma
- Neuroblastoma
- Neurofibroma
- Oligodendroglioma
- Teratoma (mature)

Intradural Extramedullary Tumors

- Ependymoma, myxopapillary type
- Epidermoid
- Lipoma
- Meningioma
- Neurofibroma
- Paraganglioma
- Schwannoma

Extradural Tumors

- Chondroblastoma
- Chordoma
- Chondrosarcoma
- Hemangioma
- Lipoma
- Lymphoma
- Meningioma
- Metastasis
- Neuroblastoma
- Neurofibroma
- Osteoblastoma
- Osteochondroma
- Osteosarcoma
- Sarcoma
- Vertebral hemangioma

PATHOLOGY

MOSTLY BENIGN COMMON TUMORS ARE

INTRAMEDULLARY

- Astrocytoma
- Ependymoma
- Hemangioblastoma (~ 25% VON-HIPPLE LINDAU Syndrome)
- Mostly seen in children/young adults (<30 yrs)

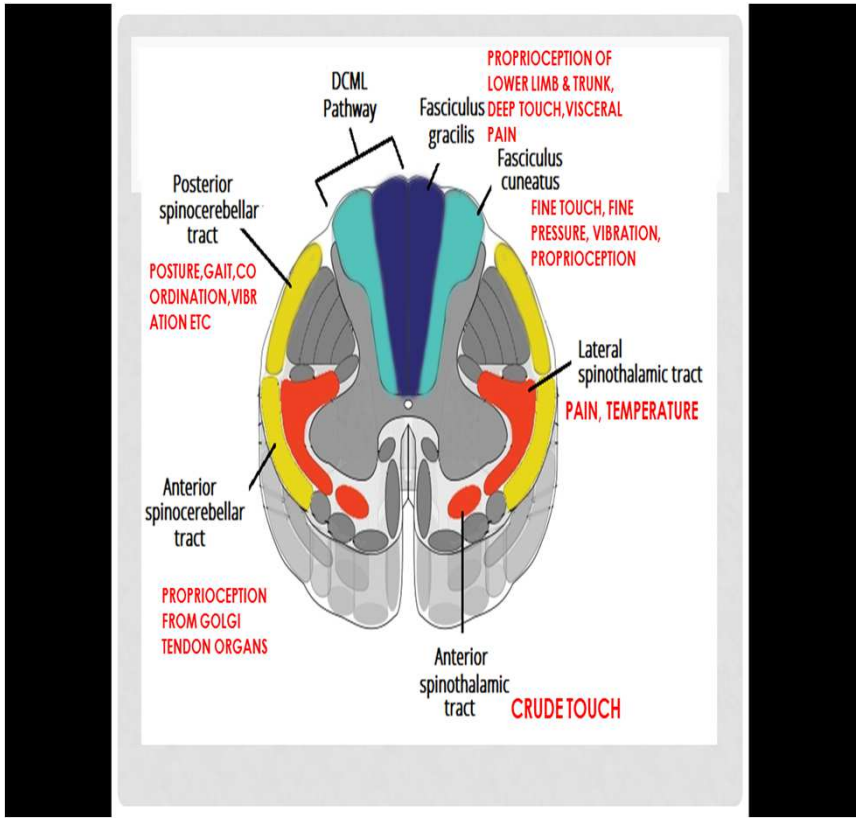
INTRADURAL EXTRAMEDULLARY

- Schwannoma
- Meningioma

EXTRADURAL

- Metastatic

CLINICAL PRESENTATION



- PAIN:-75% OF PTS (long prodrome, mths-yrs)
- NUMBNESS
- WEAKNESS:-75%
- SENSORY CHANGES:-65%
- SPHINCTER DYSFUNCTION:-15%
- CONUS MEDULLARIS & FILUM TERMINALE:-BOWEL & BLADDER INVOLVEMENT

DIAGNOSTIC WORK UP

HISTORY

PHYSICAL EXAMINATION

NEUROLOGICAL EXAMN:-

MOTOR,SENSORY AND REFLEXES

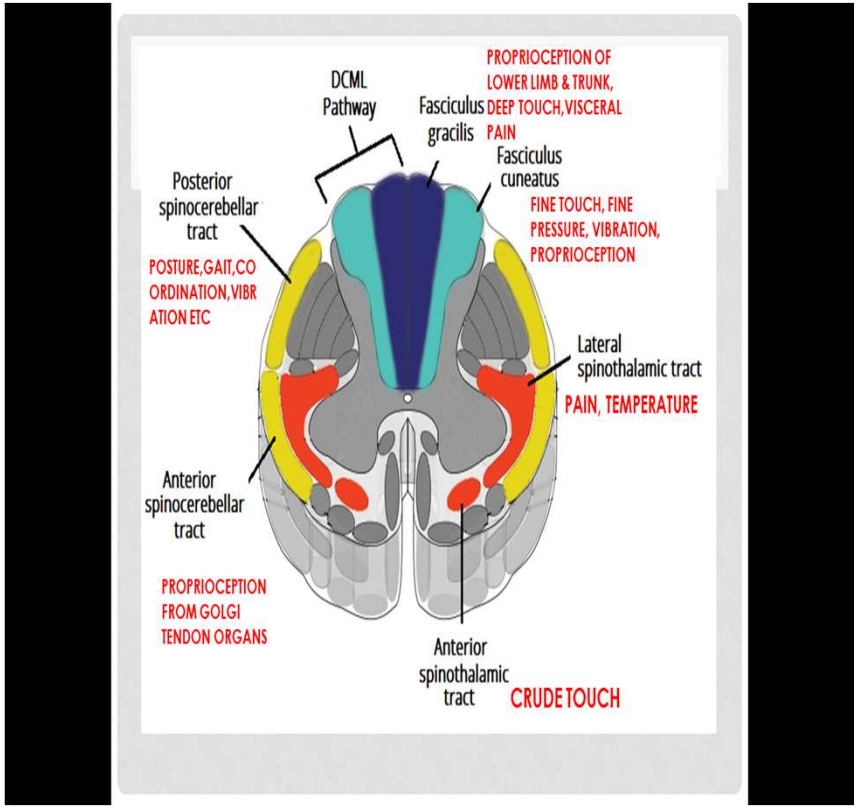
- **BROWN SEQUARD SYNDROME**
- **AUTONOMIC REFLEXES**
- **BOWEL AND BLADDER SYMOTOMS FOLLOWED BY PAIN:-FILUM TERMINALE**

IMAGING

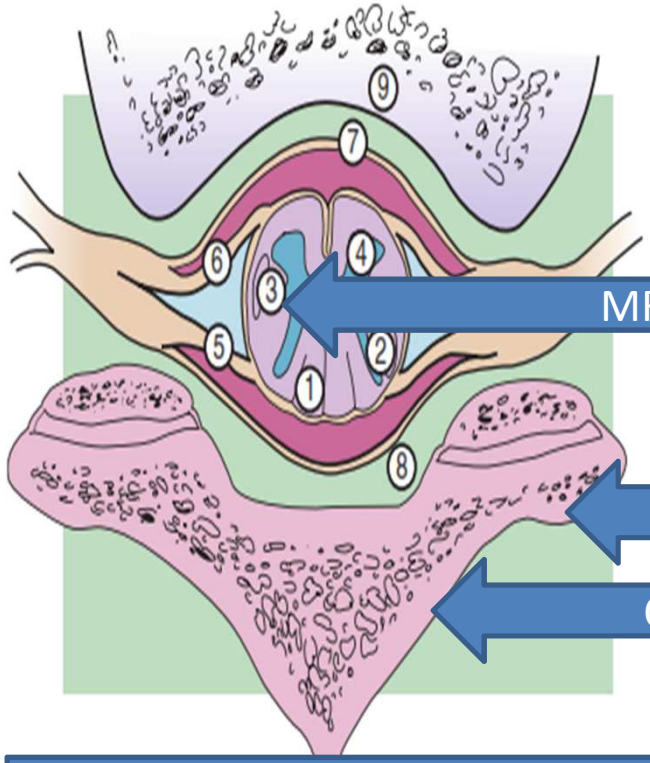
- **PLAIN X-RAY**
- **MRI OF WHOLE SPINE**
- **CT SCAN**
- **INTRA OP USG**

LAB STUDY

- **CSF CYTOLOGY**
- **CSF BIOCHEMISTRY**
- **NO CSF STUDY BEFORE MRI, AS SYMPTOMS MAY INCREASE**



NEURO IMAGING



MRI

IDEAL FOR INTRAMEDULLARY/EXTRAMEDULLARY INTRA DURAL PATHOLOGY
TOTAL NEURO AXIS EVALUATION
SAG,CORONAL,,T1,T2,CONTRAST STUDY
ALL SPINAL CORD GLIOMAS ENHANCES WITH CONTRAST
LIPOMAS-BRIGHT ON T1 WITHOUT CONTRAST
AND SIGNAL DISAPPEARS ON FAT SUPPRESSION

PLAIN X-RAY

CALCIFICATION
EROSION OF PEDICLE
SCALLOPING OF POST WALL OF SPINAL CORD

CT SCAN

IDEAL FOR EXTRA DURAL PATHOLOGY

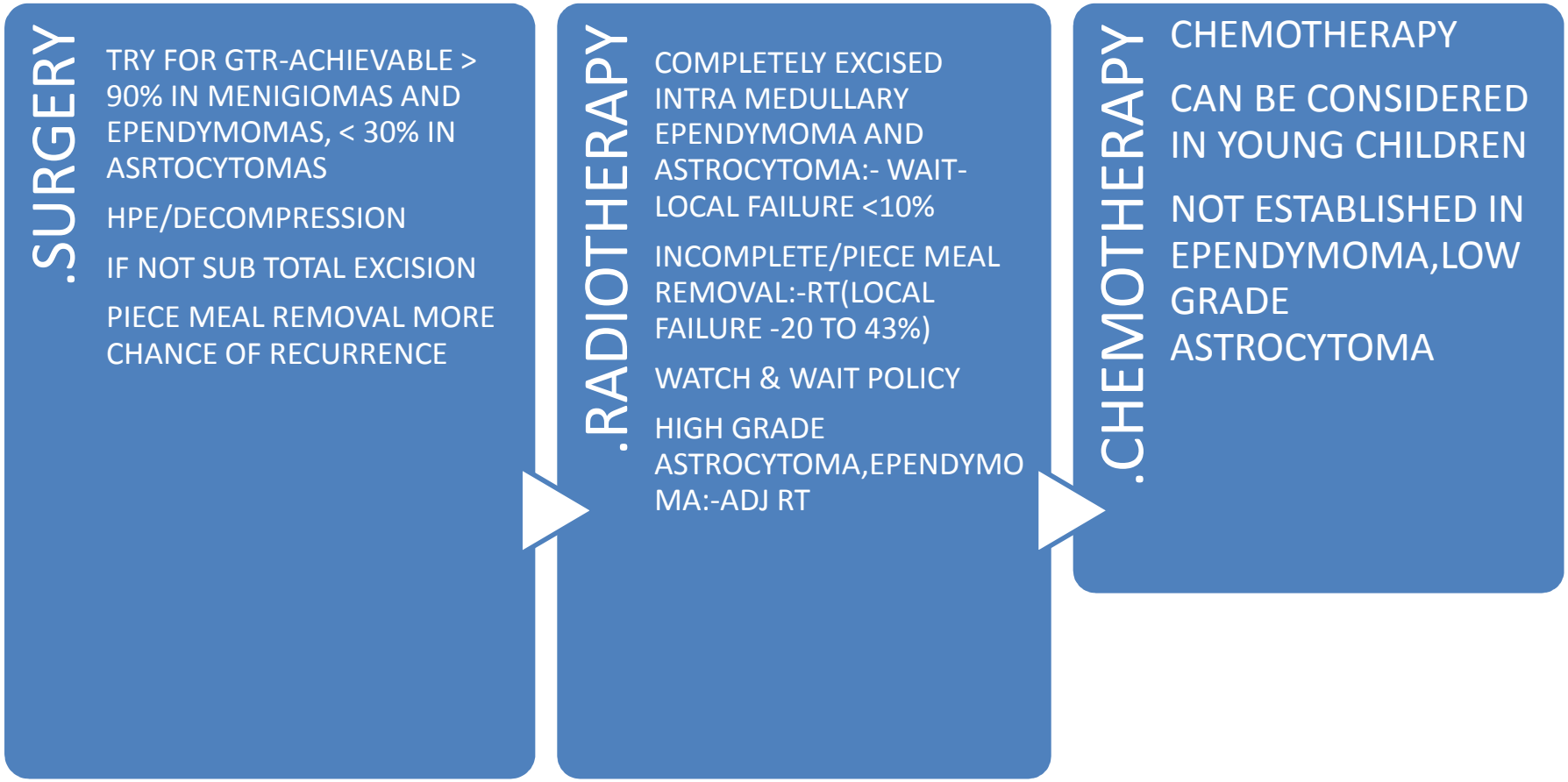
IMAGING OF ENTIRE NEURO AXIS IS RECOMMENDED AT THE INITIAL WORK UP IN CASE OF EPENDYMOMA/GBM/AA

PROGNOSTIC FACTORS

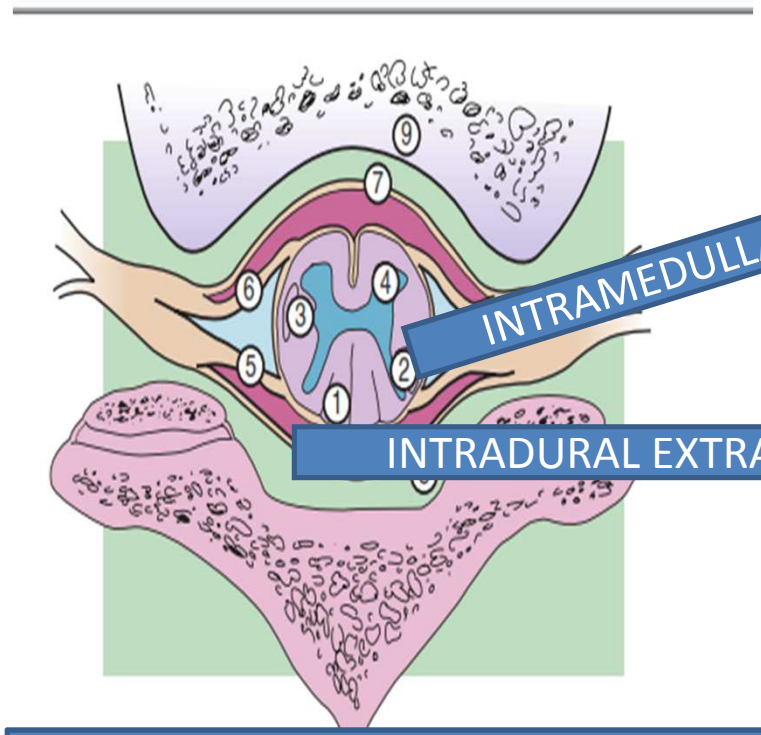
- **TUMOR RELATED** –
  - **Histologic type:-**EPENDYMOMA BETTER SURVIVAL THAN ASTROCYTOMA
    MYXOPAPILLARY BETTER SURVIVAL THAN OTHERS EPENDYMOMA
    CAPSULAR EPENDYMOMA BETTER SURVIVAL THAN UNCAPSULATED ONE
  - **Grade:-**HIGH GRADE POOR SURVIVAL
  - **Tumor extent:-** EXTENSIVE INVOLVEMENT HAVE POOR SURVIVAL
  - **Location :-**ROSTRAL LESION WORST SURVIVAL
- **PATIENT RELATED**
  - **Age :-** YOUNGER AGE BETTER SURVIVAL
  - **Presenting neurologic function:-**FEWER SYMPTOMS,BETTER NEUROLOGICAL FUNCTION BETTER SURVIVAL
- **TREATMENT RELATED FACTOR**
  - **Tumor resectibility:** EXTENT OF SURGERY CORRELATES WITH TIME OF PROGRESSION

- PTS WITH ROSTRAL TUMOR HAVE A WORSE SURVIVAL AND NEUROLOGICAL OUTCOME THAN PTS WITH CAUDAL TUMOR
- ROSTRAL PART HAVE GREATER CONCENTRATION OF FUNCTIONS PER UNIT VOLUME
- ROSTRAL LESIONS MOSTLY ASTROCYTOMA,CAUDAL LESION EPENDYMOMA

TREATMENT


.SURGERY
TRY FOR GTR-ACHIEVABLE > 90% IN MENIGIOMAS AND EPENDYMOMAS, < 30% IN ASRTOCYTOMAS
HPE/DECOMPRESSION
IF NOT SUB TOTAL EXCISION
PIECE MEAL REMOVAL MORE CHANCE OF RECURRENCE
.RADIOTHERAPY
COMPLETELY EXCISED INTRA MEDULLARY EPENDYMOMA AND ASTROCYTOMA:- WAIT-LOCAL FAILURE <10%
INCOMPLETE/PIECE MEAL REMOVAL:-RT(LOCAL FAILURE -20 TO 43%)
WATCH & WAIT POLICY
HIGH GRADE ASTROCYTOMA,EPENDYMOMA:-ADJ RT
.CHEMOTHERAPY
CHEMOTHERAPY
CAN BE CONSIDERED IN YOUNG CHILDREN
NOT ESTABLISHED IN EPENDYMOMA,LOW GRADE ASTROCYTOMA

SURGERY



INTRAMEDULLARY

RADICAL TOTAL GROSS EXCISION
- MICRO SURGERY
- CUSA
- CO2 LASER
- INTRA OP USG
- SUB TOTAL EXCISION

INTRADURAL EXTRAMEDULLARY

COMPLETE EXCISION
SUB TOTAL EXCISION

PIECE MEAL EXCISION OF EPENDYMOMA:- MORE CHANCE OF RECURRENCE

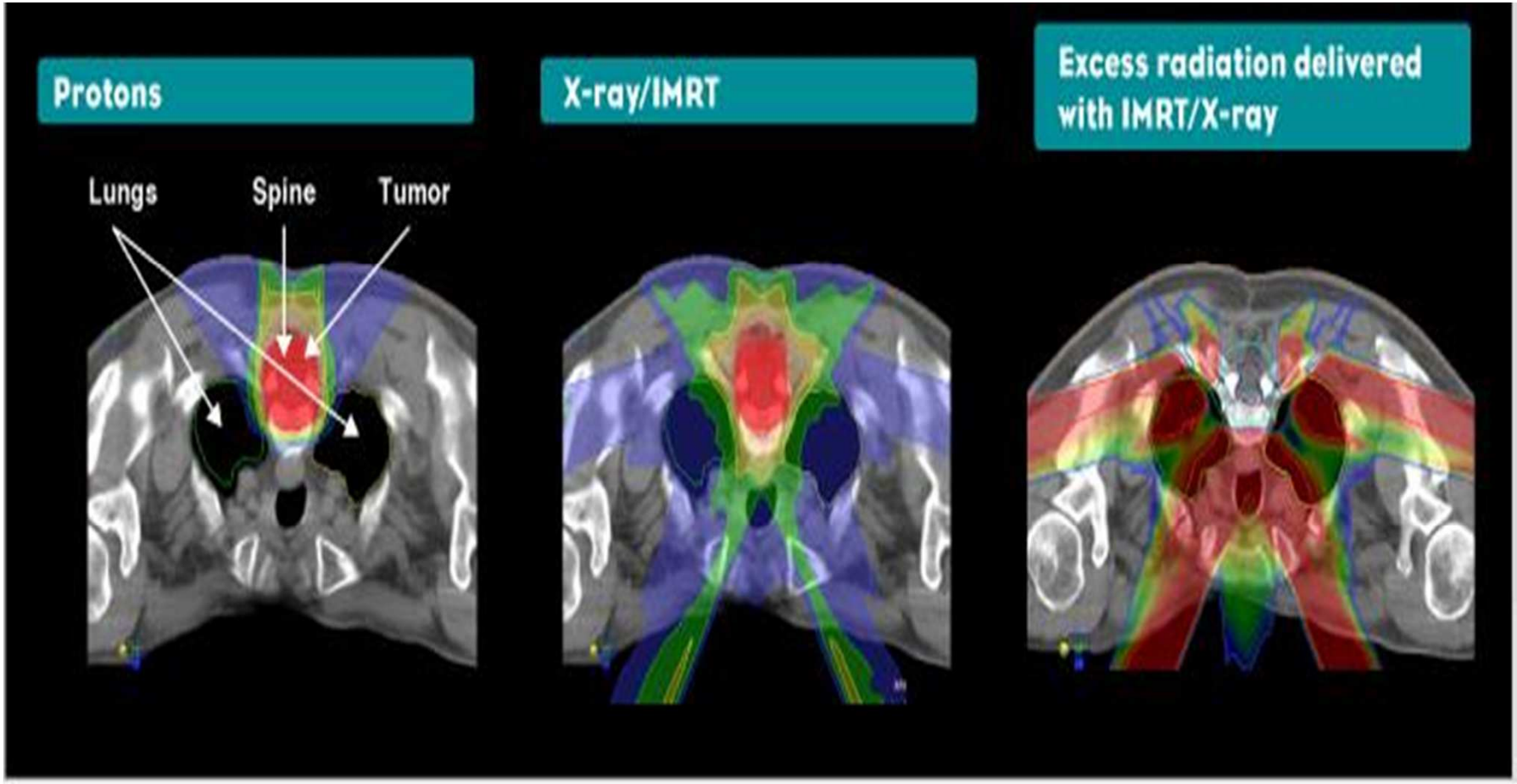
RADIOTHERAPY

- CONTROVERSIAL SINCE SC TUMORS ARE INDOLENT (SC TOXICITY)
- NOT INDICATED IN COMPLETELY EXCISED INTRA MEDULLARY EPENDYMOMA AND ASTROCYTOMA LOW GRADE
- INCOMPLETE / PIECE MEAL EXCISION :-ADJUVANT RADATION
- FOLLOW UP AFTER SURGERY AND SECOND SURGERY/ADJUVANT RT IF RECURRENCE PARTICULARLY IN CHILDREN
- RT INDUCED SPINAL DEFORMITY DUE TO DAMAGE OF EPIPHYSEAL PLATE, SOFT TISSUE FIBROSIS AND CONTRACTUE.
- MOST TUMORS ARE LOW GRADE ASTROCYTOMA/LOW GRADE EPENDYMOMA

- CLINICAL USE OF POST OP RT IS GUIDED BY THE PATTERN OF FAILURE
- IN HIGH GRADE ASTROCYTOMAS & EPENDYMOMAS ADJUVANT RT IS RECOMENDED IRRESPECIVE OF EXTENT OF RESECTION (PMH – RJ 2000, MAYO DATA RJ 1986)

RADIATION TECHNIQUE

- **CONVENTIONAL TECHNIQUE**
  - **CERVICAL:-PARALLEL OPPOSED PORTAL**
  - **THORACIC:-DIRECT POSTERIOR OR POSTERIOR WEDGE**
  - **LUMBAR & CAUDA EQUINA:-OPPOSED AP/PA FIELDS**
  - **IN FEMALE :-LATERAL TECHNIQUES ARE USED TO AVOID THE DOSE TO OVARY**
- **DEPTH OF SPINAL CORD:-DETERMINED FROM CT/MRI**
- **BEAM ENERGY:-**
  - **CERVICAL:-4 TO 6 MV**
  - **THORACIC & LUMBAR:-COMBINATION OF LOW ENERGY (4-6 MV) & HIGH ENERGY(8 TO 25 MV)**
- **DOSE:-**
  - **LOW GRADE ASTROCYTOMA AND EPENDYMOMA GTR/50.4GY AFTER STR**
  - **HIGH GRADE ASTROCYTOMA:-54 GY**
  - **HIGH GRADE EPENDYMOMA CSI = 36GY + BOOST 50.40GY – 54GY**
  - **MENINGIOMAS : 50.4 GY/1GY BD OR 16 GY/Fx 80% IDL BY SBRT**
- **MARGINS:-**
  - **3-5CMS, THECAL SAC AT S2-3 NEEDS TO BE COVERED IN CAUDAL EPENDYMOMAS**
- **IMRT**
- **SRT,PROTON BEAM THERAPY**


Protons
X-ray/IMRT
Excess radiation delivered with IMRT/X-ray
Lungs
Spine
Tumor

RADIATION TOXICITY

- **REVERSIBLE MYELOPATHY (2-6 MTHS) – L'HERMITTE'S SIGN IS SEEN CHARACTERIZED BY SHOCK LIKE SENSATION IN HANDS AND FEET WHEN NECK IS FLEXED. LASTS FOR WEEKS . REQUIRES NO TREATMENT.**

- **PROGRESSIVE MYELOPATHY (13-29 MTHS) - CHARACTERIZED BY PARASTHESIA,→ PROGRESSIVE MOTOR WEAKNESS →PAIN/TEMP LOSS→BOWEL/BLADDER DYSFUNCTION**

- **SPINAL CORD TOLERANCE– QUANTEC GUIDELINES SHOW FOR CONVENTIONAL RT A DOSE OF 50GY,60GY,69GY IS ASSOCIATED WITH 0.2%,6%.50% RATES OF MYELOPATHY.**

CHEMOTHERAPY

- LIMITED ROLE
- USED AFTER ALL MODALITIES ARE EXHAUSTED OR IN <3 YR AGE GROUP TO BORROW TIME FOR RT
- PLATINUMS & ETOPOSIDES ARE MOST ACTIVE AGENTS FOR EPENDYMOMA
- TEMOZOLAMIDE MAY BE USED IN SPINAL GLIOMA. USED AS CONCURRENT WITH RT AND THEN MAINTAINANCE THERAPY
- IN<3YEAR AGE GROUPS– INTENSIVE TREATMENT WITH CARBOPLATIN,PROCARBAZINE, VINCRISTINE, CYCLOPHOSPHAMIDE, ETOPOSIDE,CISPLATIN AGENTS

ASRTROCYTOMA

- AGE– PAEDIATRIC AND ADOLESCENT
- SITE – MOSTLY CERVICAL & THORACIC
- MOSTLY FOCAL BUT WHOLE CORD INVOLVEMENT MAY OCCUR.
- MAJORITY ARE LOW GRADE (WHO GR I & II)
- ACCOMPANYING SYRINX IN 40 % CASES.
- COMPLETE SURGICAL RESECTION IS OFTEN IMPOSSIBLE.
- JUVENILE PILOCYTIC ASTROCYTOMA (WHO GR I) DUE TO ITS NON INFILTRATIVE NATURE CAN BE TREATED WITH RADICAL RESECTION
- FIBRILLARY(GR II), ANAPLASTIC (GR III), GBM(GR IV)– DUE TO LOCAL INFILTRATION RESECTION ENBLOCK NOT POSSIBLE → ONLY BIOPSY/SUBTOTAL RESECTION DONE → ADJUVANT RADIOTHERAPY

EPENDYMOMA

- Seen in adults.
- Site – caudal location compared to astrocytoma.
- Variants – Cellular, Epithelial, Tanycytic, Subependymoma, Myxopapillary, Mixed type, Ependymo blastoma
- Rarely malignant.
- Sometimes associated with syrinx
- When associated with NF 2 are multiple
- Longer duration => Severe symptoms
- > 10 Yrs follow-up required for Ependymomas→ recurrence 5-10%

- MYXOPAPILLARY VARIANT OF EPENDYMOMA–
  - MOSTLY OCCURS IN FILUM TERMINALE.
  - HAS HIGH PROPENSITY FOR CSF SPREAD HENCE CSF CYTOLOGY SHOULD BE DONE
  - IF ENCAPSULATED CAN BE RESECTED EN-BLOCK
  - BUT WHEN IT IS ADHERENT/INFILTRATIVE PIECEMEAL RESECTION DONE → HIGH PROBABILITY OF LOCAL RECURRENCE → NEED ADJUVANT RADIOTHERAPY

NERVE SHEATH TUMORS

- ARISE FROM SCHWANN CELLS
- TYPES – SCHWANNOMA, NEUROFIBROMA,GANGLIONEUROMA
- MAJORITY ARE INTRADURAL ARISING FROM DORSAL SENSORY NERVE ROOT
- WHEN IT HAS BOTH INTRA AND EXTRA DURAL COMPONENT IT IS CALLED DUMBBELL LESION.

SCHWANNOMA –

- MOST FREQUENTLY SEEN IN CERVICAL AND LUMBAR REGION
- PRESENT WITH RADICULAR SENSORY CHANGE WITH WEAKNESS BEING A LESS COMMON SIGN
- MOSTLY SOLITARY BUT WHEN ASSOCIATED WITH NF2 & SCHWANNOMATOSIS THESE ARE MULTIPLE
- PATHOLOGICAL TYPES – CONVENTIONAL (MC),CELLULAR,MELANOTIC ,PLEXIFORM(DO NOT UNDERGO MALIGNANT CHANGE)
- GROW ECCENTRICALY WITHOUT NERVE INFILTRATION
- TREATMENT – RADICAL SURGICAL RESECTION(HEMILAMINECTOMY)
- RECURRENCE IS RARE AND THEY USUALLY DONOT UNDERGO MALIGNANT TRANSFORMATION.

(2)**NEUROFIBROMA**

- COMMONLY SEEN IN NF1
- CERVICAL SPINE MOST COMMONLY AFFECTED
- OFTEN MULTIPLE, BENIGN
- ENCASES THE NERVE ROOT HENCE EXCISION WITHOUT SACRIFICING THE NERVE IS DIFFICULT

(3) **GANGLIONEUROMA**

- Mostly benign and paraspinal
- Arises from sympathetic nervous system
- Pathological types
    a. Ganglioneuroma (Extradural, Dumbbell)
    b. Gangliocytoma
    c. Gangliogangliomа (Intradural, Intramedullary)
-

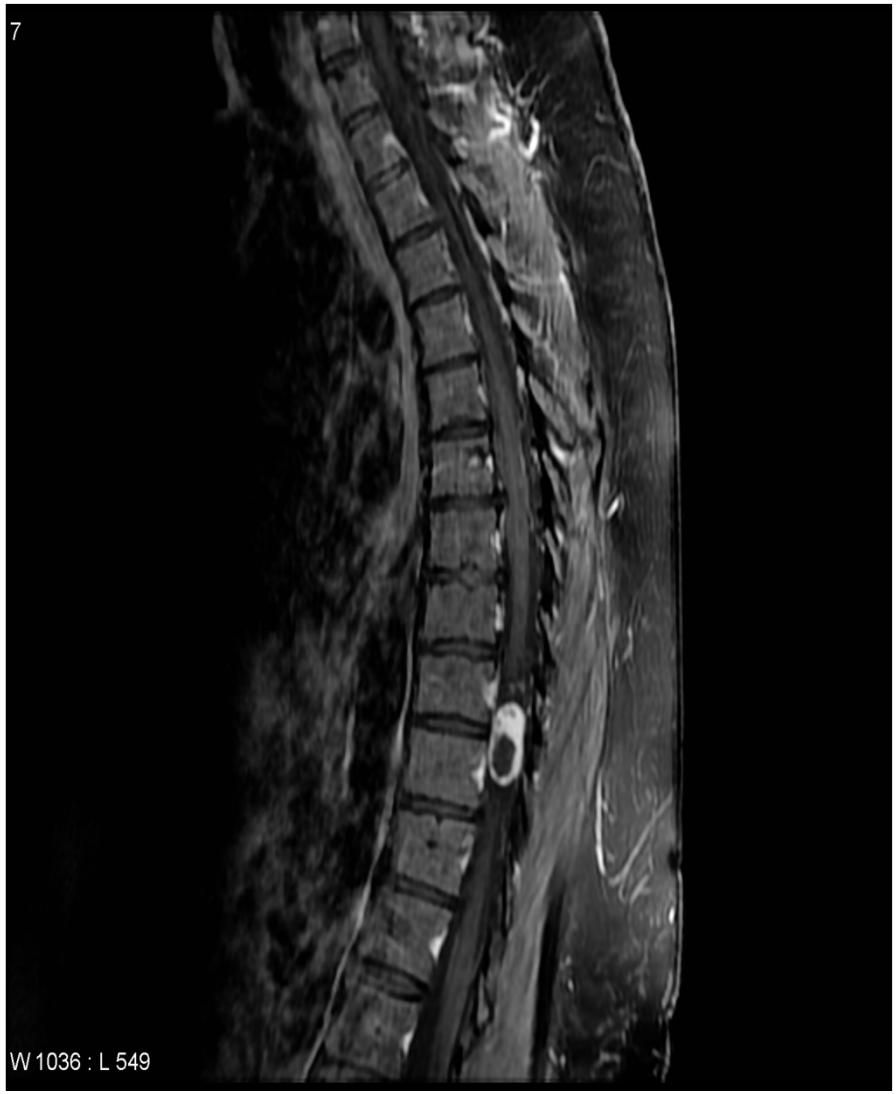
DIAGNOSIS

•X ray– posterior scalloping of vertebral body with widening of neural foramina

•CT– Hypodense paraspinal/ intraspinal mass

•MRI– T1- Hyperintense compared to muscle

T2 Very hyperintense with central hypo intense (target sign)



Meningioma

- SLOW GROWING,BENIGN
- WELL ENCAPSULATED
- 80% IN THORACIC AREA FOLLOED BY CERVICAL AREA
- DEFICITS ARE DRAMATICALLY REVERSIBLE
- TREATMENT IS TOTAL RESECTION INCLUDING DURAL ATTACHMENT & CAUTERISATION OF ADJACENT DURA
- SUBTOTAL RESECTION HAS HIGH RISK OF RECURRENCE.
- 90% ARE LOW GRADE, WHO GRADE I - II

DIAGNOSIS

•CT– ISO/HYPERDENSE LESION WITH INTENSE CONTRAST ENHANCEMENT

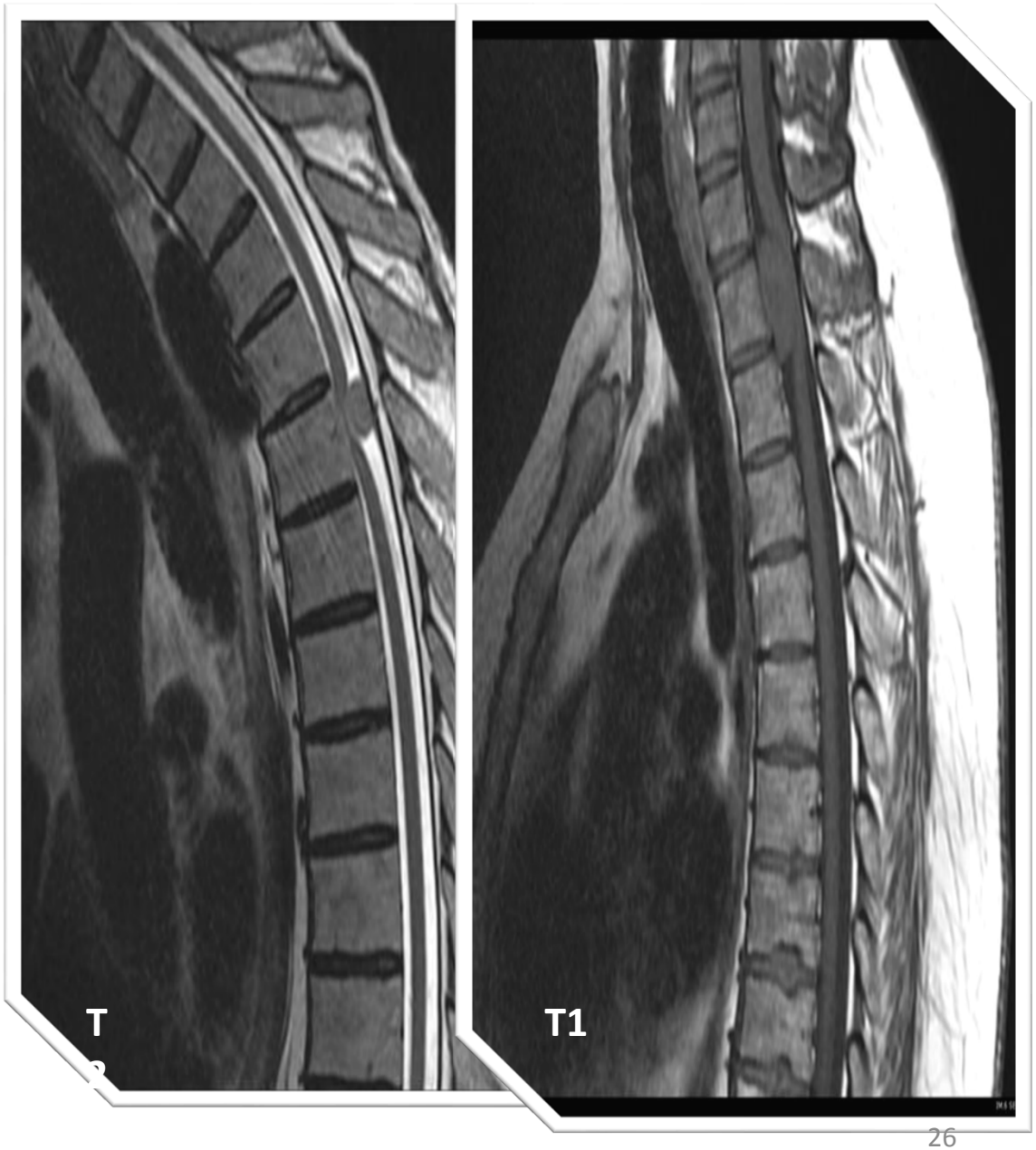
20-30% HAVE CALCIFICATIONS

•MRI–

T1- HYPO/ISO INTENSE TO CORD, WELL CIRCUMSCRIBED

T2 -SLIGHT HYPERINTENSE TO THE CORD

CALCIFICATION MAY BE DETECTED CSF VASCULAR CLEFT SIGN, DURAL TAIL MAY BE SEEN.



HEMANGIOBLASTOMA

- HIGHLY VASCULAR TUMORS
- MAY BE ASSOCIATED WITH VON- HIPPEL-LINDAU
- USUALLY DORSAL
- SOMETIMES MULTIPLE
- RENAL CELL CARCINOMA MUST BE SEARCHED FOR
- TREAMENT – SURGICAL RESECTION

Thank You