and a malignant neoplasm in the spine is frequently impossible to make on radiographic grounds alone.

Angiography of tumors of an extremity is frequently performed prior to the definitive operation as well as prior to delivering intraarterial chemotherapy or reducing intraoperative bleeding by embolization. Although tumor neovascularity is frequently nonspecific, the involvement of major neurovascular bundles may compromise attempts at limb salvage resections and necessitate amputation. Such considerations are not of great importance in the evaluation of tumors of the spine, in which most operative procedures are intralestional; however, they are important in evaluating sacral tumors in order to identify tumor neovascularity and reduce this by preoperative embolization. If a tissue diagnosis has already been obtained, the authors believe that angiography should also be performed in patients with highly vascular tumors of the true vertebra with a view to reduction of tumor vascularity by the use of either polyvinyl alcohol (Ivalon) or absolute alcohol. In addition, it may be necessary to identify segmental feeding vessels of the spinal cord if extensive tumor resections are planned.

Tissue diagnosis by open biopsy is frequently required to plan a multidisciplinary approach that may include either preoperative chemotherapy (neoablative) or preoperative irradiation. If a soft-tissue mass is present, a carefully planned needle biopsy may provide accurate diagnosis of a malignant neoplasm (spindle cell versus round cell versus pleomorphic tumor) in almost all cases. For vertebral lesions, computed tomography-directed percutaneous needle biopsies may provide accurate diagnosis in more than 80 per cent of cases; most nondiagnostic studies result from attempting needle biopsy of purely osteoblastic tumors. When open or core biopsy is required, principles of operative oncology must be carefully adhered to. Biopsy sites must be placed so that they can be subsequently excised; tissue compartments must not be contaminated by extensive blunt dissection; hemostasis must be meticulous, with the liberal use of gelatin sponge (Gelfoam); and cortical bone windows must be plugged with methyl methacrylate. "Decompressive laminectomy" is to be condemned, since this results in widespread tissue contamination. If a posterior approach is chosen, a small hemilaminectomy should suffice, if performed under radiographic control. Fresh specimens of tissue obtained in difficult or complex cases should be sent for electron microscopy and immunoperoxidase studies. Tissue culture should also be done to detect the presence of chronic infections.

**Diagnosis and Management of Specific Tumors**

**CHORDOMAS**

Chordomas are slowly growing, locally aggressive neoplasms of the axial skeleton and account for 1 to 4 per cent of all malignant bone tumors. Their presumed origin is from embryonic notochord. This structure is closely associated with the development of the axial skeleton between the fourth and seventh weeks of embryonic life and then regresses so that the only remnant of the notochord in adult life is the nucleus pulposus.

Approximately 50 per cent of chordomas arise in the sacrococcygeal region, 35 per cent at the clivus, and 15 per cent in the true vertebrae. Ectopic chordomas arising in the maxilla, sinuses, larynx, and other soft tissues are occasionally reported. However, there is no convincing evidence for the origin of the tumor from the one structure traditionally associated with the notochord, i.e., the intervertebral disc. Until recently, cure by operation has been believed to be rare because these tumors are seldom diagnosed at a stage that would permit total complete excision.

Over a 35-year-period, a total of 88 patients with chordoma were treated at the Memorial Sloan-Kettering Cancer Center. The male to female ratio was approximately 2 to 1, and the ages of the patients ranged from 2 to 74 years, with a median age of 56 years. This tumor occurs predominantly in the fifth through seventh decades of life, which is also the peak incidence for the occurrence of metastatic cancer to the spine. The presenting symptoms and signs vary with the location of the tumor. In general, symptoms associated with sacrococcygeal tumors were present for

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*See references 57, 73, 75, 87, 93, 120, 126, 128, 130, and 173.

*See references 34, 38, 67, 89, 125, and 151.
more than 1 year before diagnosis in the authors' original series. The most frequent symptoms are pain in the low back or alteration in bowel function (change in bowel habits, tenesmus, or bleeding per rectum). Many patients are therefore treated for degenerative arthritis, disc disease, coccygodynia, or hemorrhoids for many months before the diagnosis of the tumor is entertained. In almost all instances, a rectal examination should confirm a palpable presacral mass that does not involve the rectal mucosa. The symptoms in patients with vertebral chordomas are generally of shorter duration. These patients usually present with back and radicular pain, although tumors in the cervical region may produce dysphagia because of an expanding retropharyngeal mass. In the lumbar region, or occasionally in the pelvis, the tumor may mimic a retroperitoneal mass.

The most consistent radiological finding in sacral chordomas is the destruction of several sacral segments associated with a soft-tissue mass anterior to the lesion (Fig. 124–1). Calcification is frequent in sacral tumors, and the reported incidence varies from 40 to 80 per cent. Unlike the central, flocculent pattern seen in cartilage tumors, the pattern of calcification in sacral chordomas is less dense, amorphous, and peripheral. Chordomas occurring in true vertebrae usually originate in a single body and produce multifocal lytic changes with reactive sclerosis (Figs. 124–2, 124–3). This may ultimately lead to vertebral collapse. Although contiguous vertebrae may be involved, the adjacent intervertebral disc spaces are usually spared. The finding of an anterolateral soft-tissue mass that is often much larger than the osseous component should suggest the diagnosis of chordoma.

Myelography should be carried out in all patients with tumors above the sacrum, as this study will reveal varying degrees of epidural extension. However, with the advent of magnetic resonance imaging (Fig. 124–4), myelography may no longer be required in the future.

More recently, the authors have encountered smaller tumors than were seen previously. Thus, 50 per cent of sacral tumors involved the third through the fifth sacral segments, which allows potentially curative resection to be performed. Three vertebral lesions were almost completely intraosseous, with minimal epidural extension.

Arteriography generally discloses avascular tumors with minimal tumor stain that is brought out only by subtraction techniques.
If marked neovascularity is seen, the diagnosis of chordoma should be reconsidered.

On gross examination these tumors appear as lobulated gray cystic or solid masses. Their consistency varies from firm, ossified, or cartilaginous in some tumors to soft myxoid, gelatinous, or even semifluid in others. In soft tissue, the tumors are circumscribed with a clearly formed pseudocapsule; however, the pseudocapsule is absent in the region of bone invasion by tumor.

Microscopically, chordomas are characterized by a distinct lobular architecture formed by several cell types, i.e., “physaliphorous” cells with ample vacuolated cytoplasm and the “signet-ring” type of cells. A thick layer of fibrous investing septa is often seen, which is often invaded by infiltrating tumor cells. The intracytoplasmic mucous droplets usually stain positively for glycogen and mucin. The smaller, better-preserved tumor lobules usually demonstrate oval or polygonal cells arranged in a lobular pattern that often resemble carcinoma cells with mucin production. On occasion, the histological distinction between chondrosarcoma and chordoma may be difficult, especially in tumors occurring in the sphenoid-occipital region. Chordomas usually stain positively with phosphotungstic acid hematoxylin and are readily impregnated by silver reticulin, whereas chondromas are largely unaffected by these reactions. More recently, a histologically distinct subtype (chondroid chordoma) has been identified; these tumors are more common in the base of the skull and are generally more indolent than other chordomas. Following radiation therapy, a prominent spindle-cell sarcoma pattern may be observed, which on occasion results in transformation into a highly malignant spindle-cell sarcoma or malignant fibrous histiocytoma.135,169

Although chordomas are thought to be locally aggressive tumors, the authors’ data suggest a considerable propensity for metastases. In various operative series, the incidence of metastases varies from 28 to 40 per cent, and metastatic spread generally occurs by hematogenous dissemination to the lung, bone, soft tissue, liver, and other sites. In the authors’ original series, metastases were more frequent in vertebral tumors when compared with tumors involving the sacrococcygeal region. They appeared uniformly throughout the course of therapy but had little influence on overall survival.

Once the diagnosis of chordoma is established, the correct treatment is complete resection when feasible. Transrectal biopsies of presacral masses should not be performed. A small biopsy through the posterior approach, with the site selected so that it can be excised at the time of definitive operation, is indicated if needle biopsy does not provide a diagnosis.

For small sacral lesions, a combined perineal and posterior sacrorectal approach suffices for tumor resection. The patient is positioned initially in the lithotomy position, and the perineal dissection is performed to free the rectum and anus from the tumor. Laterally, the attachments of the sacrum and coccyx are cut. The presacral space is dissected and packed off, and the patient is turned prone for the sacrectomy. This involves developing skin...
for rectal cancers. If the rectum can be spared, the first part of the operation is an anterior extraperitoneal dissection to free the rectum from the sacrum and presacral tumor. This approach is performed through a low-lying transverse abdominal incision with the patient in the supine position. The recti are transected, and the muscular layers lateral to the recti are sectioned down to the paretal peritoneum. This is then retracted from either side, and the posterior paretal peritoneum is dissected free, with bilateral mobilization of the ureters. The hypogastric vessels are mobilized, and both dissections should meet in front of the sacral promontory. The internal hypogastric vessels are ligated to minimize blood loss; similarly, the median sacral artery should also be ligated. Both the internal iliac artery and the internal iliac vein are divided separately, and the ends are secured by suture ligatures. The lateral and median sacral arteries and veins are divided and ligated. The periosteum should then be stripped off the sacrum, starting at the promontory and continuing down to the level selected for the osteotomy. This osteotomy can be through the canals of S1 nerves or above these canals. The sympathetic trunk passes anterior to the S1 nerve roots at this level and has to be cut. The lumbarosacral nerve trunks, including the L4–L5 nerve roots, pass anterior to the sacral wing and the sacroiliac joint, and they are mobilized so that they can be protected when the osteotomy is performed. Using an osteome or a high-speed drill, the anterior cortical wall of the sacrum is cut through the level of the proposed osteotomy. This osteotomy must be wide enough so that it can be palpated by a finger inserted underneath the edge of the ilium in the bottom of the greater sciatic notch. The anterior wound is now closed primarily after insertion of drainage tubes.

The posterior portion of the operation begins with the patient turned to the prone position. A long midline incision to include the lamina of L5 is made, but the more inferior skin and underlying subcutaneous tissues over the lower part of the sacrum have to be removed because of presumed infiltration by tumor. If a biopsy has been performed, the entire tract should also be excised. Skin flaps are raised on either side and the gluteus maximus muscle is transected away from the sacrum. The piriformis muscle is also cut. The superior gluteal nerve is preserved as it runs under the edge of the ilium at the bottom of
the greater sciatic notch. The sacrotuberous, sacrospinous, and ligamentous attachments to the coccyx are released. Bone is removed over the S1 segment, and the dural sac is exposed through a partial laminectomy, which should include L5. The dural sac should be ligated and divided to prevent cerebrospinal fluid leakage. The level of transection will depend on whether the S1 segments have to be preserved. Once the dural sac is ligated, the sacrum is osteotomized between the S1 and S2 segments. If the S1 nerves have to be sacrificed, the line of the osteotomy should be oblique and in the plane of the S1 canal. Laterally on either side, the osteotomy should include a portion of the posterior ilium. When making this cut, it is important to palpate from behind the osteotomy cuts that were made through the pelvis anteriorly. Once the transection of the bone has been completed and all the sacral roots to the sciatic nerve have been severed bilaterally at the level of the greater sciatic notch, the specimen can be removed in one piece. After careful hemostasis, the posterior edge of the resection is terminated by suturing the flaps with a drain.

The major functional effects of such sacral resections include loss of bladder and sphincter function, as well as anorectal function. In addition, genital function will be impaired, with loss of sensibility of the external genitalia. If the S1 nerves are preserved bilaterally, the motor deficit in the lower extremities will be minor. The major loss will be related to gluteus maximus impairment. If both S1 nerves have to be sacrificed, function of the foot will be impaired mainly as a result of loss of plantar flexion. However with time, innervation by L5 may suffice to minimize this loss as well. Surprisingly, no major loss of pelvic girdle strength is noted following such an extended operation.

Since the majority of patients do not undergo curative resection, additional therapy, including postoperative irradiation, has to be considered. Chordomas are radioresistant tumors, although most series have reported subjective improvement in pain and reduction of tumor mass following irradiation. Cummings and co-workers believe that the response of this tumor to radiation may be extremely slow, and when the dose—symptomatic response rates were reviewed, no significant dose response effect was seen in the range 3500 to 8500 rads (35 to 85 Gy).34,35 In the present authors' experience, conventional external radiation therapy has had no major effect in delaying symptomatic tumor recurrence and therefore its value following tumor resection is not established. However, Rich and associates recommend postoperative radiation therapy for all patients with residual disease, especially for those with tumors located in the true vertebrae and the base of the skull.135 In two small series, high-dose particle beam therapy has been employed for unresectable or partially resected tumors using proton beams or helium/neon beams.135,140,145 The results, as determined by local control, are impressive, but the follow-up period for these specialized techniques is relatively short in view of the long natural history of the tumor. In general, the current status of radiation therapy would appear to be that of an effective palliative agent, with doses of about 4000 to 4500 rads being most likely to be of benefit.

A variety of combination chemotherapeutic agents used in the treatment of sarcomas have been tried for recurrent chordomas or metastatic disease due to this tumor, but no significant effect has been discerned. In view of the fact that smaller tumors are now being discovered, the disease-free survival at 5 years should be in the range of 30 to 50 per cent at present, compared with 10 per cent in the past.

**OSTEOSARCOMA-(OSTEOGENIC SARCOMA)**

Osteosarcoma is a high-grade malignant spindle cell tumor that is characterized by the production of osteoid directly by the stroma. It is the most common primary malignant tumor of bone (excluding myeloma), with an incidence of one to three cases per million population. Approximately 500 to 1500 cases are diagnosed annually in the United States. The majority of tumors arise at the growing ends of long bones in the region of the knee and less than 5 per cent arise from the axial skeleton.10,38 Most series suggest a slight male preponderance of 1.3–1.6 to 1.0. Although the majority of tumors of long bones develop during the first two decades of life, tumors arising from the spine have a slightly older age distribution. Histologically, these tumors are further subdivided into fibroblastic, chondroblastic, osteoblastic, or mixed variants. In addition, some tumors have numerous areas...
tumors are stage IIb at diagnosis. In addition to plain radiography, computed tomography and myelography are required to outline the extent of tumor if a magnetic resonance imaging scan is not available. Radionuclide bone scans and computed tomography or plain tomography should be performed to establish the stage of the tumor prior to definitive therapy. If a soft-tissue component of the tumor is evident, a carefully placed needle biopsy should be performed to establish the histological diagnosis of malignancy. Unfortunately, the majority of patients referred for treatment have frequently undergone decompressive laminectomy for neural decompression, resulting in contamination of tissue planes. Patients with primary malignant bone tumors of the spine should be evaluated carefully to determine the optimal method of establishing a tissue diagnosis and to decide whether an anterior approach can be used in association with subtotal spondylectomy at the initial operation.

Prior to 1972, the prognosis for all patients with osteosarcoma was bleak. The only treatment possible for lesions of an extremity was amputation. This was followed shortly by the appearance of metastatic disease in the lungs, with a 5-year survival of 20 per cent. This

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*See references 1, 9, 15, 17, 22, 56, 59, 70, 79, 104, 106, 110, 113, 118, 119, 139, 141, 142, 168, and 172.

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Figure 124–5. Osteogenic sarcoma of C3. A. Lateral radiograph shows destruction and almost complete collapse of the vertebral body of C3 with posterior protrusion of the bony component into the spinal canal. There is anterior displacement of the air shadow of the hypopharynx due to a paraspinal soft-tissue mass. B. Corresponding computed tomography in the axial plane. There is significant compromise of the anteroposterior diameter of the spinal canal. Destructive bony change is accompanied by a lobulated anterior and left lateral soft-tissue mass.