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Spine Update

Primary Bone Tumors of the Spine

Terminology and Surgical Staging

Stefano Boriani, MD,* James Neal Weinstein, DO, MS,† and Roberto Biagini, MD‡

Appropriate application of an oncologic staging system is required to evaluate the relationship among histologic types, management, and outcome of primary bone tumors. A commonly accepted terminology for surgical procedures and for definition of tumor extent is needed for surgical planning and clinical reviews. The principles of the Enneking system for classifying stages of tumors are emphasized and applied to the spine using a practical approach for surgical staging. [Key words: spine tumor, staging, terminology] **Spine 1997; 22:1036–1044**

Evaluating the management and outcome of primary spine tumors has been limited by a variable and loose interpretation of terminology and staging. Moreover, there are difficulties in applying the oncologic staging system for long bones proposed by Enneking to the spine.[8–10] The result has been a disorganized approach, with management more often determined by the available surgical skill than by the histologic type of the tumor and its location.[18] This has lead to an unsatisfactory assessment of the various types of management, which often are used with no consideration of the accepted principles of management in oncology.

As a result of the contributions of pioneers in this field, the techniques of en bloc surgery in the spine are well known, particularly for the thoracolumbar area.[12,13,15–17,19–22,24,25] These procedures began to evolve in 1968, but few were based on established principles for treatment in oncology.[15,20,21] What failed to evolve was the application of a common terminology and a staging system, especially one dedicated to the planning of the surgical management of spine tumors. In fact, even in the most recent reports, terms such as "resection," "excision," "radical," and "vertebrectomy" are used with different meanings.[11,18,25] An exacting terminology is an essential requirement for correctly studying the relationships among tumor types, management, and outcome.

The aim of this report is to apply to the spine the terms accepted by most oncologists as applicable for musculoskeletal tumors of the limbs.[6,8–10] An appropriate terminology and staging system dedicated to spine tumors should encourage more consistent surgical planning and improve the ability to exchange reliable information between institutions.

■ Terminology[9]

"Curettage" describes the piecemeal removal of the tumor. As such, it is always an intralesional procedure.

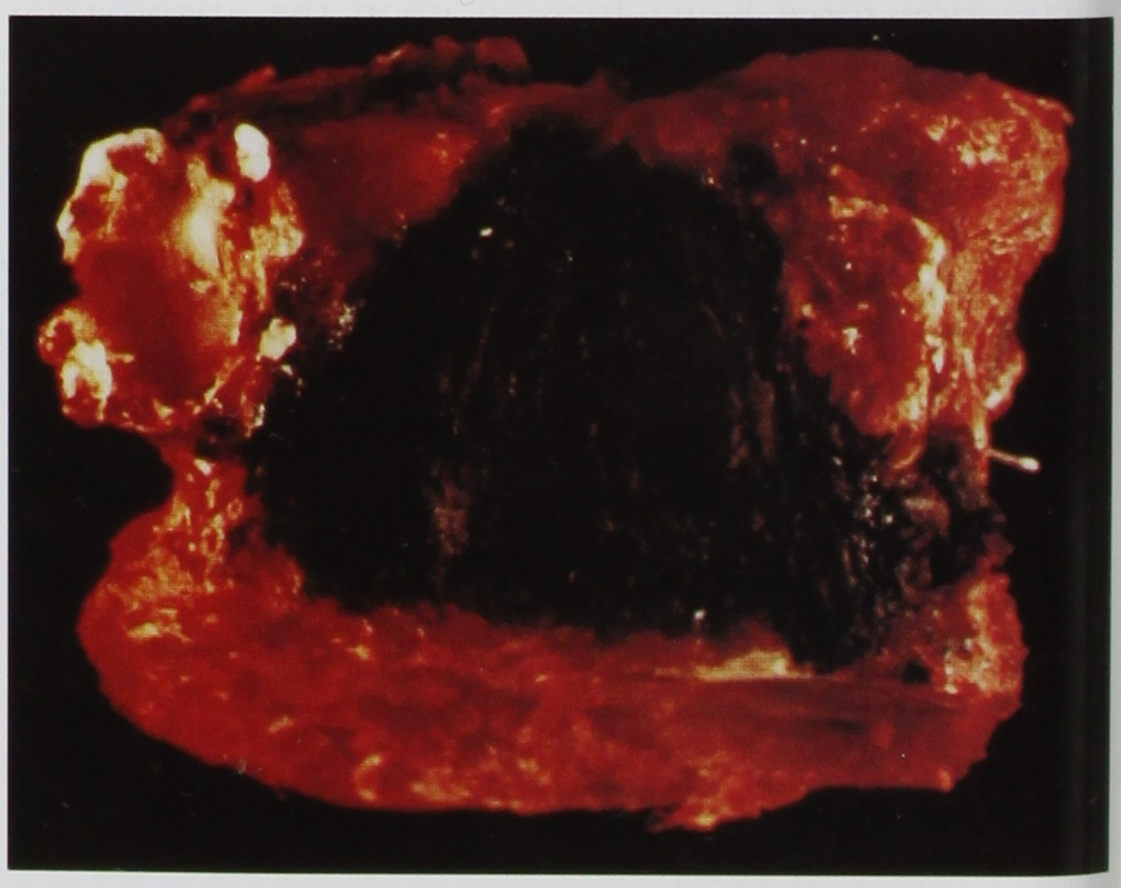

Figure 1. Chordoma of L3 submitted to en bloc resection of the vertebral body. Painting the surface of the surgical margin with China ink allows the depth of healthy tissue (if any) to be evaluated to define the margin as intralesional, marginal, or wide. In this case the biopsy had been performed incorrectly through the epidural space. The histologic study demonstrated that the margin achieved was "wide" all around the vertebra, but marginal or intralesional in the epidural space. The final definition was wide, contaminated. No radiotherapy was used, and there was no recurrence of the tumor at the lastest evaluation (5 years later).

From the *Department of Orthopaedics and Traumatology, Ospedale Maggiore, Bologna, Italy, †Dartmouth Medical School, Hanover, New Hampshire, and ‡Rizzoli Institute, Clinica Ortopedica dell'Università di Bologna, Bologna, Italy.
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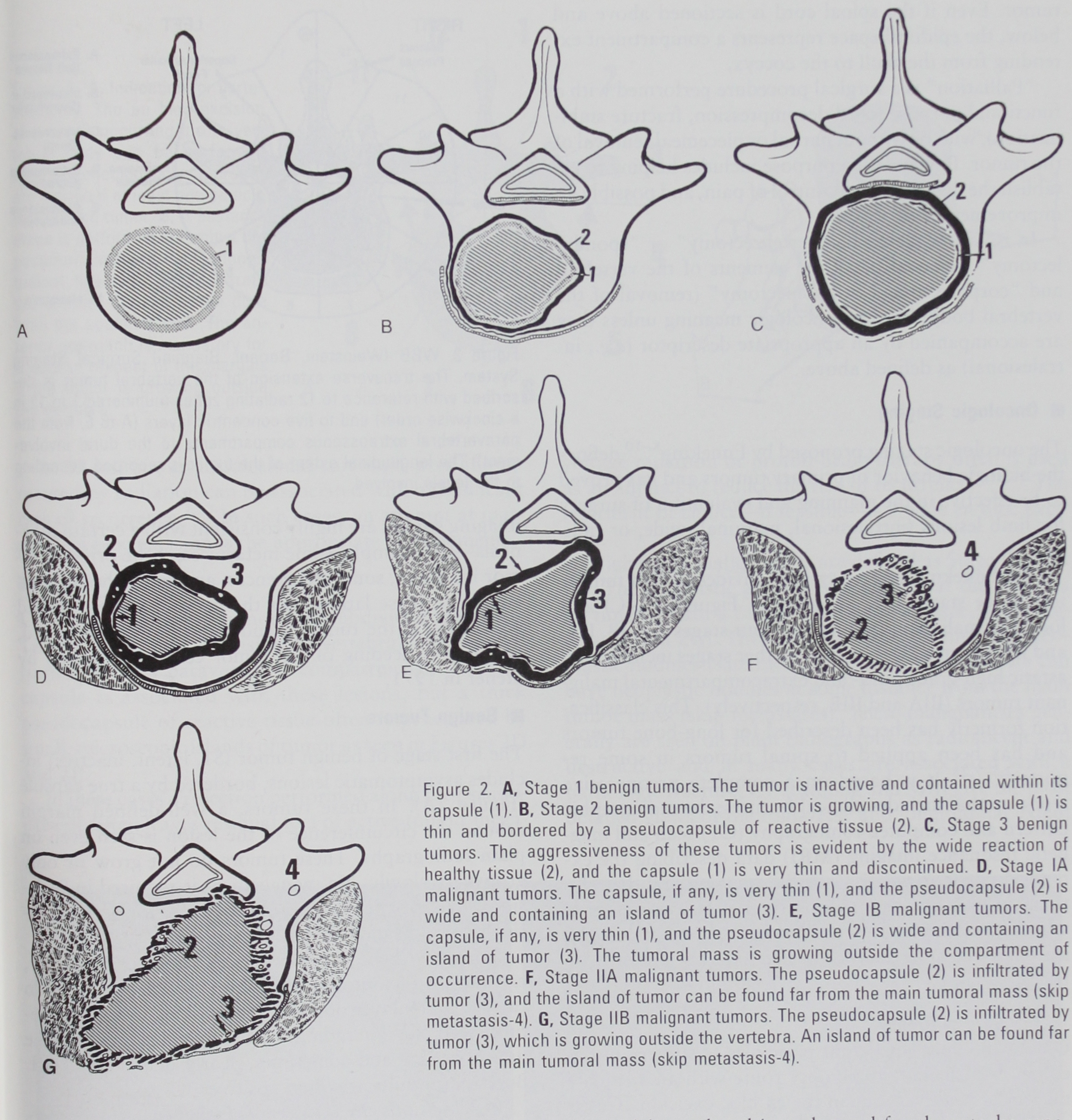

Figure 2. **A,** Stage 1 benign tumors. The tumor is inactive and contained within its capsule (1). **B,** Stage 2 benign tumors. The tumor is growing, and the capsule (1) is thin and bordered by a pseudocapsule of reactive tissue (2). **C,** Stage 3 benign tumors. The aggressiveness of these tumors is evident by the wide reaction of healthy tissue (2), and the capsule (1) is very thin and discontinued. **D,** Stage IA malignant tumors. The capsule, if any, is very thin (1), and the pseudocapsule (2) is wide and containing an island of tumor (3). **E,** Stage IB malignant tumors. The capsule, if any, is very thin (1), and the pseudocapsule (2) is wide and containing an island of tumor (3). The tumoral mass is growing outside the compartment of occurrence. **F,** Stage IIA malignant tumors. The pseudocapsule (2) is infiltrated by tumor (3), and the island of tumor can be found far from the main tumoral mass (skip metastasis-4). **G,** Stage IIB malignant tumors. The pseudocapsule (2) is infiltrated by tumor (3), which is growing outside the vertebra. An island of tumor can be found far from the main tumoral mass (skip metastasis-4).

"En bloc" indicates an attempt to remove the whole tumor in one piece, together with a layer of healthy tissue (Figure 1). The specimen then must be submitted to careful gross and histologic studies to further define the procedure as "intralesional," "marginal," or "wide." The term "intralesional" is appropriate if the surgeon has cut within the tumor mass; "marginal" is appropriate if the surgeon has dissected along the pseudocapsule, the layer of reactive tissue around the tumor; and "wide" is appropriate if separation has occurred outside the pseudo capsule, removing the tumor with a continuous shell of healthy tissue.[8,9] This wide en bloc procedure can be called "excision" or "resection." Both of these terms are too widely used and interchanged for them to be separated. However, the authors of the present report prefer to define resection as "en bloc excision." To avoid confusion and to compare results, it is essential to distinguish the longer, more difficult, and risky removal of the whole tumor in one piece (en bloc) from a simple intralesional procedure, even though this sometimes may mean the piecemeal removal of the whole vertebra.

"Radical resection" means the en bloc removal of the tumor and the whole compartment of tumor origin. It is obvious that this can be possible for a tumor arising in the scapula (scapulectomy) or in the tibia (above knee amputation), but it is absolutely impossible for a spine

tumor. Even if the spinal cord is sectioned above and below, the epidural space represents a compartment extending from the skull to the coccyx.

"Palliation" is a surgical procedure performed with a functional purpose (cord decompression, fracture stabilization), with or without partial or piecemeal removal of the tumor. In general its purpose includes helping to establish the diagnosis, the control of pain, and possibly an improvement in function.

In isolation the terms "vertebrectomy" or "spondylectomy" (removal of all the elements of the vertebra) and "corporectomy" or "somectomy" (removal of the vertebral body) have no oncologic meaning unless they are accompanied by an appropriate descriptor (*e.g.*, intralesional) as defined above.

■ Oncologic Staging

The oncologic staging proposed by Enneking[8–10] defines the biologic behavior of primary tumors and has proved to be effective in the planning and evaluation of surgery for limb lesions (intralesional, marginal, wide, or radical).

The Enneking staging system[9] divides benign tumors into three stages (S1, S2, and S3; Figures 2A–C) and localized malignant tumors into four stages (IA, IB, IIA, and IIB; Figures 2D–G). Two further stages include metastatic high-grade intra- and extracompartmental malignant tumors (IIIA and IIIB, respectively). This classification formerly has been described for long-bone tumors and has been applied to spinal tumors in some reports.[2,5–7,28] It is based on a complete preoperative work-up that includes clinical features; the radiographic pattern and computed tomography (CT) scan and magnetic resonance imaging (MRI) data describing the extension of the tumor; its peculiar imaging and the relationship with the neighboring tissues; an isotope scan, which gives information about local aggression and the systemic diffusion; and histologic findings obtained by biopsy.

The Role of Biopsy

One of the most important principles of surgery in oncology is to include the biopsy route with an adequate margin of healthy tissue in the en bloc excision. This is sometimes impossible in the spine if an approach through anatomic planes is used. Unfortunately, a biopsy of a tumor arising in the body or expanding in the epidural space (frequently contained by a continuous pseudocapsule) is sometimes performed by laminectomy (Figures 1A and 1B): this is the worst approach to these tumors because contamination of the epidural space is inevitable, increasing the risk of recurrence. A better solution is the transpedicular approach, filling the empty pedicle with acrylic cement. Trocar biopsy (under CT scan guidance for accurate tissue selection) is the best way to reduce the spread of tumor cells. In some rare cases, biopsy can be avoided, such as when findings on

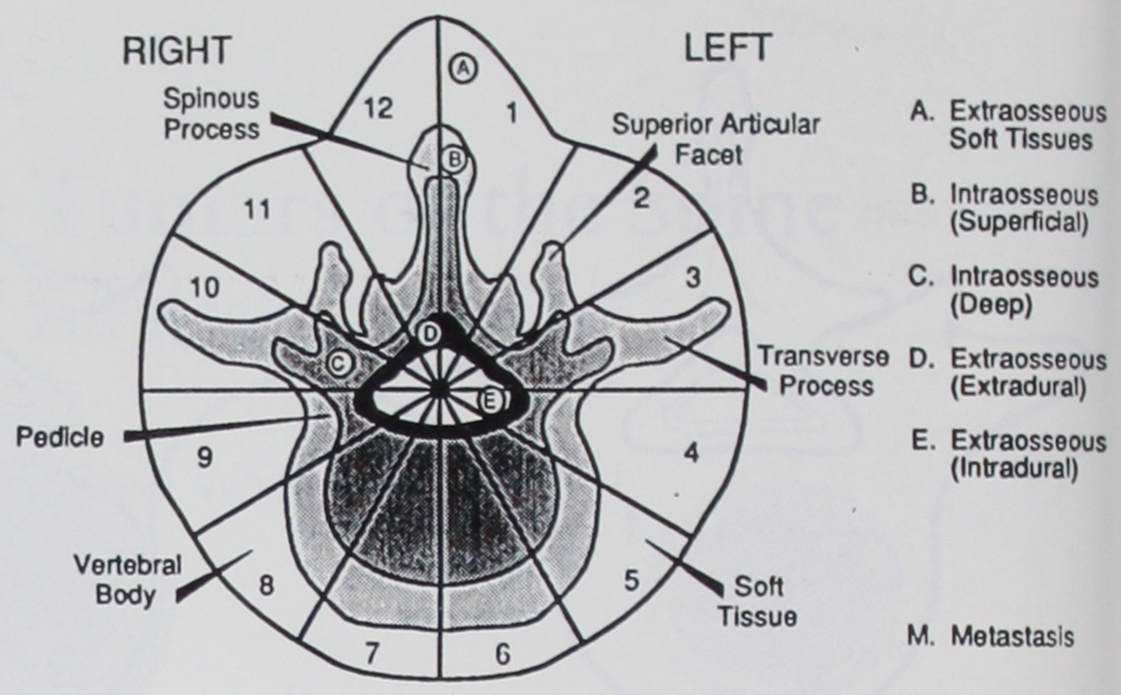

Figure 3. WBB (Weinstein, Boriani, Biagnini) Surgical Staging System. The transverse extension of the vertebral tumor is described with reference to 12 radiating zones (numbered 1 to 12 in a clockwise order) and to five concentric layers (A to E, from the paravertebral extraosseous compartments to the dural involvement). The longitudinal extent of the tumor is recorded according to the levels involved.

imaging studies are highly consistent with a certain diagnosis—for example, some metastases, if the primary tumor is known, some recurrences, and some chondrosarcomas. In these latter cases the myxoid content would spread out if the tumor pseudocapsule were breached, resulting in seeding of the tumor cells as discussed by Stener in 1971.[21]

■ Benign Tumors

The first stage of benign tumor (S1, latent, inactive) includes asymptomatic lesions, bordered by a true capsule (Figure 2A). In these tumors, a well-defined margin around the circumference of the lesion is seen even on plain radiographs. These tumors do not grow or only grow very slowly. No management is required in S1 lesions, unless palliative surgery is needed for decompression or stabilization.

Stage two benign tumors (S2, active) grow slowly, causing mild symptoms. The tumor is bordered by a thin capsule and a layer of reactive tissue (Figure 2B), sometimes seen on plain radiographs as an enlargement of the tumor outline and sometimes clearly defined by MRI. Bone scan results are often positive.

In S2 lesions an intralesional excision can be performed with a low rate of recurrence.[2] The incidence of recurrences can be further lowered by local adjuvants (cryotherapy, embolization, and radiation therapy).

The third stage of benign tumors (S3, aggressive) includes rapidly growing benign tumors: the capsule is very thin, incomplete, or absent (Figure 2C). The tumor invades neighboring compartments and often has an associated wide, reactive, hypervascularized pseudocapsule, which sometimes is permeated by neoplastic digitations. Bone scan results are usually positive, fuzzy limits are seen on plain radiographs, CT scans show the tumor extension, and MRI clearly defines a pseudocapsule and its relationship to the neurologic structures.

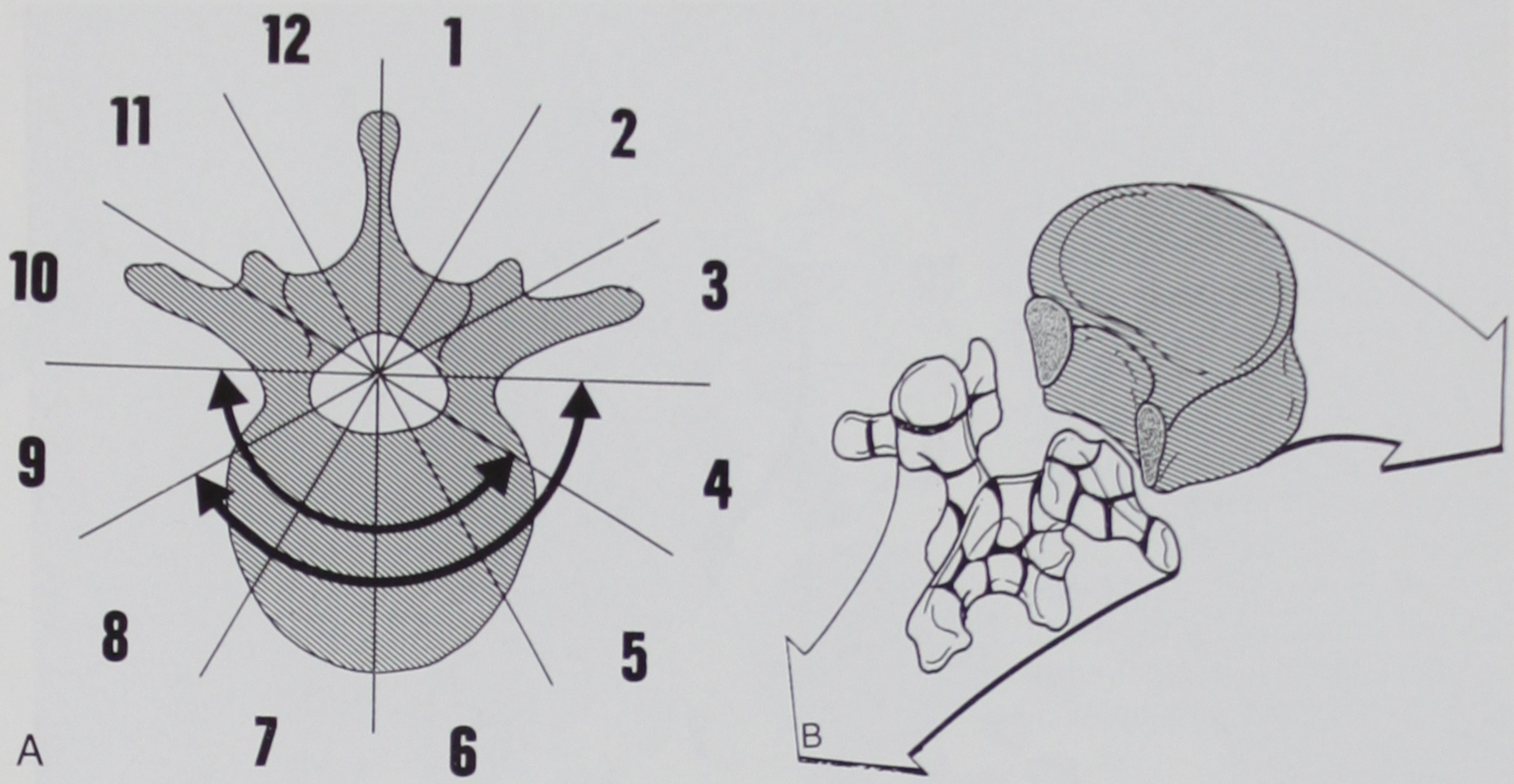

Figure 4. **A,** Indication for vertebrectomy. The en bloc excision of a tumor occurring in the vertebral body can be performed with an oncologically appropriate margin if at least one pedicle is free from tumor. **B,** A posterior stage is performed to remove the posterior elements, cut the longitudinal ligament, and separate the anterior surface of the dura from the posterior wall. The anterior approach is mandatory for a careful respect of the margins if the tumor is growing outside the vertebra.

In S3 lesions, an intralesional curettage, even if augmented by radiation can be associated with a significant rate of recurrence.[2,27] In such cases, an attempt at marginal en bloc excision is the appropriate management.

■ Malignant Tumors

Low grade malignant tumors are subdivided into Stage 1A (the tumor remains inside the vertebra) and Stage 1B (tumor invades paravertebral compartments). No true capsule is associated with these lesions, but a thick pseudocapsule of reactive tissue often is penetrated by small, microscopic islands of tumor as seen in Figures 2D and 2E.

In these cases a resection performed along the pseudocapsule often leaves residual foci of active tumor; megavoltage radiation or proton-beam therapy often is used as an adjunct to reduce the risk of recurrence.[5,19,23] The treatment of choice—if feasible—is a wide en bloc excision.

High grade malignancies are defined as Stage IIA and IIB. The neoplastic growth is so rapid that the host has no time to form a continuous reactive tissue layer (Figures 2F and 2G). There is continuous seeding with neoplastic nodules (satellites). Moreover, these tumors can have neoplastic nodules at some distance from the main tumor mass (skip metastases). These malignancies generally are seen on plain radiographs as radiolucent and destructive and in many cases are associated with a pathologic fracture; CT scanning and MRI give the most detailed views of the transverse and longitudinal extent

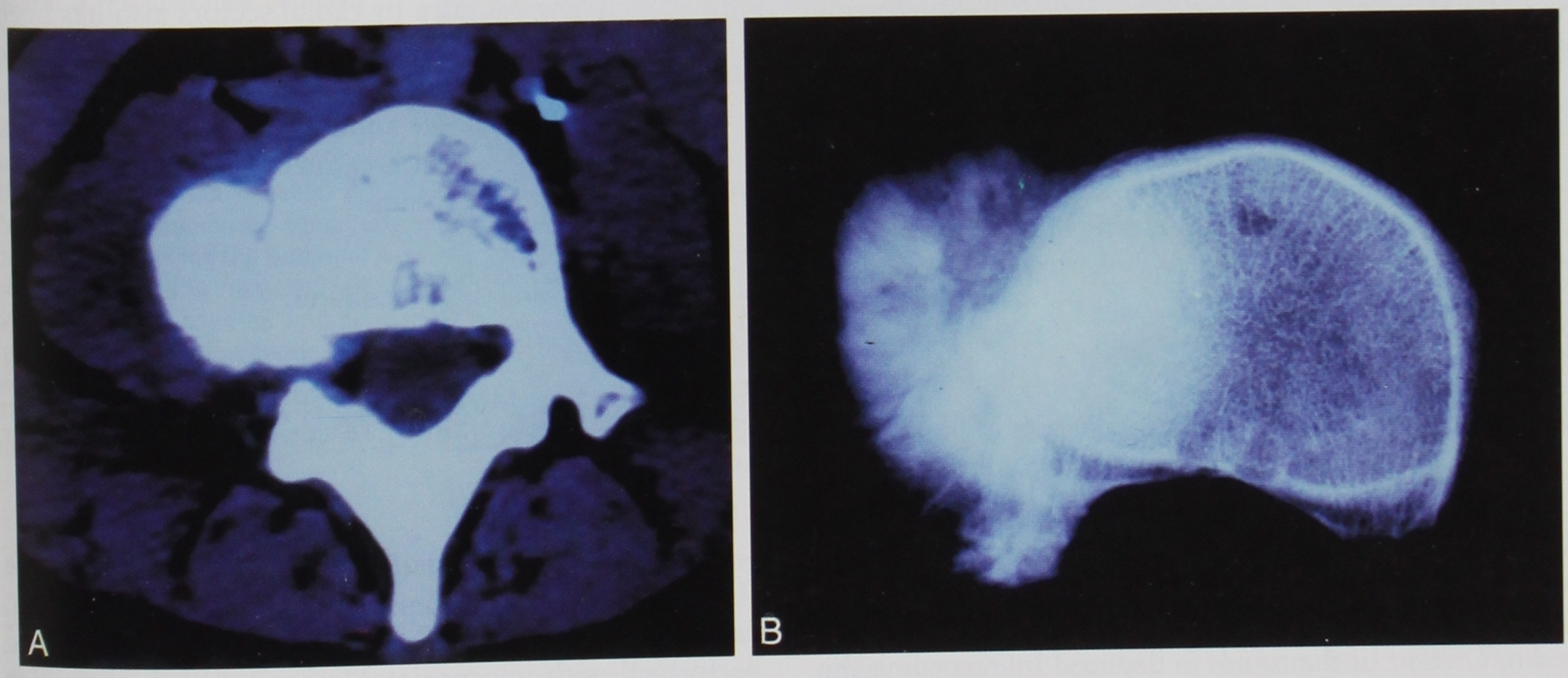

Figure 5. Metastatic osteosarcoma arising from the L4 vertebral body, occupying the left pedicle and expanding into the psoas muscle. The patient had been treated with en bloc excision and neo-adjuvant therapy for proximal humerus osteosarcoma 6 months earlier. No biopsy had been performed on the vertebral tumor. **A,** Enneking staging: stage 11B (high-grade malignant, extracompartmental). WBB staging: zones 4 to 7, layers A to C. **B,** Radiographs of the specimen showing an en bloc resection including the psoas muscle. The histologic study confirmed that a "wide" margin had been achieved. One year later there was further localization in the spine and in the lungs. The patient died 2 years after spine surgery.

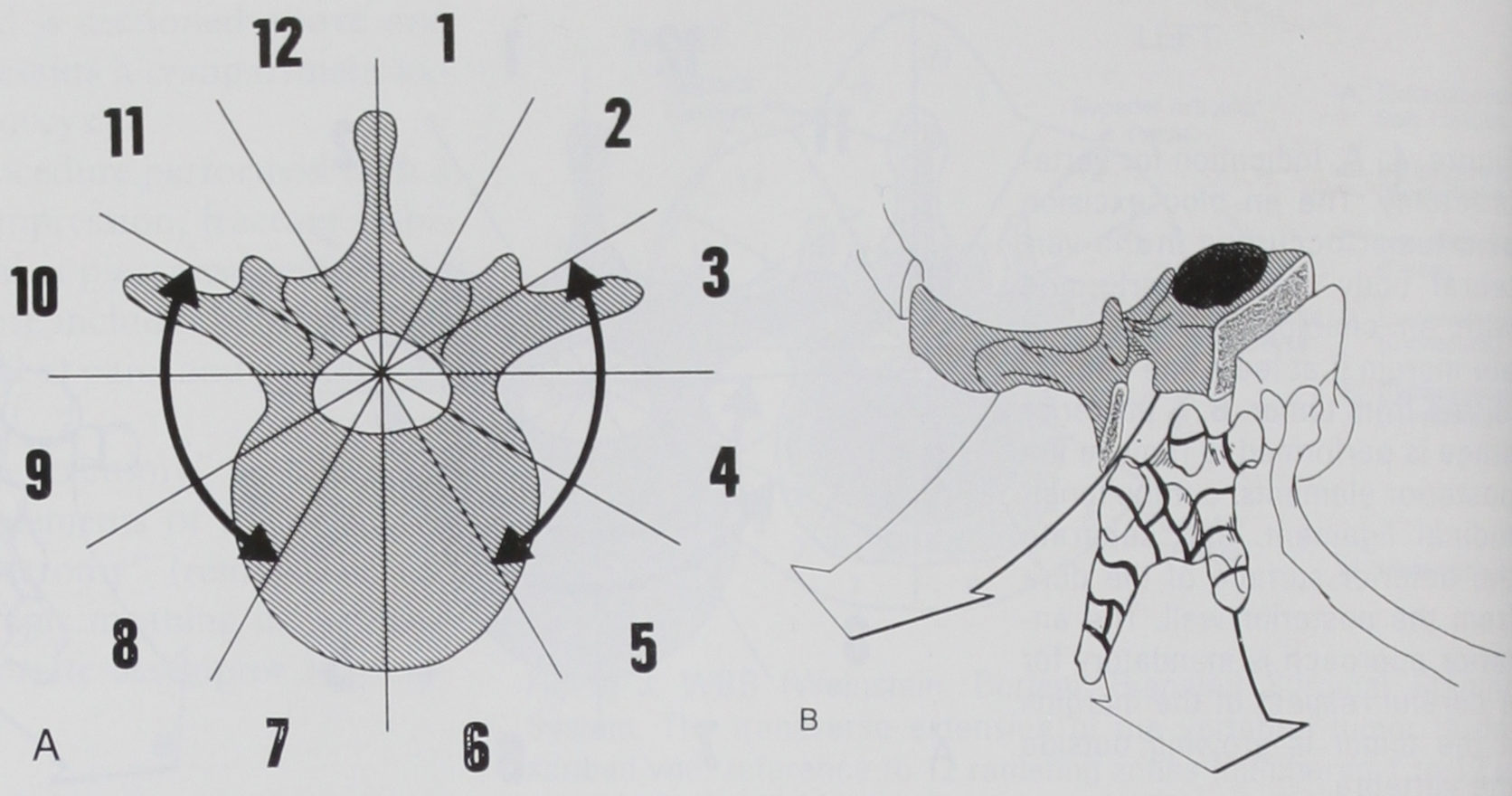

Figure 6. **A,** Indication for sagittal resection. The en bloc excision of a tumor arising eccentrically in the body, the pedicle, or the transverse process, is performed when the tumor occupies the zones 2 to 5 (or 8 to 11). **B,** A posterior stage is needed to remove the posterior healthy elements. A combined posterior and anterior approach is required to safely perform the en bloc excision.

of these tumors and may confirm the absence of a reactive tissue margin. Invasion of the epidural space is rapid in stage B, particularly in small cell tumors (Ewing sarcoma, Lymphomas) and is characterized by infiltrating tumor spread beyond the cortical border of the vertebra with no gross destruction.

The margin of the en bloc excision must be wide at the very least, because it is not possible to achieve a radical margin in the spine. Adjuvant courses of radiation and chemotherapy (according to the tumor type) must be considered for the local control and in an attempt to prevent distant spread.

Stages IIIA and IIIB describe the same lesions as IIA and IIB, but with distant metastasis.

Now having considered surgical resection terminology and oncologic staging, let us look at applying this knowledge to surgical staging for spine tumors.

■ Surgical Staging

Surgical staging is appropriate only after the diagnosis has been established and oncologic staging has been determined. The first attempt to propose a staging classification for purposes of surgically managing primary spine tumors was introduced by Weinstein[26,27] and subsequently was modified according to the Rizzoli Institute (Bologna, Italy) experience to identify each lesion in a systematic fashion.[2,5–7] This WBB staging system, as it is now called, has been subjected to clinical evaluation.[5,28]

Description of Surgical Staging for Spine Tumors

In the transverse plane, the vertebra is divided into 12 radiating zones (numbered 1 to 12 in a clockwise order) and into five layers (A to E, from the paravertebral extraosseous region to the dural involvement). The longitudinal extent of the tumor is deduced by recording the spine segment(s) involved.

Computed tomography scanning, MRI, and sometimes angiography of the tumor are the imaging techniques needed to describe the transverse and longitudinal expansion of these tumors. It is the authors' view that this system (Figure 3) allows a more rational approach to the surgical planning, provided that all efforts are made to perform surgery along the required margins.

This method has evolved from a previously described anteroposterior staging system.[26] The major advantage of the clock-face radiating zone system is that it emphasizes the limitations to performing en bloc excision because of the presence of the spinal cord in the longitudinal median axis of the vertebra. To save this vital structure and to control the epidural space (layer D), the surgeon is compelled to resect wedge sectors of the vertebra. For example, if the tumor occupied an eccentric area, the surgeon is compelled to remove the healthy contralateral parts of the posterior arch, dissect the pseudocapsule from the dura, dislocate the dural sac, and create radiating sections by chisel or osteotome. The approach to the vertebra by radial sectors also has been described recently by Lassale et al,[14] who confirmed the usefulness of this concept.

And finally how do we apply the oncologic and surgical staging systems to the treatment of patients with tumors of the vertebral column?

■ Planning of Surgical Procedures

There are three major methods for performing en bloc excisions in the thoracolumbar spine: "vertebrectomy," "sagittal resection" and "resection of the posterior arch."

Vertebrectomy (Marginal/Wide en bloc Excision of the Vertebral Body)

En bloc tumor excision of the vertebral body can be performed with appropriate "margins" if the tumor is confined to zones 4 to 8 or 5 to 9 (Figure 4A), which means that it is centrally located and that at least one pedicle is free from tumor. The procedure can be performed in two stages (Figure 4B) or in one stage.[3,19,22,25]

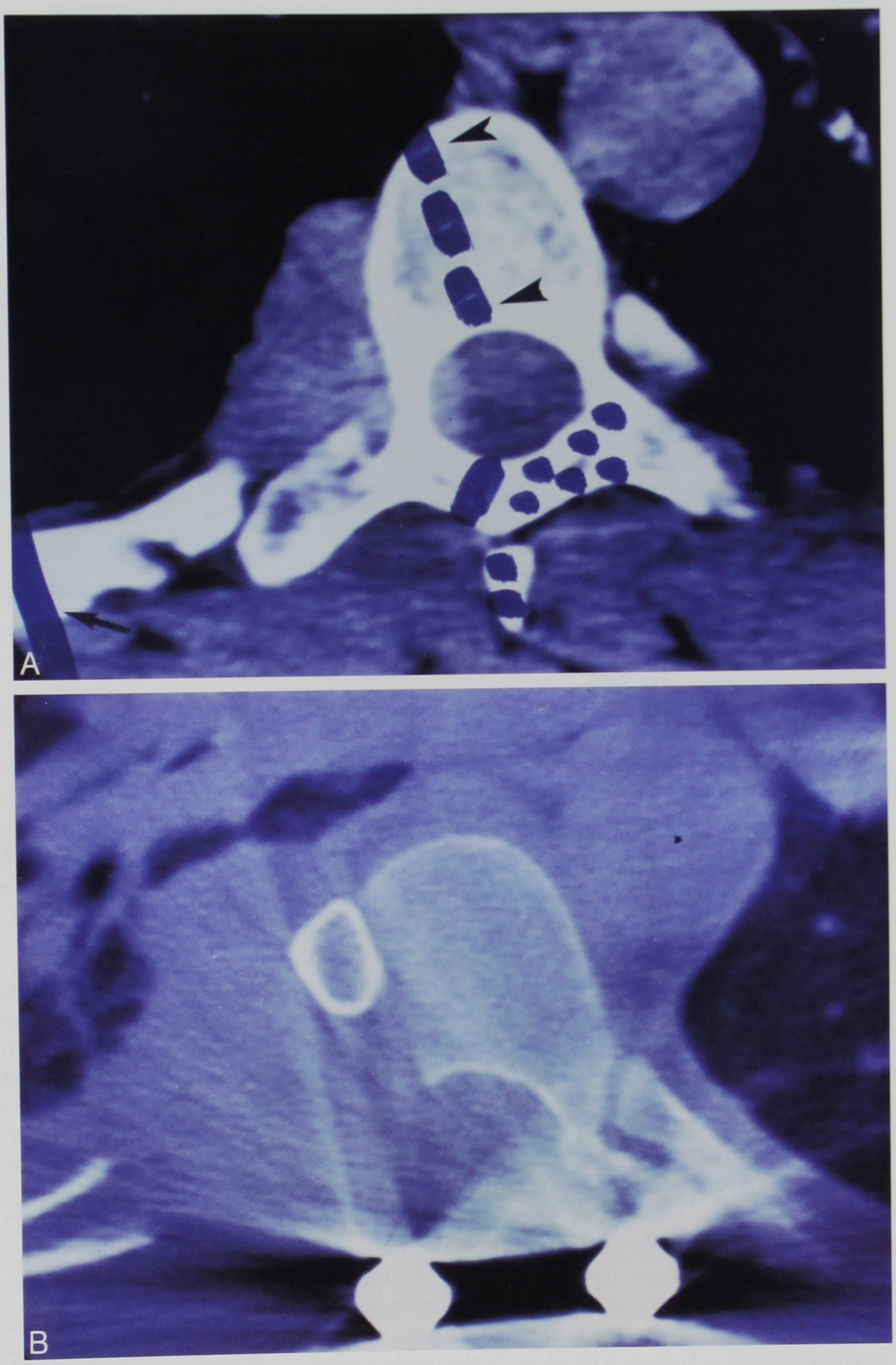

Figure 7. Liposarcoma arising from the foramen of T7, eroding the transverse process, the pedicle, and the body, and expanding into the thorax, covered by the pleura. **A,** Preoperative planning indicated removal of the posterior healthy elements (dotted area) by cutting the ribs (arrow) and the body (arrow head) posterior to anterior after protecting the viscera and the vessels. **B,** Postoperative computed tomography scan showing the outcome of the procedure. A rib was fixed by screw to the superior and inferior vertebra. A posterior device was used for stabilization. There was no recurrence of the tumor 2 years later.

The posterior approach (with patient in the prone position) involves excision of the posterior elements, which enables the anulus fibrosus and the posterior longitudinal ligament to be sectioned. It also allows careful hemostasis of the epidural venous plexus to be achieved and posterior stabilization to be performed.

The anterior approach (transpleural thoracotomy, retroperitoneal abdominal, or thoracoabdominal approach) allows the ligature of segmental vessels (at the lesional level, above and below), proximal and distal discectomies (or the section by chisel through the neighboring vertebrae according to the preoperative planning), the en bloc removal of the vertebral body (Figures 1, 5A, and 5B), and anterior reconstruction.[22] The main advantages of performing the vertebrectomy through a bilateral approach are that ligation of the segmental vessels is made easier and that it permits dissection of the tumor from the anterior elements entirely under direct

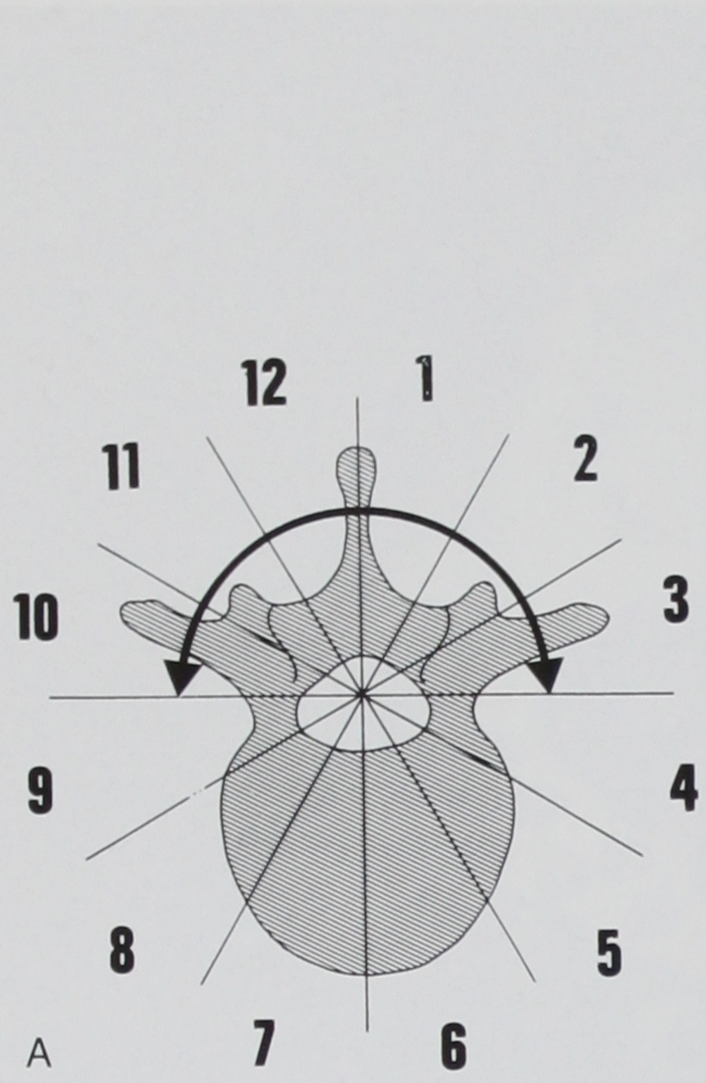

Figure 8. **A,** Indication for resection of the posterior arch. The en bloc excision of a tumor arising in the arch is performed when the tumor occupies the zones 10 to 3. The pedicles must be free from tumor to obtain an oncologically appropriate specimen. **B,** This procedure is performed by posterior approach.

vision, thus achieving a better margin when the tumor has expanded anteriorly.

Sagittal Resection (Marginal/Wide)

This approach is most appropriate when the tumor occupies zones 3 to 5 (or 8 to 10), which means that it arises and develops eccentrically within the body, the pedicle, or the transverse process (Figure 6A). En bloc excision of more than one level can be performed and may include, if necessary, one or more ribs. A combined anterior and posterior approach allows 300 degrees of the circumference of the thoracic and lumbar vertebrae to be viewed (Figure 6B).[1]

The first step is the same as in vertebrectomy. The posterior healthy structures are removed (Figures 6B and 7A), including the pedicle to make room for the dural displacement. The nerve root or roots of the affected segment are ligated, if necessary. Then the patient is placed in a lateral decubitus position. In the thoracic spine, the midline posterior incision is combined with an oblique thoracotomy incision on the rib of the affected level, producing a T-shaped incision. In the lumbar spine and at the thoracolumbar junction, a classic retroperitoneal (abdominal or thoracoabdominal) approach is performed. The vertebra is cut by chisel or osteotome far from the tumor (at least one zone is free from tumor) after protecting the major vessels (isolated by the anterior approach), obtaining an en bloc excision (Figure 7B).

Resection of the Posterior Arch (Marginal/Wide)

When the tumor is located between the zones 10 and 3, en bloc excision can be performed by a posterior approach (Figures 8A, 8B, 9A, and 9B).[4] To achieve this result, a wide laminectomy must expose the dural sac above and below the tumor. Lateral dissection must expose the pedicles, which are sectioned by osteotome or Gigli saw (Figures 8B and 9B).

■ Conclusions

The purposes of this update are to clarify the meanings of the common surgical terms used in oncology and to describe a precise application of the Enneking staging system to the spine. Primary tumors of the spine are rare, but the principles of treatment should be based on those that have proved to be effective in the management of primary tumors of the extremities. A complete work-up is needed, including achievement of a histologic diagnosis to stage the lesion. The oncologic staging is based on an understanding of the biologic behavior of the tumor and helps to decide which kind of treatment ("surgical margin") is most applicable. It is recommended that surgical planning be made on the basis of a surgical staging system specifically developed for tumors of the vertebral column. The general adoption of a stricter use of terminology is essential. It increases the accuracy of the assessment of such factors as aggressiveness of the condition, the extension of the lesion, the management technique used, and the outcome, which enables the establishment of more valid multicenter studies.

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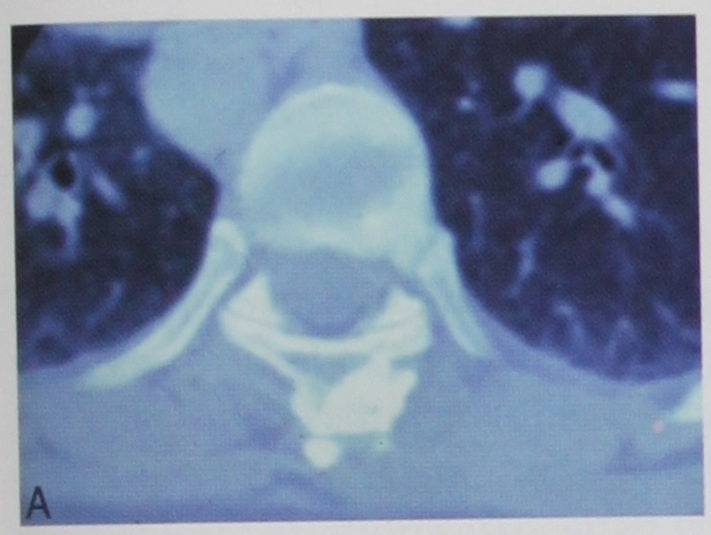

Figure 9. Recurrent chondrosarcoma of T8. **A,** The tumor occupied the posterior arch without involving the pedicles. No biopsy was performed. **B,** The lesion was removed en bloc together with the scar of the previous excision (see Figure 8B); note the inner surface of the canal (arrow) and the cut pedicle (arrow head). There was no recurrence of the tumor 42 months later.

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Address reprint requests to

S. Boriani, MD
Department of Orthopaedics and Traumatology
Ospedale Maggiore
Largo B. Nigrisoli 2
Bologna
Italy