

# Spinal Tumors

## Good, Bad, and Ugly

Sohail Bajammal, MB ChB, MSc

September 18, 2006

Hamilton General Hospital – Weekly Spine Rounds

*Permission to use for educational uses with reference to the source*

# 1<sup>st</sup> Objective

*To recognize red flags and  
avoid delayed diagnosis*

## 2<sup>nd</sup> Objective

*Most of what  
you need to know  
for the Royal College Exam*

# Things to cover

- Characteristics
- Presentation
- Evaluation
- Treatment

# Tumors of the Spine

- Vertebral tumors:
  - Primary: 2-5%
    - Primary benign
    - Primary malignant
    - Benign more common than malignant
  - Secondary (metastases): 95-98%
- Spinal cord tumors (neurosurgery):
  - Extra-dural
  - Intra-dural:
    - extra-medullary
    - intra-medullary

# Facts for Differential

- Young patients (20s-30s) more likely to be benign tumors (except osteosarcoma and Ewing's sarcoma)
- Benign lesions tend to favor posterior elements

# Benign Tumors

# Osteoid Osteoma

- Age: 1<sup>st</sup> 3 decades, peak 15
- 10-25% of osteoid osteoma occur in spine
- 70% of painful juvenile scoliosis are due to osteoid osteoma
- Pain, worse at night, responds to NSAIDs

# Osteoid Osteoma

- Investigations:
  - X-ray: nidus surrounded by a halo, scoliosis
  - Bone scan: most sensitive, target sign
  - CT: most specific
- Treatment:
  - Medical: NSAIDs, observation
  - Surgical: if progressing or not responding
    - Excision
    - Percutaneous radiofrequency ablation

# Osteoblastoma

- 40% of osteoblastoma involves spine
- Histologically similar to osteoid osteoma, but larger in size, different presentation
- Clinical Presentation:
  - Pain, activity, not as responsive to NSAIDs
  - Larger size (>2cm) → cortical expansion → radiculopathy
  - Scoliosis: less common than osteoid osteoma

# Osteoblastoma

- Investigations:
  - X-rays: more readily detected
- Treatment:
  - Slowly progressive → less likely non-operative
  - Surgical:
    - Curettage: 15% recurrence, 50% in high grade
    - Marginal resection: less recurrence
- Malignant transformation: very rare

# Giant Cell Tumor (GCT)

- Characteristics:
  - 5-10% of GCT involves spine
  - Most common in sacrum
  - Vertebral body
- Clinical Presentation:
  - Age: 30s-40s, more in women
  - Variable: slowly growing to locally aggressive with metastases
  - Delayed of diagnosis
  - Pain and radiculopathy

# Giant Cell Tumor (GCT)

- Investigations:
  - X-rays: well-demarcated, radiolucent lesion with cortical expansion and local remodeling
- Treatment:
  - En bloc resection: optimal, but higher morbidity
  - Curettage: acceptable option, higher recurrence
- Prognosis:
  - Poorer than GCT in appendicular skeleton
  - Recurrence: 80% in stage III
  - Metastases to lung: 10%

# Osteochondroma

- <10% of all osteochondroma
- More in cervical
- If multiple → hereditary multiple exostoses
- Slowly growing → rare mechanical or compressive symptoms
- Treatment:
  - Mainly observation
  - Resection: if symptomatic

# Eosinophilic Granuloma (EG) Langerhans Cell Histiocytosis

- Benign, self-limiting process of well-demarcated bone resorption, ? etiology
- 1st – 2<sup>nd</sup> decade, Male 2:1
- Spine involved in 10-15% of EG
- Common sites: skull, pelvis, ribs, shoulder
- Associated with 2 systemic diseases:
  - Hand-Schüller-Christian disease
  - Letterer-Siwe disease

# Eosinophilic Granuloma (EG)

## Langerhans Cell Histiocytosis

- Investigations:
  - Spine X-rays: Vertebra plana (D/D)
  - Skeletal survey
  - Abdominal U/S: hepatosplenomegaly (HSC)
- Treatment:
  - Observation: because self-limiting
  - Surgical resection: if progressive kyphosis or progressive neurological symptoms
  - Low dose radiotherapy: if not amenable for surgery

# Vertebra Plana (FETISH)

- Fracture
- Eosinophilic granuloma
- Tumor
  - Metastases
  - Myeloma
  - Ewing's
  - Osteosarcoma
  - ABC
- Infection
  - TB
  - Osteomyelitis (disc involvement)
- Steroids
- Hemangioma



# Aneurysmal Bone Cyst (ABC)

- Characteristics:
  - Spine involved in 10-30% of ABC
  - Posterior element of thoracolumbar spine
  - May involve multilevel adjacent segments
  - 1<sup>st</sup> – 2<sup>nd</sup> decade
- Investigations:
  - X-rays: cortical expansion and thinning, “bubbly” appearance
  - MRI: fluid/fluid level

# Aneurysmal Bone Cyst (ABC)

- Treatment Options:
  - Curettage
  - Wide local excision
  - Embolization
  - Radiation
- Prognosis:
  - Recurrence: 15-30%

# Hemangioma

- Characteristics:
  - Most common tumor of the spine
  - Commonly incidental finding
  - 10% of autopsy
  - Single lesion in 2/3 of cases
  - Mainly in vertebral body, thoracic spine
- Clinical Presentation:
  - Neural compression by cortical or soft tissue expansion

# Hemangioma

- Investigations:
  - X-rays:
    - Able to detect only if involves 30-40% of body
    - Vertical trabecular striations like a honeycomb
  - CT or MRI: for subtle lesions
- Treatment Options:
  - Low dose radiation
  - Embolization
  - Surgical resection & stabilization: if instability
  - Vertebroplasty and kyphoplasty

# Differential Diagnosis (Anterior Spine)

- Non tumor
  - Infection (discitis)
  - TB
- Benign
  - Neurofibroma
  - Hemangioma
  - GCT
  - EG
  - ABC (more posterior)
- Malignant
  - Metastases
  - Myeloma

# Differential Diagnosis (Posterior Spine)

- Benign
  - Osteochondroma
  - Osteoblastoma
  - Osteoid osteoma
  - ABC
- Malignant
  - Metastases

# Primary Malignant Tumors

# Osteosarcoma

- 3-14% of malignant tumors of spine
- 2% of all osteosarcoma in the body
- Mainly in vertebral body, lumbosacral
- Bimodal age:
  - 10 – 25 yr: primary
  - Older than 50yr: secondary (radiation, Paget's)
- Many histological types
- Poorer prognosis and older age than appendicular osteosarcoma

# Osteosarcoma

- Treatment:
  - Neoadjuvant chemotherapy → surgical resection → Adjuvant chemotherapy
  - If not amenable for surgical resection: chemo and radiotherapy
- Bad prognostic factors:
  - Metastases at diagnosis
  - Large size
  - Sacral location
  - Intralesional resection

# Chondrosarcoma

- Characteristics:
  - 2<sup>nd</sup> most common primary malignant bone tumor (after chordoma)
  - 7-12% of all spine tumors
  - Age: 40s, more in men
- Treatment:
  - Resistant to radiotherapy and chemotherapy
  - Surgical excision

# Chordoma

- Characteristics:
  - Most common primary malignant tumor of spine (excluding lymphoproliferative disorders)
  - Age: 50s – 60s, Males: 3x more common
  - Remnants of the primitive notochord → midline
  - Sacrococcygeal > Base of skull > V. body (C)
- Clinical Presentation:
  - Gradual onset, disregarded, Pain, numbness, weakness, constipation or incont.
  - Sacrococcygeal lesions palpable by DRE

# Chordoma - Diagnosis

- X-rays: midline, lytic or mixed lytic and blastic
- CT: check involvement of local structures (rectum, vessels)
- MRI: check involvement of dura & roots
- Biopsy: posterior midline, never trans-rectal
  - Histology: lobular framework of physaliphorous cells

# Chordoma - Treatment

- Highly resistant to chemo and radiotherapy
- Radiotherapy for positive margins or palliative
- Lesions above S3: usually requires anterior and posterior approach for excision
- Unilateral retention of all roots: near normal bowel, bladder, and sexual function
- Sacrificing S2 → incontinent
- Metastases: liver, lungs, lymph nodes, peritoneum

# Multiple Myeloma

- Characteristics:
  - B-cell lymphoproliferative diseases
  - Rapidly progressive and highly lethal (20% survival at 5 yr)
  - Age: 60s – 70s
- Investigations:
  - X-ray: looks normal; Bone scan: cold
  - CT and MRI: delineate lesion
  - Serum and urine protein electrophoresis
  - 20% of cases: only urine is positive

# Multiple Myeloma - Treatment

- Very radiosensitive → main modality
- Chemotherapy for systemic component
- Bracing: for lesion <50% of vertebral body
- Surgery indicated for:
  - Stabilization of the spine
  - Decompression of neurological elements
  - Local control if recurrence or no response to radiation therapy
- Follow with MRI and serum electrophoresis

# Metastatic Tumors

# Significance

- The spine is the most common site for skeletal metastases
- Metastatic lesions are the most common tumors of the spine (95-98%)
- Vertebral body affected first
- Approximately 70% of patients who die of cancer have evidence of vertebral metastases on autopsy

# Common Primary Sites

- Breast (21%)
- Lung (14%)
- Prostate (7.5%)
- Renal (5%)
- GI (5%)
- Thyroid (2.5%)

# Level of Metastases

- Thoracolumbar 70%
- Lumbosacral 20%
- Cervical 10%

# Clinical Presentation

- Pain (85%)
  - Hyperemia, expansion, nerve compression, cord compression, pathologic fractures & instability
- Weakness (34%)
  - Spinal cord compression in 20%
- Mass (13%)
- Constitutional Symptoms

# History

- Age: high level of suspicion
- Details of the pain:
  - insidious or acute,  $\pm$  trauma, axial,  $\pm$  radiculopathy, unrelenting, non-mechanical, worse at night, change in features if chronic
- Personal history of cancer
- Constitutional symptoms
- Review of systems: thyroid, breast, chest, GI, GU & skin
- Any age-specific screening tests by GP
- Social history: smoking, alcohol, exposure to carcinogen
- Family history of malignancy

# Physical Exam

- Thorough examination of thyroid, breast, lung, abdomen, pelvis, prostate, skin, lymph nodes (referrals)
- Spine:
  - Look: alignment
  - Feel: focal tenderness
  - Move: ROM
  - Neurological examination: gait, power, sensation, reflexes (DTR, abdominal, Babinski, Hoffman), clonus

# Evaluation

- History
- Physical Exam
- Laboratory:
  - CBC, ESR, CRP, Lytes, BUN, Creatinine
  - Ca, PO<sub>4</sub>, Alk Phosph
  - Urinalysis: routine, Bence-Jones Proteins
  - Special: PSA, thyroid Fxn, serum and urine protein electrophoresis, liver function tests, stool guaiac, CEA
- Radiological
- Biopsy

# Radiological Evaluation

- Local:
  - X-ray of spine: AP, lateral, oblique
    - “winking owl” sign: pedicle destruction
    - Vertebral body destruction is not visible until 30-50% of trabeculae are involved
    - Negative x-ray does not rule out tumor
  - Bone Scan: screening, cold in MM
  - CT: bony architecture
  - MRI + gadolinium: gold standard



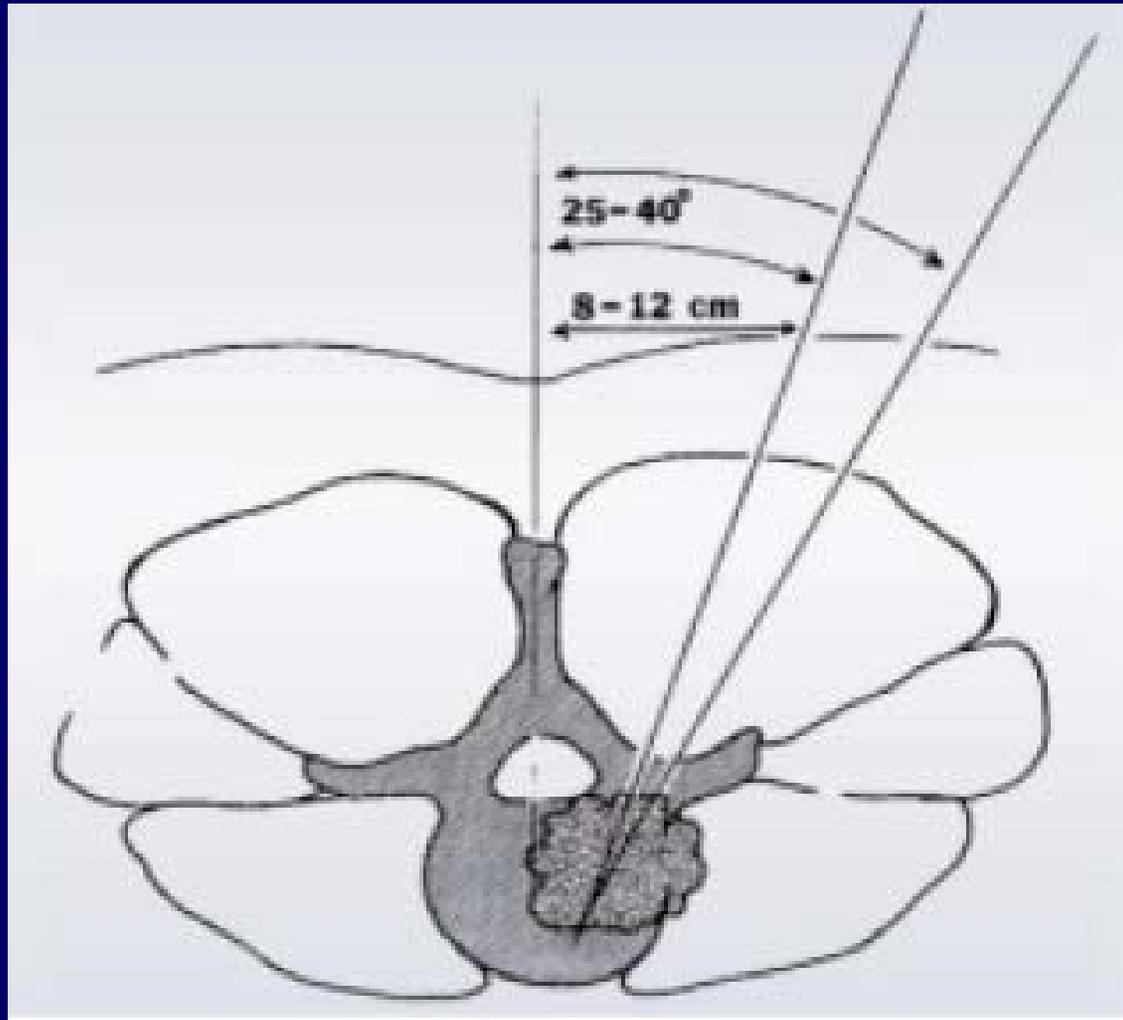
# Radiological Evaluation

- Staging:
  - CT chest, abdomen and pelvis with oral and IV contrast
  - Bone Scan
  - Mammogram

# Biopsy

- Indicated if primary diagnosis is unclear after workup:
  - Remote history of malignancy with long disease-free interval
- Options:
  - CT-guided: most accessible lesion, minimal morbidity, tattoo tract for later excision
  - Open: cost, delay, definitive for benign tumors
- Culture every tumor and biopsy every infection

# Percutaneous Biopsy



# Goal of Management

Maximize quality of life

# To achieve the goal....

- Provide pain relief
- Improve or maintain neurologic function
- Restore or maintain the structural integrity of the spinal column

# Options of Treatment

- Orthotic
- Steroids
- Radiotherapy
- Chemotherapy
- Hormonal Therapy
- Surgery
- Combination

**Multi-disciplinary approach**

# Pitfall

Aggressive chemotherapeutic regimens for patients with spinal pain not responding to conventional therapy without ruling out subtle mechanical etiology



Severe depression of bone marrow that surgery or radiotherapy are no longer feasible

# Decision Making (Prognostic Decision Rules)

- Frankel et. al. *Paraplegia* 1969
- Harrington *JBJS(A)* 1986
- Tokuhashi et. al. *Spine* 1990
- Tomita et. al. *Spine* 2001

# Frankel 1969

- A: Complete sensory & motor loss
- B: Complete motor loss; incomplete sensory loss
- C: Some motor function below level of involvement; incomplete sensory loss
- D: Useful motor function below level of involvement; incomplete sensory loss
- E: Normal motor & sensory function

# Harrington 1986

Non-operative

- I. No significant neurologic compromise
- II. Involvement of bone with minimal neurological impairment, but without collapse

Radio

- III. Major neurologic impairment without significant involvement of bone

Operative

- IV. Vertebral collapse with pain resulting from mechanical causes or instability, but with no significant neurologic compromise
- V. Vertebral collapse or instability with major neurologic compromise

# Tokuhashi et. al. 1990

- Retrospective analysis of 64 patients
- Scoring system for preoperative evaluation of metastatic spine prognosis
- Six parameters employed, each 0-2
- Total score 0-12 predicts the surgical intervention (excisional vs. palliative)

Table 1. Evaluation System for the Prognosis of Metastatic Spine Tumors

	Score
1. General condition (performance status) Karnofsky	
Poor (PS 10-40%)	0
Moderate (PS 50-70%)	1
Good (PS 80-100%)	2
2. Number of extraspinal bone metastases foci	
$\geq 3$	0
1-2	1
0	2
3. Number of metastases in the vertebral body	
$\geq 3$	0
2	1
1	2
4. Metastases to the major internal organs	
Unremovable	0
Removable	1
No metastases	2
5. Primary site of the cancer	
Lung, stomach	0
Kidney, liver, uterus	1
Others, unidentified	2
Thyroid, prostate, breast	2
Rectum	2
6. Spinal cord palsy (Frankel's)	
Complete	0
Incomplete	1
None	2
Total = 12	

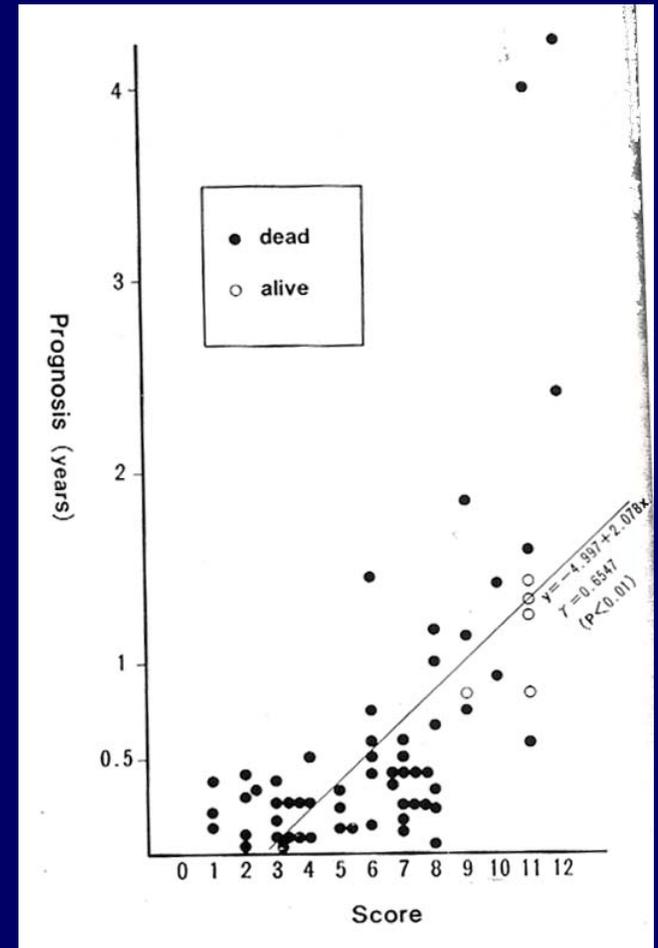
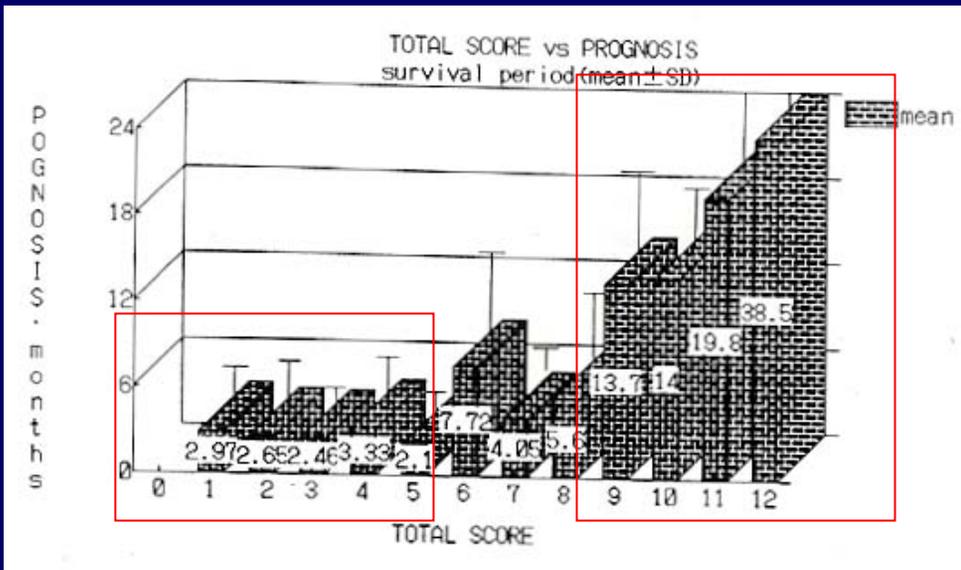
■  $\geq 9$ :

- Excision
- Survival > 12 months

■  $\leq 5$ :

- Palliative
- Survival < 3 months

# Tokuhashi et. al. 1990



# Tokuhashi et. al. 1990

- Validated by Enkaoua et. al. 1997
  - Retrospective analysis of 71 patients
- No statistical background of points 0, 1 & 2
- The important value of each prognostic factor was not considered.

# Tomita et. al. 2001

- Phase 1 (1987-1991):
  - Retrospective analysis of 67 patients to evaluate predictors
  - Hazard ratios were analyzed & standardized
- Phase 2 (1993-1996):
  - Prospective validation of 61 patients
- Total Score 2-10, based on:
  - Grade of malignancy of the primary tumor
  - Visceral Metastases to vital organs
  - Bone metastases

# Tomita et. al. 2001

## Phase 1

Scoring System			
Point	Prognostic factors		
	Primary tumor	Visceral mets.*	Bone mets.**
1	slow growth <small>(breast, thyroid, etc.)</small>	/	solitary or isolated
2	moderate growth <small>(kidney, uterus, etc.)</small>	treatable	multiple
4	rapid growth <small>(lung, stomach, etc.)</small>	un-treatable	/

\* No visceral mets. = 0 point.

\*\* Bone mets. including spinal mets.

# Tomita et. al. 2001

## Phase 1

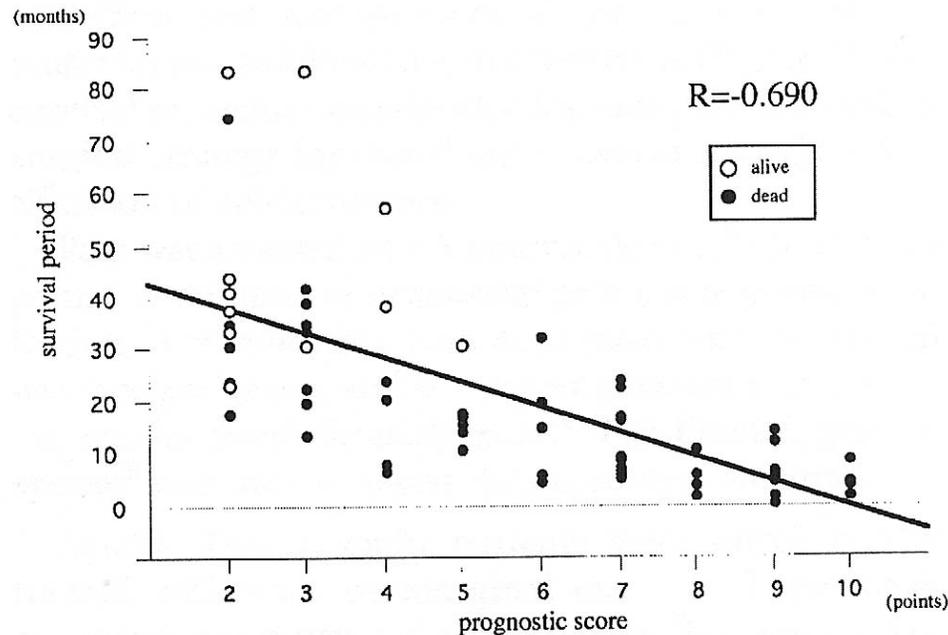


Figure 2. Relation between the prognostic score and the survival period. There was a high-grade correlation between the prognostic score and survival period, with a correlation coefficient of  $-0.690$  ( $P < 0.0001$ ).

# Tomita et. al. 2001

## Phase 2

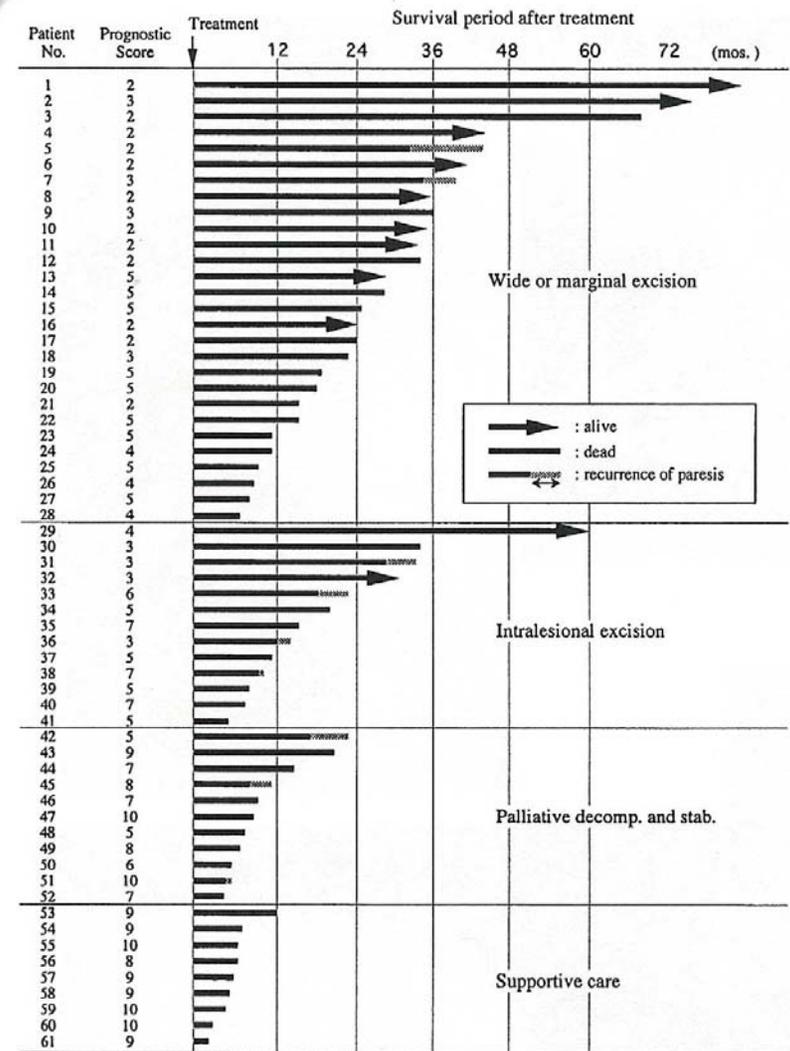


Figure 4. Survival periods of each patient treated with the surgical strategy for spinal metastases. Twenty-eight patients were oncologically treated with wide or marginal excision. Thirteen patients were treated with intralesional excision such as piecemeal subtotal excision, eggshell curettage, or through debulking. Eleven patients were treated with palliative decompression and stabilization. Nine patients were treated with terminal supportive care.

# Indications of Surgery

1. Intractable pain unresponsive to non-operative
2. Growing tumor resistant to other measures
3. Patients reached spinal cord tolerance after prior radiation therapy
4. Spinal instability: pathologic fractures, progressive deformity, or neurologic deficits
5. Clinically significant neural compression, especially by bone or bone debris
6. The need for definitive histologic diagnosis



# Spinal Instability

- White and Punjabi:

“the ability of the spine, under physiologic loads, to prevent initial or additional neurologic damage, severe intractable pain, and gross deformity”
- Grubb & Kostuik:
  - 6 columns (3 columns of Denis, right and left):
    - if >3 involved → unstable
  - >20° angulation → unstable

# Spinal Instability

Taneichi et. al., Spine 1997

- 100 thoracic & lumbar osteolytic lesions followed
- Suggested that criteria of impending collapse:
  - Thoracic Spine (T1-T10)
    - 50-60% of vertebral body with no destruction of other structures
    - 25-30% of vertebral body and costovertebral joint destruction
  - Thoracolumbar & Thoracic Spine (T10-L5)
    - 35-40% of vertebral body
    - 20-25% of vertebral body with posterior element destruction

# Principles of Surgical Treatment

- Establish diagnosis, if not done
- Decompression
- Realignment
- Stabilization

# Staging Systems

(mainly for primary malignant tumors)

## Enneking Oncologic Staging System

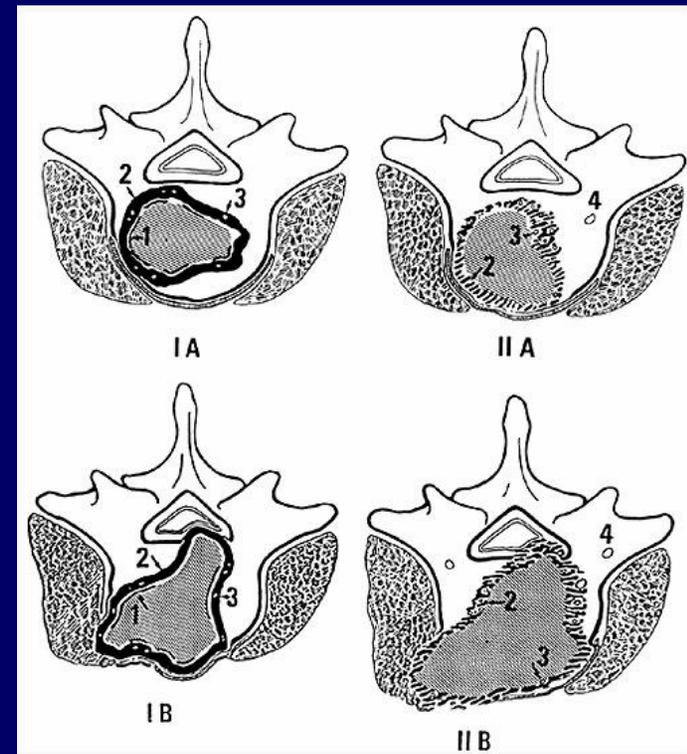
– Stage I: low grade

- A: intra-compartmental
- B: extra-compartmental

– Stage II: high grade

- A: intra-compartmental
- B: extra-compartmental

– Stage III: distant metastasis



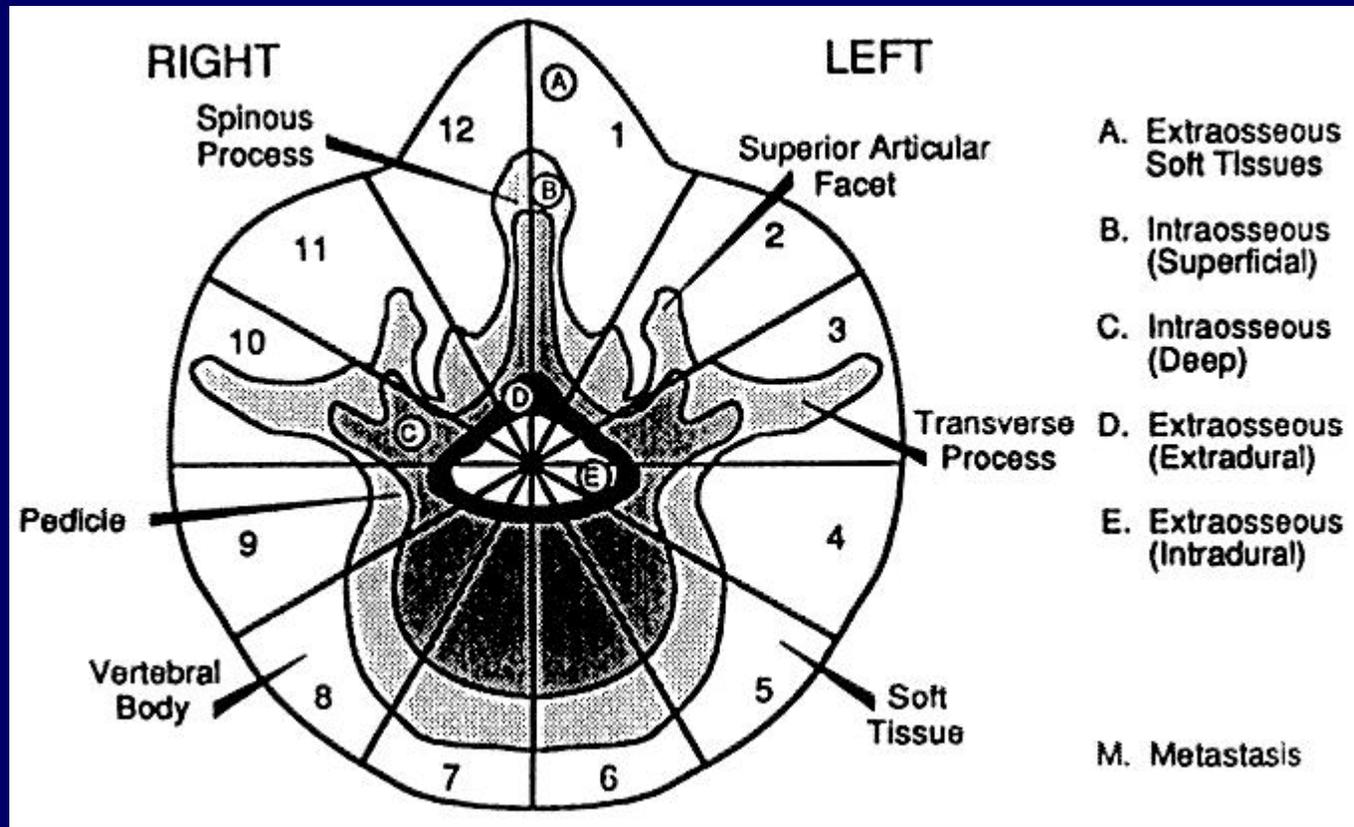
# Staging Systems

(mainly for primary malignant tumors)

## Boriani-Weinstein-Biagini Surgical Staging System (Spine 1997)

- 12 triangular segments (1-12 clockwise)
- 5 layers (A to E): soft tissue to dural
- Longitudinal extension: levels involved

# Boriani-Weinstein-Biagini



# Surgical Options

- Approach: anterior, posterior, A+P, or posterolateral
- Reconstruction: bone graft, cement, or cages
- Pre-operative embolization (renal cell ca, thyroid, Ewing's)
- ? Postoperative radiotherapy: after 3-6 wks

# Radiotherapy

- Goal is to debulk, promote calcification or ossification (3 months), relieve pain (90-90%)
- Of patients that are ambulatory at presentation, 70% will remain so
- Can be used when myelopathy due to soft tissue but not if due to bone or deformity (Harrington III)
- Combine with surgery if failure of radiation at that level (deformity or neurological worsening)

# Radiotherapy

- Radiosensitivity
  - Myeloma & Lymphoma: most radiosensitive
  - Prostate, Breast, Lung and Colon: moderately
  - Thyroid, Kidney, Melanoma: not radiosensitive
- Dose
  - 5,000 cGy in 25 fractions over 5 weeks (C & L-spine)
  - 4,500 cGy over 4½ -5 weeks in T-spine

# Radiotherapy As Only Treatment

- Radiosensitive tumor not previously irradiated
- Stable or slowly progressive neurological deficit
- Soft-tissue spinal canal compromise (not bone)
- Widespread spinal metastases with multilevel neural compression
- No evidence of spinal instability
- Patient's condition (or prognosis) precludes surgery

# Adjuvant Radiotherapy

- Done after operative stabilization / decompression
- Wait 3 weeks for wound healing before starting radiation
- If allograft / autograft bone was used, wait 6/52 for incorporation before starting

# Bottom Line

- Tumor type
- Tumor location
- Extent of spinal column involvement
- Number & distribution of metastases
- Life expectancy
- Neurologic status
- Comorbid medical conditions
- Nutritional status
- Immune status
- Patient & family wishes

# References

- OKU 8, AAOS, 2005
- OKU Spine 3, AAOS, 2006
- Core Knowledge in Orthopaedics - Spine
- ICL 49, 2000

**Thank You**