

Collège Hospitalier et Universitaire de Chirurgie Pédiatrique DESC de Chirurgie Pédiatrique

Session de Mars 2008 - PARIS

Tumeurs Malignes du Rachis

G. BOLLINI

We wilL exclude from this presentation the tumor involving the sacrum which are particular entities more prone to be discussed in a chapter entitled sacral and pelvi-sacral malignant bone tumor.





It is often difficult to determine whether a malignant bone tumor in children involve a rib and then spread to the adjacent vertebra(e) or if it is a primary malignant spine tumor invading the adjacent rib(s).

That the reason why in such cases in thoracic area we will discuss of costo-vertebral bone neoplasm



Primary malignant sarcomas of the spine are extremely rare. It has been estimated that they account for 2% to 10% of all primary bone neoplasms

4767 patients in the Vienna Bone Tumor Registry



1715 primary malignant tumors



44 spine tumors = 2.5%

As this localization of malignant bone tumor is very rare most of the publication in the literature deal with both children and adult patients.....

And often with benign as well as malignant spinal bone tumours

5 Ewing Sarcomas, 1 Chondrosarcoma, 1 Malignant Fibrous Histiocytoma, 1 Epitheliod Sarcoma

| Patient Number | Age/ Gender | Tumor Location | Histologic Diagnosis | Tumor Removal | Resection Margins | Local Recurrence |
|-------------------|----------------|-------------------|--------------------------------|---------------|--------------------------|---------------------|
| 1 | 37/M | L2 | Chondrosarcoma | En bloc | Free of tumor | No |
| 2 | 18/M | L4 | Chondrosarcoma | En bloc | Free of tumor | No |
| 3 | 15/M | L5 | Ewing's sarcoma | Piecemeal | Macroscopic tumor | No |
| 4 | 13/M | L2-L4 | Ewing's sarcoma | Piecemeal | Free of tumor | No |
| 5 | 22/M | C3-C7 | Chondrosarcoma | Piecemeal | Free of tumor | Yes |
| 6 | 67/M | L3 | Chondrosarcoma | Piecemeal | Microscopic tumor | No |
| 7 | 44/M | C3,C4 | Chondrosarcoma | Piecemeal | Microscopic tumor | No |
| 8 | 7/M | L2-L5 | Ewing's sarcoma | Piecemeal | Microscopic tumor | No |
| 9 | 27/M | T10-T12 | Myxoid liposarcoma | En bloc | Free of tumor | No |
| 10 | 44/F | T10 | Hemangioendothelial sarcoma | Piecemeal | Macroscopic tumor | No |
| 11 | 29/F | T10 | Giant cell tumor | En bloc | Free of tumor | No |
| 12 | 22/M | T2 | Ewing's sarcoma | En bloc | Free of tumor | No |
| 13 | 28/M | T10,T11 | Chondrosarcoma | En bloc | Free of tumor | No |
| 14 | 21/F | L4 | Chondrosarcoma | Piecemeal | Free of tumor | No |
| 15 | 32/M | C4 | Chondrosarcoma | Piecemeal | Macroscopic tumor | Yes |
| 16 | 19/M | L1 | Ewing's sarcoma | Piecemeal | Macroscopic tumor | Yes |
| 17 | 66/F | L3 | Chondrosarcoma | En bloc | Macroscopic tumor | Yes |
| 18 | 70/F | L3 | Osteosarcoma | Piecemeal | Microscopic tumor | Yes |
| 19 | 32/M | C4-T1 | Osteosarcoma | Piecemeal | Macroscopic tumor | Yes |
| 20 | 7/M | L1,L2 | Ewing's sarcoma | En bloc | Macroscopic tumor | No |
| 21 | 80/M | L3 | Fibroblastic osteosarcoma | Piecemeal | Microscopic tumor | No |
| 22 | 50/M | Τ7 | Osteogenic sarcoma | En bloc | Free of turnor | No |
| 23 | 62/F | L3 | Fibroblastic osteosarcoma | Piecemeal | Microscopic tumor | Yes |
| 24 | 16/F | C1,C2 | Malignant fibrous histiocytoma | Piecemeal | Macroscopic tumor | Yes |
| 25 | 72/M | T11 | Malignant fibrous histiocytoma | FILTOURC | Free of tumor | IN(O) |
| 26 | 4/F | C1,C2 | Epitheliod sarcoma | Piecemeal | Microscopic tumor | Yes |
| 27 | 34/F | L4,L5 | Osteosarcoma | En bloc | Microscopic tumor | Yes |
| 28 | 13/M | T7-T9 | Ewing's sarcoma | Piecemeal | Macroscopic tumor | No |
| 29 | 30/F | T7-T9 | Giant cell rich osteosarcoma | Piecemeal | Microscopic tumor | Yes |
| 30 | 72/M | T2,T3 | Myofibroblastic sarcoma | En bloc | Free of tumor | Yes |

Relationship Between Surgical Margins and Local Recurrence in Sarcomas of the Spine

Talac, Robert MD, PhD; Yaszemski, Michael J. MD, PhD; Currier, Bradford L. MD; Fuchs, Bruno MD; Dekutoski, Mark B. MD; Kim,
Choll W. MD, PhD; Sim, Franklin H. MD,Clin Ortop Rel Res Volume 397, April 2002, pp 127-132

Charles G. Fisher 26 patients

Table 1. Patient Diagnosis and Preoperative Staging

| 1 56 15 Chordoma S4-S5 1b N/A 2 56 78 MFH S1-S3 2b N/A 3 29 37 Osteosarcoma T5-T7 1b 8-12 4 53 79 Osteoblastoma (recurrent) T12-L2 S3 11-8 5 51 13 Osteoblastoma (recurrent) L5 S3 1-4 6 38 6 Pancoast tumor T3-T4 2b 2-5 7 31 17 Osteosarcoma T9-T11 2b 2-6 9 20 9 Osteosarcoma L1-L3 2b 1-6 10 53 13 Chordoma L5-Cx 1b N/A 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 3-4 13 20 12 Mesenchymal hamartoma C6-T1 <t< th=""><th>WBB Level</th></t<> | WBB Level |
|---|-----------|
| 2 56 78 MFH S1-S3 2b N/A 3 29 37 Osteosarcoma T5-T7 1b 8-12 4 53 79 Osteoblastoma (recurrent) T12-L2 S3 11-8 5 51 13 Osteoblastoma (recurrent) L5 S3 1-4 6 38 6 Pancoast tumor T3-T4 2b 2-5 7 31 17 Osteosarcoma T9-T11 2b 2-5 8 36 5 MPNST C5-T1 2b 2-6 9 20 9 Osteosarcoma L1-L3 2b 1-6 10 53 13 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 | N/A |
| 3 29 37 Osteosarcoma T5-T7 1b 8-12 4 53 79 Osteoblastoma (recurrent) T12-L2 S3 11-8 5 51 13 Osteoblastoma (recurrent) L5 S3 1-4 6 38 6 Pancoast tumor T3-T4 2b 2-5 7 31 17 Osteosarcoma T9-T11 2b 2-5 9 20 9 Osteosarcoma L1-L3 2b 1-6 10 53 13 Chordoma S1-S3 1b N/A 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 3-4 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma L | N/A |
| 4 53 79 Osteoblastoma (recurrent) T12–L2 S3 11–8 5 51 13 Osteoblastoma (recurrent) L5 S3 1–4 6 38 6 Pancoast tumor T3–T4 2b 2–5 7 31 17 Osteosarcoma T9–T11 2b 2–5 8 36 5 MPNST C5–T1 2b 2–6 9 20 9 Osteosarcoma L1–L3 2b 1–6 10 53 13 Chordoma S1–S3 1b N/A 11 64 6 Chordoma L5–Cx 1b N/A 12 41 4 Hemangicedothelioma L1 S2 3–4 13 20 12 Mesenchymal hamartoma C6–T1 S2 3–4 14 45 9 Giant cell tumor C7–T2 S3 7–12 15 67 6 Chordoma L4–S1 1b N/A 16 16 7 Chondrosarcoma L4 | A-D |
| 5 51 13 Osteoblastoma (recurrent) L5 S3 1-4 6 38 6 Pancoast tumor T3-T4 2b 2-5 7 31 17 Osteosarcoma T9-T11 2b 2-5 8 36 5 MPNST C5-T1 2b 2-6 9 20 9 Osteosarcoma L1-L3 2b 1-6 10 53 13 Chordoma S1-S3 1b N/A 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma L4 1b 6-10 18 50 4 Giant cell tumor C3 S3 <td>A-D</td> | A-D |
| 6 38 6 Pancoast tumor T3-T4 2b 2-5 7 31 17 Osteosarcoma T9-T11 2b 2-5 8 36 5 MPNST C5-T1 2b 2-6 9 20 9 Osteosarcoma L1-L3 2b 1-6 10 53 13 Chordoma S1-S3 1b N/A 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma L4 1b 6-10 18 50 4 Giant cell tumor C3 S3 4-11 19 50 5 Chordoma C2 1b 1-6 | A-C |
| 7 31 17 Osteosarcoma T9-T11 2b 2-5 8 36 5 MPNST C5-T1 2b 2-6 9 20 9 Osteosarcoma L1-L3 2b 1-6 10 53 13 Chordoma S1-S3 1b N/A 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2-T5 2b 9-12 17 65 3 Chordoma C2 1b 6-10 18 50 4 Giant cell tumor C3 S3 4-11 19 50 5 Chordoma C2 | A-C |
| 8 36 5 MPNST C5-T1 2b 2-6 9 20 9 Osteosarcoma L1-L3 2b 1-6 10 53 13 Chordoma S1-S3 1b N/A 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2-T5 2b 9-12 17 65 3 Chondrosarcoma L4 1b 6-10 18 50 4 Giant cell tumor C3 S3 4-11 19 50 5 Chordoma S2 1b N/A 20 70 4 Chordoma S2 | A-D |
| 9 20 9 Osteosarcoma L1–L3 2b 1–6 10 53 13 Chordoma S1–S3 1b N/A 11 64 6 Chordoma L5–Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4–9 13 20 12 Mesenchymal hamartoma C6–T1 S2 3–4 14 45 9 Giant cell tumor C7–T2 S3 7–12 15 67 6 Chordoma L4–S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2–T5 2b 9–12 17 65 3 Chondrosarcoma L4 1b 6–10 18 50 4 Giant cell tumor C3 S3 4–11 19 50 5 Chordoma C2 1b 1–6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 | A-E |
| 10 53 13 Chordoma S1-S3 1b N/A 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2-T5 2b 9-12 17 65 3 Chordoma L4 1b 6-10 18 50 4 Giant cell tumor C3 S3 4-11 19 50 5 Chordoma C2 1b 1-6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1-6 22 46 3 Cho | A-D |
| 11 64 6 Chordoma L5-Cx 1b N/A 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2-T5 2b 9-12 17 65 3 Chondrosarcoma C3 S3 4-11 19 50 4 Giant cell tumor C3 S3 4-11 19 50 5 Chordoma C2 1b 1-6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1-6 22 46 3 Chordoma S4-S5 1b N/A | N/A |
| 12 41 4 Hemangioedothelioma L1 S2 4-9 13 20 12 Mesenchymal hamartoma C6-T1 S2 3-4 14 45 9 Giant cell tumor C7-T2 S3 7-12 15 67 6 Chordoma L4-S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2-T5 2b 9-12 17 65 3 Chondrosarcoma L4 1b 6-10 18 50 4 Giant cell tumor C3 S3 4-11 19 50 5 Chordoma C2 1b 1-6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1-6 22 46 3 Chordoma S4-S5 1b N/A | N/A |
| 13 20 12 Mesenchymal hamartoma C6–T1 S2 3–4 14 45 9 Giant cell tumor C7–T2 S3 7–12 15 67 6 Chordoma L4–S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2–T5 2b 9–12 17 65 3 Chondrosarcoma L4 1b 6–10 18 50 4 Giant cell tumor C3 S3 4–11 19 50 5 Chordoma S2 1b 1–6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1–6 22 46 3 Chordoma S4–S5 1b N/A | A-D |
| 14 45 9 Giant cell tumor C7–T2 S3 7–12 15 67 6 Chordoma L4–S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2–T5 2b 9–12 17 65 3 Chondrosarcoma L4 1b 6–10 18 50 4 Giant cell tumor C3 S3 4–11 19 50 5 Chordoma S2 1b N/A 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1–6 22 46 3 Chordoma S4–S5 1b N/A | A-B |
| 15 67 6 Chordoma L4–S1 1b N/A 16 16 7 Chondrosarcoma 36 M T2–T5 2b 9–12 17 65 3 Chondrosarcoma L4 1b 6–10 18 50 4 Giant cell tumor C3 S3 4–11 19 50 5 Chordoma C2 1b 1–6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1–6 22 46 3 Chordoma S4–S5 1b N/A | A-D |
| 16 16 7 Chondrosarcoma 36 M T2-T5 2b 9-12 17 65 3 Chondrosarcoma L4 1b 6-10 18 50 4 Giant cell tumor C3 S3 4-11 19 50 5 Chordoma C2 1b 1-6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1-6 22 46 3 Chordoma S4-S5 1b N/A | N/A |
| 17 65 3 Chondrosarcoma L4 1b 6–10 18 50 4 Giant cell tumor C3 S3 4–11 19 50 5 Chordoma C2 1b 1–6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1–6 22 46 3 Chordoma S4–S5 1b N/A | A-C |
| 18 50 4 Giant cell tumor C3 S3 4–11 19 50 5 Chordoma C2 1b 1–6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1–6 22 46 3 Chordoma S4–S5 1b N/A | A–D |
| 19 50 5 Chordoma C2 1b 1-6 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1-6 22 46 3 Chordoma S4-S5 1b N/A | C-D |
| 20 70 4 Chordoma S2 1b N/A 21 18 9 Ewing's sarcoma 18 M L5 2b 1-6 22 46 3 Chordoma S4–S5 1b N/A | A-D |
| 21 18 9 Ewing's sarcoma 18 M L5 2b 1-6 22 46 3 Chordoma S4-S5 1b N/A | N/A |
| 22 46 3 Chordoma S4–S5 1b N/A | A-C |
| | N/A |
| 23 17 7 Osteoblastoma L1 S2 10–12 | A-C |
| 24 31 10 Chondrosarcoma T7-T10 1b 3-5 | A-C |
| 25 26 3 Ewing's sarcoma (recurrent) T2-T3 2b 1-6 | A-C |
| 26 57 20 Chondrosarcoma T1 2b 3–6 | A-B |

Note. A summary of patient demographic data and preoperative staging according to both the Enneking and Weinstein Boriani Biagini (WBB) classification (where applicable).

Fisher, Charles G. MD, MHSc; Keynan, Ory MD; Boyd, Michael C. MD; Dvorak, Marcel F. MD Spine Volume 30(16), 15 August 2005, pp 1899-1908

Hasegawa Kazuhiro 13 patients

| 20000000000 | 77-7004-55/550-660 | | | | | | |
|-------------|--------------------|-----|------------------------|--------|---------------|---------------|------------------|
| Case No. | Age (yr) | Sex | Diagnosis | Level | F. U c | lassification | Resected Area |
| 1 | 45 | F | Chondrosarcoma | T2,3 | | 6 | T(1),2,3,4,(5) |
| 2 | 55 | F | MET (breast) | Т6 | | 2 | T(5),6,(7) |
| 3 | 18 | М | Chondrosarcoma | T5 | 156 M | 5 | T(3),4,5,6 |
| 4 | 40 | F | MET (thyroid cancer) | T12 | | 5 | T(11),12,(L1) |
| 5 | 61 | F | Chondrosarcoma | Т6 | | 6 | T4,5,6,7,8,(9 |
| 6 | 22 | М | Osteosarcoma | L1 | | 1 | (T12),L1,(2) |
| 7 | 41 | M | Chordoma | L2 | | 6 | L1,2,3 |
| 8 | 39 | М | GCT | T7 | | 6 | T(6),7,(8) |
| 9 | 53 | М | MET (RRC) | T10,11 | | 6 | T(8),9,10,11 |
| 10 | 45 | F | MET (laryngeal cancer) | T7 | | 3 | T(6),7,(8) |
| 11 | 31 | F | GCT | T12 | | 4 | T11,12,L1 |
| 12 | 54 | М | MET (RRC) | Т6 | | 3 | T(5),6,(7) |
| 13 | 38 | F | GCT | L3 | | 6 | L(2),3,(4) |
| | | | | | | | |

Table 1. Details of the 13 Cases Who Underwent Margin-Free Spondylectomy

MET (breast) indicates metastatic spine tumor of breast cancer; MET (thyroid cancer), metastatic spine tumor of thyroid cancer; GCT, giant-cell tumor; MET (RC), metastatic spine tumor of renal cell cancer; MET (laryngeal cancer), metastatic spine tumor of laryngeal cancer; Level, vertebral level of tumor origin; Classification, surgical classification by Tomita *et a*^β; Resected area, extirpated vertebral level. The value in parenthesis indicates the osteotomized normal vertebral level.

Margin-Free Spondylectomy for Extended Malignant Spine Tumors: Surgical Technique and Outcome of 13 Cases Hasegawa, Kazuhiro ; Homma, Takao ; Hirano, Toru ; Ogose, Akira ; Hotta, Tetsuo ; Yajiri, Yoichi; Nagano, Junji ; Inoue, Yoshiya Spine Volume 32(1), 1 January 2007, pp 142-148 P Krepler : 2 Ewing Sarcoma, 1 Chondrosarcoma

F.U. 27 M – 58 M

No evidence of disease

2 Neurological deficit

| Case | Gender | Age (yrs) | Histology | Site | Neurology preoperatively | Chemotherapy | Follow-up (mths) | Surgical margins | Oncological state* |
|------|--------|-----------|----------------------|--------|-----------------------------|--------------|---------------------|---------------------|-----------------------|
| 1 | М | 45.8 | Leiomyosarcoma | T7 | No | No | 24 | Marginal | DOD |
| 2 | M | 28.1 | Osteogenic sarcoma | T12 | No | Yes | 67 | Wide | NED |
| 3 | M | 36.9 | Spindle-cell sarcoma | T6,7,8 | No | Yes | 46 | Wide | AWD |
| 4 | F | 16.2 | Chondrosarcoma | T2,3 | Paraparesis T3 | Yes | 58 | Wide | NED |
| 5 | F | 6.2 | Ewing's sarcoma | L3 | Weakness L3 | Yes | 46 | Wide | NED |
| 6 | F | 34.2 | Schwannoma | T10 | Paraparesis T10 | Yes | 12 | Marginal | DOD |
| 7 | Μ | 18.3 | Ewing's sarcoma | L3 | No | Yes | 27 | Wide | NED |

* DOD, died of disease; NED, no evidence of disease; AWD, alive with disease

Total vertebrectomy for primary malignant tumours of the spine Krepler, P.; Windhager, R.; Bretschneider, W.; Toma, C. D.; Kotz, R. JBJS Volume 84-B(5), July 2002, pp 712-715 Shital Parikh :

2 Osteosarcomas (1recurrence) 1 Chondrosarcoma 1 Paraganglioma

1 Ewing tumor (died) 1 Chordoma 1 Hodgkin Lymphoma (died)

Table 1. Patient Characteristics and Location of Malignant Tumors

| | | c. | | A | Delas is | | | | | | | |
|------------------|----|----|------|---------------------|-----------------------|---|----|---|--------------|---|-----------|----------------|
| Histological | | | 3X | Age at Diagnosis | Delay in Diagnosis | | | | | | | |
| Diagnosis | Ν | М | F | (yr) | (mo) | С | CT | Т | TL | L | LS | S |
| Osteosarcoma | 2 | 2 | 1000 | 12.2 | 5 | | | 1 | - <u></u> | | <u></u> s | 1 |
| Rhabdomyosarcoma | 2 | 2 | | 6.7 | 10.5 | | — | 2 | | 1 | _ | _ |
| Ewing sarcoma | 1 | 1 | | 6.8 | 3 | | _ | | 1 | | | _ |
| Chondrosarcoma | 1 | 1 | | 18 | 18 | | | 1 | | - | | |
| Paraganglioma* | 1 | | 1 | 11.1 | 12 | | - | | _ | 1 | — | - |
| Ependymoma* | 1 | 1 | | 15.7 | 1 | - | 1 | 1 | _ | | - | _ |
| Hodakin lymphoma | 1 | | 1 | 18 | 2 | | | | (a <u></u>) | 1 | <u></u> | <u> 17 – 7</u> |
| Chordoma** | 1 | - | 1 | 5.7 | 60 | _ | - | _ | _ | 1 | _ | |
| Total | 10 | 7 | 3 | 11.7 | 13.9 | | 1 | 5 | 1 | 4 | - | 1 |

Intramedullary, intradural.

* Extradural.

3 metastatic: 2 Rhabdomyosarcomas and 1 Ependymoma (died)

Orthopaedic Implications in the Management of Pediatric Vertebral and Spinal Cord Tumors: A Retrospective Review Parikh, Shital N. MD; Crawford, Alvin H. MD Spine Volume 28(20), 15 October 2003, pp 2390-2396

Steven Beer: 2 GCT, 2 Chordomas, 1 Ewing Sarcoma (Long term F.U. 1951-1976)

| Diagnosis | Site | Age (yr) Sex | Treatment | Result | Follow-up (yr |
|---------------------------|--|-----------------|--|--|-----------------------|
| Aneurysmal bone cyst | S1–S3 right sacral ala | 15/M | Radical excision and fusion | Normal | 20 |
| Aneurysmal bone cyst | L3 body and pedicle right | 11/F | Radical excision, no fusion | Normal function, developed scolio- sis requiring immobilization | 22 |
| Eosinophilic granuloma | C7 body | 8/F | Resection and fusion | Normal | 20 |
| Osteoid osteoma | S2 pedicle | 15/M | Partial excision elsewhere | Recurrence | |
| | | | Radical excision/radiation | Normal | 20 |
| Osteoid osteoma | C4 pedicle | 10/M | Radical excision | Normal | 23 |
| Osteoid osteoma | L5-S1 pedicle | 8/M | Partial excision | Persistent pain, residual tumor | |
| | | | Excision | Normal | 15 |
| Osteoblastoma | C2 anterior arch | 9/M | Transoral excision and fusion | Recurred 6 mo | |
| | | | Immobilization/radiation | Normal | 26 |
| Osteoblastoma | L5 and S1 lamina | 8/F | Radical excision | Normal | 16 |
| Osteoblastoma | L3-L5 posterior arches | 6/M | Partial excision | Recurred 1 yr | |
| | | | Radical excision and radiation | Normal | 18 |
| Osteoblastoma | C7–T2 pedicles and transverse processes | 13/F | Excision with anterior and posterior fusion | Recurred 6 mo | |
| | | | Radical excision and radiation | Normal | 32 |
| Osteochondroma | T3-T4 pedicle and lam- ina | 16/M | Excision | Normal | 11 |
| Osteochondroma | T4–T5 pedicle | 10/F | Excision | Normal | 25 |
| Osteochondroma | T3 body | 15/M | Partial excision | Normal | 10 |
| Hemangioma | L2-L3 bodies | 14/M | Partial excision and radiation | Normal | 30 |
| Hemandioma | T5 lamina and body | 14/M | Partial excision | Partial recovery | 44 |
| Giant cell tumor | T5–T7 bodies | 13/M | Excision with fusion | Normal | 19 |
| Giant cell tumor | T2-T3 body and pedicle | 14/F | Excision | Partial recovery | 30 |
| Chordoma | Sacroiliac region | 3/F | Radical resection and radiation | Died 1 vr | |
| Chordoma | L5-S1 body | 2/M | Radical excision | Normal | 36 |
| Ewing's sarcoma | T1-T3 body | 16/F | Partial excision, radiation, and chemotherapy | No recovery | No recurrenc 15 yr |

Primary Tumors of the Spine in Children: Natural History, Management, and Long-term Follow-up Beer, Steven J. MD; Menezes, Arnold H. MD

Spine Volume 22(6), 15 March 1997, pp 649-658

Katsuro Tomita: 1 Osteosarcoma, 1 Chondrosarcoma

F.U. 2 Y and 3.1 Y

Disease Free

| | | | Chemotherapy | | Radiotherapy | | Frankel's Grade | | | |
|-------------|--------------|-----------------------------------|--------------|--------|--------------|--------|-----------------|--------|-------------------|--------------------------------|
| Patient No. | Age (yr)/Sex | Histology | Preop | Postop | Preop | Postop | Preop | Postop | Follow-up (yr) | Physical State at Follow-up |
| 1 | 19/F | Osteosarcoma | No | Yes | No | No | В | E | 6.5 | CDF |
| 2 | 7/M | Chondrosarcoma (recurrence) | No | No | No | No | C | D | 3.1 | NED |
| 3 | 17/M | Osteosarcoma | Yes | Yes | No | No | E | E | 2.0 | CDF |
| 4 | 42/F | Giant cell tumor (recurrence) | No | No | Yes | No | E | E | 2.0 | CDF |
| 5 | 60/F | Malignant fibrous histiocytoma | No | No | No | No | C | D | 0.6 | DOD |
| 6 | 73/M | Solitary plasmacytoma | No | No | No | No | С | D | 3.5 | CDF |
| 7 | 36/M | Giant cell tumor | No | No | No | No | E | F | 2.0 | CDF |

CDF = continuously disease-free; NED = no evidence of disease; DOD = dead of disease.

Total En Bloc Spondylectomy: A New Surgical Technique for Primary Malignant Vertebral Tumors Tomita, Katsuro MD*; Kawahara, Norio MD*; Baba, Hisatoshi MD†; Tsuchiya, Hiroyuki MD*; Fujita, Takuya MD*; Toribatake, Yasumitsu M Spine Volume 22(3), 1 February 1997, pp 324-333 Marco Rex : 13 Ewing Sarcoma 19 Y of age (7-26)

F.U. 65 M (2-218)

Anterior column alone was involved in 4 patients

Posterior column alone was involved in 1 patient

Both columns were involved in the remaining 8 patients



Ewing's Sarcoma of the Mobile Spine Marco, Rex A. W. MD; Gentry, J Brett MD†; Rhines, Laurence D. MD‡; Lewis, Valerae O. MD¶; Wolinski, J P. MD§; Jaffe, Norman MD†; Gokaslan, Ziya L. MD§ Spine Volume 30(7), 1 April 2005, pp 769-773* Ewing and PNET tumors
Chondrosarcoma
Osteosarcoma
Chordoma
Rhabdomyosarcoma





CLINICAL ONSET

Pain : backache or radicular

Exacerbated when the patient was supine

Interval between the onset of symptoms and diagnosis ranged from 1 week to more than one year

26 weeks for benign tumors

20 weeks overall

11 weeks for malignant tumors



Neurological deficit such as myelopathy to plexopathy and peripheral neuropathy in

91% of the malignant spine tumors

64% of the benign spine tumors

BIOPSY AND SURGICAL STAGING

Tools for diagnosis and surgical staging



Computed tomography scanning MRI Bone scintigraphy Angiography TEP scan



SURGICAL STAGING

Katsuro Tomita

Classifications of different anatomic sites of the vertebra.

- 1 =vertebral body.
- 2 = pedicle.
- 3 = lamina, transverse, and spinous processes.
- 4 = spinal canal (epidural space).
- 5 = paravertebral area



Total En Bloc Spondylectomy: A New Surgical Technique for Primary Malignant Vertebral Tumors Tomita, Katsuro MD*; Kawahara, Norio MD*; Baba, Hisatoshi MD†; Tsuchiya, Hiroyuki MD*; Fujita, Takuya MD*; Toribatake, Yasumitsu MD* Spine Volume 22(3), 1 February 1997, pp 324-333

SURGICAL STAGING



Total En Bloc Spendylectomy: A New Surgical Technique for Primary Malignant Vertebral Tumors Tomita, Katsuro MD*; Kawahara, Norio MD*; Baba, Hisatoshi MD†; Tsuchiya, Hiroyuki MD*; Fujita, Takuya MD*; Toribatake, Yasumitsu MD* Spine Volume 22(3), 1 February 1997, pp 324-333

SURGICAL STAGING

| Туре | Stage | Characteristics | | |
|-----------|---------------------|--|--|--|
| Malignant | I IA IB | Low grade without metastases Intracompartmental Extracompartmental | | |
| | II IIA IIB | High grade without metastases Intracompartmental Extracompartmental | | |
| | III IIIA IIIB | Low/high grade with metastases Intracompartmental Extracompartmental | | |

Enneking WF: A system of staging musculoskeletal neoplasms. Clin Orthop 1986, 204:9–24



| | - Stage IA | Stage IB | Stage IIA | Stage IIB |
|-----------------------|--|--|---|--|
| Grade | G1 | G1 | G2 | G2 |
| Site | T1 | T2 | T1 | T2 |
| Metastasis | MO | MO | MO | MO |
| Clinical course | Symptomatic indolent growth | Symptomatic mass, indolent growth | Symptomatic rapid growth | Symptomatic rapid growth, fixed mass, pathologic fracture |
| lsotope scan | Increased uptake | Increased uptake | Increased uptake, beyond radiographic limits | Increased uptake, beyond radiographic limits |
| Radiographic grade | II | II | | III ^o |
| Angiogram | Modest neovascular reaction, involvement of neurovascular bundle | Modest neovascular reaction, involvement of neurovascular bundle | Marked neovascular reaction, no involvement of neurovascular bundle | Marked neovascular reaction, involvement of neurovascular bundle |
| CT scan | Irregular or broached capsule- intracompartmental | Extracompartmental extension or location | Broached (pseudo) capsule– intracompartmental | Broached (pseudo) capsule- extracompartmental |

Staging and treatment of primary tumors of the spine. Boriani S, Bandiera S, Biagini R, Picci P. Current Opinion in Orthopedics. 10(2):93-100, April 1999

SURGICAL STAGING WBB (Weinstein, Boriani, Biagini)



12 radiating zones (numbered 1 to 12 in a clockwise order)

5 concentric layers (A to E, from the paravertebral extraosseous compartments to the dural involvement)

The longitudinal extent of the tumor is recorded according to the levels involved. *Primary Bone Tumors of the Spine: Terminology and Surgical Staging. Boriani and al. Spine 1997; 22(9); 1036-1044*

B

Posterior resection



Recurrent chondrosarcoma of T8

Primary Bone Tumors of the Spine: Terminology and Surgical Staging. Boriani and al. Spine 1997; 22(9); 1036-1044

Anterior resection



L 4 metastatic osteosarcoma

Primary Bone Tumors of the Spine: Terminology and Surgical Staging. Boriani and al. Spine 1997; 22(9); 1036-1044



Total vertebrectomy for primary malignant tumours of the spineKrepler, P.; Windhager, R.; Bretschneider, W.; Toma, C. D.; Kotz, R.JBJS Volume 84-B(5), July 2002, pp 712-715

SURGERY 9. 1995

Prone position Instrumentation L2 L4 Intralesional resection



Liposarcoma arising from the foramen of T7

Primary Bone Tumors of the Spine: Terminology and Surgical Staging. Boriani and al. Spine 1997; 22(9); 1036-1044



The sagittal resection can be performed the patient in a prone position trough a single midline posterior skin incision

Osteosarcoma (bone metastasis of a femoral osteosarcoma) Adjuvant chemotherapy

Sagittal resection









DURA MATTER





LUNG

SPINE









Died at two years F.U. from lung metastasis No evidence of local recurrence

3/2001

MRI

CT SCAN





Telangiectasic osteosarcoma











The sagittal resection can be performed the patient in a prone position trough a single midline posterior skin incision



Hasegawa Kazuhiro and al Spine 2007 32 (1)



SURGERY

- 4 hemi corporectomies
- Non vascularized fibula anteriorly
- Hardware and fusion posteriorly from T1 to T9
- Post operative complications :
- Fibula displacement..... reoperated on
- Chylothorax
- Margins: free of disease
- 18% of viable cells tumor
- Post operative chemotherapy course
- F.U. 6 years disease free





Prone position Posterior single approach 3 Ribs 4 hemispondylectomies Fibula anteriorly

Hardware and fusion posteriorly



COMPLICATION





Right lower limb monoparesia post operatively Complete recovery at two years F.U. Surgical decompression

Laminectomy
Intratumoral resection
<u>Post-op:</u>
1/ regression of the paraplegia
2/ histology : PNET (Ewing)





Intercostal nerve roots on the tumor side to be cut





Complications

bleedingLeft chest dysesthesia4 Years F.U. disease free

Surgery is very demanding

Surgery needs surgeons expert in both spinal and tumor surgery

Surgery leeds to complications

| Chondrosarcoma92Osteosarcoma76Ewing's sarcoma75Liposarcoma10Hemangioendothelial sarcoma11Giant cell tumor10Malignant fibrous histiocytoma21Epithelial sarcoma11Myofibroblastic sarcoma10 | Local currences |
|--|--------------------|
| Osteosarcoma76Ewing's sarcoma75Liposarcoma10Hemangioendothelial sarcoma11Giant cell tumor10Malignant fibrous histiocytoma21Epithelial sarcoma11Myofibroblastic sarcoma10 | 3 |
| Ewing's sarcoma75Liposarcoma10Hemangioendothelial sarcoma11Giant cell tumor10Malignant fibrous histiocytoma21Epithelial sarcoma11Myofibroblastic sarcoma10 | 5 |
| Liposarcoma10Hemangioendothelial sarcoma11Giant cell tumor10Malignant fibrous histiocytoma21Epithelial sarcoma11Myofibroblastic sarcoma10 | 1 |
| Hemangioendothelial sarcoma11Giant cell tumor10Malignant fibrous histiocytoma21Epithelial sarcoma11Myofibroblastic sarcoma10 | 0 |
| Giant cell tumor10Malignant fibrous histiocytoma21Epithelial sarcoma11Myofibroblastic sarcoma10 | 0 |
| Malignant fibrous histiocytoma21Epithelial sarcoma11Myofibroblastic sarcoma10 | 0 |
| Epithelial sarcoma11Myofibroblastic sarcoma10 | 1 |
| Myofibroblastic sarcoma 1 0 | 1 |
| | 1 |
| 30 16 | 12 🗖 |

Relationship Between Surgical Margins and Local Recurrence in Sarcomas of the Spine Talac, Robert MD, PhD; Yaszemski, Michael J. MD, PhD; Currier, Bradford L. MD; Fuchs, Bruno MD; Dekutoski, Mark B. MD; Kim, Choll W. MD, PhD; Sim, Franklin H. MD, Clin Ortop Rel Res Volume 397, April 2002, pp 127-132

PROGNOSIS

82 patients with primary neoplasm of the spine

0% Patients undergoing curettage

Five year survival $\leftarrow 18.7\%$ Patients with incomplete resection

75% Patients with complete resection

Primary tumor of the spine. Weinstein JN, McLain RF. Spine 1987; 12: 843-51

2 of 22 (9%) patients with Ewing Sarcoma involving the trunk survived 5 years after treatment with radiation therapy alone

The curability of Ewing's endothelioma of bone in children

Phillips RF, Higinbotham NL. J Pediatr 1967; 70: 391 - 7

5 years survival rate < 19 % in patients with Ewing Sarcoma involving the trunk after treatment with radiation therapy alone

Ewing's sarcoma of bone. Experience with 140 patients

Wilkins RM, Pritchard DJ, Burger EO Jr and al Cancer (Phila) 1986; 58: 2551 – 5.

Radiotherapy + Chemotherapy in Ewing sarcoma

3 patients developed a local recurence
5 / 13 patients disease free at last F.U
Disease-free survival rate 46% at 5 years

34% at 10 years

Ewing's Sarcoma of the Mobile Spine

Marco, Rex A. W. MD*; Gentry, J Brett MD†; Rhines, Laurence D. MD‡; Lewis, Valerae O. MD¶; Wolinski, J P. MD§; Jaffe, Norman MD†; Gokaslan, Ziya L. MD§ Spine Volume 30(7), 1 April 2005, pp 769-773

GOLD STANDARD FOR THE MANAGEMENT OF MALIGNANT SPINAL TUMOR IN CHILDREN Biopsy ideally per cutaneously CT guided Neo adjuvant chemotherapy MRI + CT scan to know exactly the limits of the tumor Spinal arteriography for Adamkievitch +/- embolization Surgery if cord compression En bloc resection as often as possible Stabilization and fusion (hardware allowing control MRI) Post operative chemotherapy according to Huvos grading

As far as a en bloc resection can be performed the prognosis is now the same that the one of the limbs malignant tumors 2 Telangiectasic Osteosarcomas; 21 and 57 Years old paraplegic patients Segmental resection of the spine including the spinal cord Both patients died 6 months after surgery

Complete Segmental Resection of the Spine, Including the Spinal Cord, for Telangiectatic Osteosarcoma: A Report of 2 Cases Murakami, Hideki MD*; Tomita, Katsuro MD*; Kawahara, Norio MD*; Oda, Makoto MD†; Yahata, Tetsutaro MD*; Yamaguchi, Takehiko MD‡ Spine Volume 31(4), 15 February 2006, pp E117-E122

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