Primary bone tumors of the spine in children

ALBERT J. FENOY, M.D., JEREMY D. W. GREENLEE, M.D., ARNOLD H. MENEZES, M.D., KATHLEEN A. DONOVAN, A.R.N.P., YUTAKA SATO, M.D., PATRICK W. HITCHON, M.D., AND JOHN C. CHALOUPKA, M.D.

Departments of Neurosurgery and Radiology, University of Iowa Hospitals and Clinics, Iowa City, Iowa

Object. Tumors originating in the vertebrae in children are difficult to treat. In this paper the authors sought to evaluate the decision-making process and outcome of surgical intervention in this population given the complex issues of spinal stability, continued skeletal growth, intraoperative blood loss, and long-term outcome.

Methods. To select patients for this study, the authors retrospectively reviewed medical records and images at the University of Iowa Hospitals and Clinics between 1996 and 2005. Their inclusion criteria were age younger than 18 years at the time of diagnosis and histopathological findings confirming that the tumor originated from vertebral bone. Sixteen patients met these requirements. In addition, the authors conducted a comparison with 45 patients in whom similar diagnoses were made prior to 1996. Gross-total resection of all nonmetastatic primary bone tumors is desired, as exemplified in 11 patients in this series; biopsy sampling only was performed in two others. Gross-total resection was also not performed in three patients with eosinophilic granuloma (EG). These three patients underwent nonsurgical treatment, which is different from how patients with EG were treated in the earlier study. Nine histopathological diagnoses were included; with a mean follow-up period of 3.7 years, the survival rate is 94%. The tumor recurred in one patient with a giant cell tumor of the sacrum. The authors performed preoperative tumor embolization and found that it was a useful adjunct to resection. Provocative testing prior to embolization was part of the protocol to reduce ischemic complications. Motion-sparing surgical procedures were performed in which a few segments were fused, preserving axial mobility.

Conclusions. Overall, early intervention offers the best symptomatic relief, which can only be rendered if sufficient clinical suspicion provokes early diagnostic imaging.

KEY WORDS • spine • primary bone tumor • pediatric neurosurgery

Primary bone tumors involving the spine are rare in children. The exact incidence varies by histological subtype, which is broad and includes osteoid osteoma, osteoblastoma, osteochondroma, osteosarcoma, EG, ES, aneurysmal bone cyst, chordoma, mesenchymal chondrosarcoma, giant cell tumor of bone, fibrous dysplasia, fibroma, angiosarcoma, and hemangioma. An example is EG in which spinal lesions have been reported to occur in 6.5 to 25% of total cases involving bone.14,43 Ewing sarcoma is the most common primary osseous spinal tumor in children,59 but less than 10% of ESSs originate in the vertebral column.70

This group of diverse tumors is difficult to treat for many reasons. Treatment issues include spinal stability, deciding whether to undertake surgical intervention, and preservation of neurological function. Unlike adults, children have not achieved complete skeletal growth, which must also be taken into account.

In a previous series conducted at our institution the authors reviewed 45 patients in whom primary bone tumors of the spine were diagnosed between 1951 and 1996.6 It was found that single-stage complete resection and stabilization were most likely to provide satisfactory long-term outcomes.6 We undertook the present report to evaluate the effects of improved technological advances and treatment on outcome in this patient population.

Clinical Material and Methods

All research was approved by the University of Iowa institutional review board. Multiple institutional databases were searched using the following inclusion criteria: age at diagnosis younger than 18 years, histopathological findings consistent with origination from vertebral bone, and diagnosis between 1996 and 2005. The databases that were searched included the institutional tumor registry, records of in- and outpatient visits, the pediatric neurosurgery database, and the pediatric neuroradiology database.

Once patients were identified as meeting these criteria, their medical records including inpatient and outpatient documentation, operative reports, and imaging findings were
Primary bone tumors of the spine in children

reviewed retrospectively. Attention was given to the chief complaint, presenting symptoms, physical and neurological findings, imaging findings, extent of disease, treatment, histopathological findings, complications, and outcome. Comparisons were then made with the previous study.

Results

Since 1996, 16 patients were identified who met the inclusion criteria. There were seven girls and nine boys who ranged in age from 2.6 to 17 years (mean 9.5 years). The follow-up period ranged from 0.3 to 8.4 years (mean 3.7 years).

The most common chief complaint was spinal pain in 12 patients, weakness in three, and an asymptomatic palpable mass in one. Additional symptoms included impaired range of motion in the neck, head tilt, urinary difficulty, numbness, and epigastric pain. The mean duration of symptoms was 13 weeks.

Examination at the time of initial presentation revealed abnormal findings in all patients. Neurological deficits were noted in six patients, of whom each had combined sensory and motor deficits. Seven patients had pain on palpation of the affected area, three had palpable masses, and one had a local lymphadenopathy that was discovered during the general physical examination. Three patients exhibited torticollis. A summary of presentations is provided in Table 1.

The imaging evaluation comprised plain radiography, CT, and MR imaging of the affected spinal levels in all patients. The levels involved were cervical in 11 patients (five subaxial), thoracic in two, and lumbosacral in three (Table 2). The tumors tended to be lytic, expansile lesions with involvement of paraspinal tissues. Two patients had intradural extension, and nine exhibited evidence of instability. On imaging studies the disease was found to be localized to one site in 13 patients; two separate lesions were observed in two patients, and a disseminated lesion was noted in another. One of the patients with two lesions (Case 4) had previously undergone excision of a symptomatic skull lesion (EG).

On comparison of presenting symptoms in the current series with that of the previous series of Beer and Menezes, the main differences observed were a shorter mean duration of symptoms (13 weeks compared with 20 weeks) and decreased incidence of neurological deficits (weakness: 38% compared with 74%); Table 1). Similarly, radicular pain was present in 19% of the current patients compared with 39% of the earlier ones. Whereas all patients in the current study presented with abnormal physical status, only 77% in the earlier series had abnormal physical findings.

The initial treatment plan for these children was based on possible diagnosis, extent of disease and neurological deficit, spinal stability, and growth potential. Gross-total resection and spinal stabilization were performed in 11 of the 16 patients. Two patients underwent percutaneous biopsy procedures alone; one (Case 9) who had disseminated disease at presentation and another (Case 5) with an asymptomatic lumbar mass. Three patients underwent nonsurgical treatment, including one in whom EG had already been diagnosed in the skull lesion (Case 4), and two in whom EG was diagnosed clinically (Cases 6 and 7). Preoperative embolization was performed in eight of the 11 patients who underwent resection (Cases 1, 3, 8, 10–12, 15, and 16).

In addition to resection four of the 11 patients received adjuvant radiation therapy (Cases 2, 3, 8, and 14), one of whom was also treated using chemotherapy (Case 14; Table 2). Of the five patients who did not undergo surgery, two received chemotherapy (Cases 4 and 9), one of whom also received radiotherapy (Case 9), and one patient was treated with radiotherapy alone (Case 5). Two patients with EG were treated using external orthoses and analgesic agents alone (Cases 6 and 7).

The following diagnoses were made based on histopathological findings: EG (in four patients); chordoma, ES, fibrous dysplasia, and osteoblastoma in two each; and aneurysmal bone cyst, giant cell tumor, mesenchymal chondrosarcoma, and fibroma in one each. Figure 1 shows the distributions of histopathological findings for both the current and previous series. Eosinophilic granuloma constituted a higher percentage of diagnoses in the current study (25% compared with 12%). With an average follow-up period of 3.7 years, the survival rate is 94%. Recurrence was noted in one patient (Case 13), requiring additional resection of a giant cell tumor of the sacrum. One patient (Case 9) presented with extensive ES at the craniovertebral border and had persistent residual tumor years later; this patient subsequently died of metastatic disease.

At the last follow-up examination, 10 patients reported resolution of their initial symptoms, whereas five noted improvement; one patient with metastatic ES died (Case 9). No other patient reported symptom worsening. Neurological deficits that were noted in six patients at presentation resolved in three (Cases 8, 14, and 15) and improved in three (Cases 2, 3, and 13) at the last examination. Radiographic fusion was demonstrated in all patients treated with internal fixation. There was no evidence of tumor on postoperative imaging studies in 13 patients, whereas three were shown to have persistent disease (Cases 2, 6, and 9). In a 6-year-old girl in whom EG had been diagnosed (Case 6), residual T-8 vertebral plana was noted at the last follow-up examination, but it was asymptomatic.

Complications in the series included recurrent pulmonary

### Table 1

<table>
<thead>
<tr>
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<tbody>
<tr>
<td>symptom/finding</td>
<td></td>
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</tr>
<tr>
<td>back or neck pain</td>
<td>13 (81)</td>
<td>35 (78)</td>
</tr>
<tr>
<td>torticolisis</td>
<td>3 (19)</td>
<td>—</td>
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<tr>
<td>radicular pain</td>
<td>3 (19)</td>
<td>18 (40)</td>
</tr>
<tr>
<td>paresthesia</td>
<td>6 (38)</td>
<td>—</td>
</tr>
<tr>
<td>weakness</td>
<td>6 (38)</td>
<td>35 (78)</td>
</tr>
<tr>
<td>paresthesia &amp; bladder dysfunction</td>
<td>2 (13)</td>
<td>6 (13)</td>
</tr>
<tr>
<td>normal examination</td>
<td>0 (0)</td>
<td>10 (22)</td>
</tr>
<tr>
<td>mean symptom duration (wks)</td>
<td>13</td>
<td>20</td>
</tr>
<tr>
<td>resolution of symptoms</td>
<td>13 (81)</td>
<td>32 (71)</td>
</tr>
<tr>
<td>after surgical treatment</td>
<td>9/11 (81)</td>
<td>—</td>
</tr>
<tr>
<td>after nonsurgical treatment</td>
<td>4/5 (80)†</td>
<td>—</td>
</tr>
<tr>
<td>improvement in symptoms</td>
<td>15 (94)</td>
<td>37 (82)</td>
</tr>
<tr>
<td>survival rate (%)</td>
<td>94</td>
<td>93</td>
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<tr>
<td>mean FU (yrs)</td>
<td>3.73</td>
<td>12.7</td>
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</tbody>
</table>

† All five patients had EG.
embolism in one patient (Case 14), which was successfully treated using a vena cava filter and anticoagulation therapy (Case 14). This patient also had radiation-induced gastritis. One patient (Case 10) had a pin-site infection during halo immobilization. No patient experienced any permanent neurological morbidity due to surgery.

### Illustrative Cases

**Case 8**

This 11-year-old boy presented with a 4-month history of low-back pain and a 1-month history of weakness in the right lower extremity. Examination revealed weakness in the right quadriceps muscle and hypesthesia and hypalgesia in the anterior thigh. His gait was unsteady, and the lumbar spine was tender on palpation. Imaging showed a pathological fracture of L-4 with retrolisthesis (Fig. 2). Magnetic resonance imaging revealed the paraspinal extension of a mass from the L-4 VB with high-grade central canal stenosis. The patient was treated using selective endovascular tumor embolization followed by an L-4 corpectomy, L3–5 arthrodesis, and instrumentation. The pathological diagnosis was ES. Chemotherapy with vincristine, cyclophosphamide, and doxorubicin was followed by fractionated radiotherapy to a total of 40 Gy. At the 4-year follow-up examination, the patient was symptomatically well with no neurological deficit and no evidence of disease on images.

**Case 3**

This 13-year-old boy presented to an outside institution with a 6-week history of progressive occipital and cervical pain, gait abnormalities, and left arm and hand weakness. Imaging studies revealed a large destructive mass involving the C-3 and C-4 VBs with intradural and retropharyngeal extension (Fig. 3). The left VA was encased in tumor. The patient underwent intradural tumor resection, and a C2–5 osteoplastic laminectomy was performed. The lesion was diagnosed as a chordoma. The boy’s neurological deficits persisted, and his left shoulder and arm pain worsened. At this time he presented to our institution. The patient underwent endovascular occlusion of the left VA and feeding vessels of the tumor. The laminoplasty was revised with accompanying right C2–5 posterolateral fusion, which resulted in resolution of his pain. This procedure was followed by left anterolateral retropharyngeal resection of the C-3 and C-4 VBs and retropharyngeal chordoma. We performed C2–5 fusion in which femoral allograft was used. The patient’s radicular pain and weakness resolved. He underwent PBRT 4 weeks later. Three years later, there was no evidence of tumor on follow-up imaging.

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**TABLE 2**

Treatment and results at the last follow-up examination in 16 patients

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Diagnosis</th>
<th>Symptoms</th>
<th>Site</th>
<th>Extent of Tumor</th>
<th>Treatment</th>
<th>Outcome (yrs)</th>
<th>FU (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>6, F</td>
<td>aneurysmal bone cyst</td>
<td>AP</td>
<td>C-2 VB</td>
<td>spinal</td>
<td>TR &amp; F</td>
<td>normal</td>
<td>1.3</td>
</tr>
<tr>
<td>2</td>
<td>7, M</td>
<td>chordoma</td>
<td>SD &amp; MD</td>
<td>clivus to C2–3 disc space</td>
<td>C3–5 VBs</td>
<td>TR &amp; F, RT</td>
<td>improved</td>
<td>1.6</td>
</tr>
<tr>
<td>3</td>
<td>13, M</td>
<td>chordoma</td>
<td>AP, GD, MD, &amp; SD</td>
<td>C-2 (dens), C-5, T-2</td>
<td>spinal &amp; para; separate skull lesion</td>
<td>chemo</td>
<td>normal</td>
<td>6.5</td>
</tr>
<tr>
<td>4</td>
<td>3, M</td>
<td>EG</td>
<td>AP &amp; tort</td>
<td>L4–5 VB, pre-vertebral soft tissue</td>
<td>spinal &amp; para</td>
<td>RT</td>
<td>normal</td>
<td>8.4</td>
</tr>
<tr>
<td>5</td>
<td>4, F</td>
<td>EG</td>
<td>mass</td>
<td>T-8 VB</td>
<td>spinal &amp; para</td>
<td>brace therapy</td>
<td>asympt persistent plana</td>
<td>6.7</td>
</tr>
<tr>
<td>6</td>
<td>6, F</td>
<td>EG</td>
<td>AP &amp; tort</td>
<td>C-2 VB</td>
<td>spinal &amp; para</td>
<td>brace therapy</td>
<td>normal</td>
<td>0.6</td>
</tr>
<tr>
<td>7</td>
<td>9, M</td>
<td>EG</td>
<td>AP, MD, GD, &amp; SD</td>
<td>L-4 VB, rt para-spinal mass</td>
<td>C1 lat mass, lungs</td>
<td>TR &amp; F, RT &amp; chemo</td>
<td>normal</td>
<td>2.9</td>
</tr>
<tr>
<td>8</td>
<td>11, M</td>
<td>ES</td>
<td>AP, mass, &amp; fever</td>
<td>C-2–3 VB, dens</td>
<td>spinal</td>
<td>RT &amp; chemo</td>
<td>improved</td>
<td>1.8</td>
</tr>
<tr>
<td>9</td>
<td>12, M</td>
<td>ES</td>
<td>AP &amp; RP</td>
<td>C-2 VB, dens</td>
<td>spinal</td>
<td>TR &amp; F</td>
<td>normal</td>
<td>0.4</td>
</tr>
<tr>
<td>10</td>
<td>12, F</td>
<td>fibroma</td>
<td>AP, tort, &amp; mass</td>
<td>C-2–3 VBs</td>
<td>spinal &amp; para</td>
<td>TR &amp; F</td>
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<td>0.3</td>
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<tr>
<td>11</td>
<td>12, M</td>
<td>fibrous dysplasia</td>
<td>AP, tort, &amp; mass</td>
<td>rt C-1 lat mass</td>
<td>spinal &amp; para</td>
<td>TR &amp; F</td>
<td>normal</td>
<td>3.0</td>
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<tr>
<td>12</td>
<td>4, M</td>
<td>fibrous dysplasia</td>
<td>AP &amp; tort</td>
<td>AP, MD, SD, &amp; B/B</td>
<td>S1–2 VBs</td>
<td>TR &amp; F (multiple)</td>
<td>improved; recurrent</td>
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<tr>
<td>13</td>
<td>17, F</td>
<td>giant cell tumor</td>
<td>AP &amp; tort</td>
<td>RP, MD, SD, &amp; B/B</td>
<td>T3–5 pst elements</td>
<td>TR &amp; F, RT, &amp; chemo</td>
<td>normal</td>
<td>5.5</td>
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<td>14</td>
<td>14, M</td>
<td>mesenchymal chondrosarcoma</td>
<td>AP &amp; tort</td>
<td>RP, MD, SD, &amp; B/B</td>
<td>C-4 VB, laminae</td>
<td>TR &amp; F</td>
<td>normal</td>
<td>7.4</td>
</tr>
<tr>
<td>15</td>
<td>13, F</td>
<td>osteoblastoma</td>
<td>AP &amp; RP, MD, &amp; SD</td>
<td>C-2 VB</td>
<td>spinal &amp; para</td>
<td>TR &amp; F &amp; normal</td>
<td>1.4</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>10, M</td>
<td>osteoblastoma</td>
<td>AP</td>
<td>C-2 VB</td>
<td>spinal &amp; para</td>
<td>TR &amp; F</td>
<td>normal</td>
<td>1.4</td>
</tr>
</tbody>
</table>

* AP = axial pain; asymp = asymptomatic; B/B = bowel/bladder; chemo = chemotherapy; ED = extradural; GD = gait disturbance; ID = intradural; mass = palpable mass; MD = motor deficit; para = paraspinal; pst = posterior; RP = radicular pain; RT = radiotherapy; SD = sensory deficit; tort = torticollis; TR & F = total resection and fusion.
Primary bone tumors of the spine in children

Case 1

This 6-year-old girl originally presented to an outside institution with complaints of neck pain after participating in cheerleading. Prolonged evaluation including a bone scan, CT, and MR imaging revealed a C-2 mass for which she underwent a CT-guided biopsy, which was nondiagnostic. The patient was placed in a cervical collar, which relieved her pain. When she was referred to our institution, physical examination disclosed decreased reflexes in the upper extremities with hyperreflexia in the lower extremities, a present Babinski sign, and a positive Romberg sign; findings from the motor and sensory examination were normal. The original CT scan revealed a marked lytic lesion within the C-2 VB that extended toward the right C-2 pedicle; this finding was confirmed on MR images. Subsequent imaging performed 3 months later showed expansion of the lesion and crossing over the midline toward the left as well as a mass in the right lateral aspect of the spinal canal displacing the C-2 subarachnoid space to the left (Fig. 4). A presumptive diagnosis of an aneurysmal bone cyst was made. We performed embolization of the right VA and tumor feeding vessels prior to undertaking a right C-2 laminectomy via a posterolateral approach to resect the tumor; the C-2 VB was reconstructed with an autologous bone graft and the posterior C1–3 levels with an interlaminar rib graft. The diagnosis of an aneurysmal bone cyst was confirmed. On discharge from the hospital, the patient was pain free and findings from her neurological examination were normal at the 1.3-year follow-up visit; cervical spine films demonstrated excellent alignment with bone fusion.

Discussion

The current series of children with primary bone tumors originating in the vertebral column illustrates three points when viewed in comparison with the larger, earlier series from this institution. First, time to diagnosis has been significantly shortened. Second, preoperative embolization can be a useful adjuvant modality to aid in resection and control blood loss. Finally, EG can often be successfully managed without surgery.

![Pie charts. Histopathological diagnoses in the current (upper) and previous (lower) series. Note the higher incidence of EG in the current series. N = number of patients.](image1)

![Fig. 2. Case 8. Upper Row: Preoperative images. Lower Row: Images obtained at the last follow-up visit. A: Lateral radiograph demonstrating the initial destruction of the L-4 body with partial collapse (upper) and the L3–5 lateral fusion construct (lower). B: Coronal CT reconstruction (upper) demonstrating right hemivertebral body collapse (upper) and an anteroposterior radiograph showing the allograft and fusion construct (lower). C: Axial Gd-enhanced T1-weighted MR image showing marked compression of the thecal sac by extradural tumor and heterogeneous signal intensity of the L-4 VB (upper). Postoperative axial T1-weighted MR image showing no thecal sac compression or residual tumor (lower). D: Parasagittal T1-weighted MR image showing a large extradural component of tumor and loss of signal intensity in the L-4 bone marrow (upper). No extradural tumor is seen after surgery, chemotherapy, and radiotherapy (lower).](image2)
Time to Diagnosis

Spinal column tumors in children are being diagnosed earlier in the course of disease, as evidenced by the shorter symptom duration. Although it is known that malignant tumors typically have a shorter duration of symptoms than benign lesions, both entities are being treated earlier than in the past, as supported by comparisons of recent reports with earlier series. As a result of earlier diagnoses, more complete resections can be undertaken, leading to better outcomes.

In our study, symptoms were present for approximately 13 weeks prior to diagnosis, whereas in the earlier series the mean symptom duration was 20 weeks. The patterns of presentation in the two series were otherwise similar: pain was the most common symptom in both, although more patients in the earlier series exhibited motor deficits (74% compared with 37%). A possible explanation for this difference is the higher percentage of patients with EG in the present series. Vertebral EG often causes pain early on in the disease course and has a low associated incidence of neurological deficit; on the other hand, pain associated with other benign spinal tumors can be insidious in onset and thereby delay patients from seeking medical attention.

Imaging Techniques

The most likely explanation for the shorter duration of symptoms is that improved imaging techniques can reliably detect smaller lesions. Computed tomography and MR imaging were both widely available at the end of the earlier series (early 1990s), but clearly these modalities were unavailable to patients in the early years of that study. As a result, tumors may not have been identified on radiographs until they were larger and therefore were more likely to cause neurological deficits.

The widespread availability of three-dimensional CT reconstruction imaging has assisted surgeons tremendously in identifying the lesion and in preoperative planning; indeed, we routinely use this imaging modality (Fig. 4). Image guidance, which has the advantage of less radiation exposure to pediatric patients, is another new modality that may be used to assist the surgeon in operative planning and resection. Computed tomography and/or MR angiography have become increasingly useful in defining pertinent vasculature with fine resolution; because these noninterventional studies are associated with less risk, they may be used routinely and can assist the surgeon in deciding when conventional angiography with or without embolization is necessary.

Preoperative Embolization

Preoperative embolization of vertebral tumors is known to be a useful adjunct to surgery, facilitating resection by minimizing blood loss and improving visualization. This procedure is also reported to aid in reduction in surgical time and allow for a safer surgical course. The volume of hypervascular tumors shrinks after successful arterial embolization, often facilitating complete resection. The estimated amount of blood loss during resection of spinal column neoplasms after embolization has been reported to range from 1.5 to 2.2 L, compared with 9.5 to 15 L lost.

Fig. 3. Case 3. A: Initial sagittal T1-weighted MR image showing the large hyperintense mass both ventral and dorsal to the VBs (upper). Axial T1-weighted image demonstrating extension of tumor through the neural foramen with severe canal stenosis (lower). B: Follow-up sagittal T1-weighted image after posterior debulking showing residual retropharyngeal tumor and development of kyphosis (upper). Axial T1-weighted image showing resection of the extradural portion of the tumor (lower). The femoral allograft is seen in the corpectomy defect. C: Lateral view angiogram showing multiple small feeding vessels to the tumor. D: Follow-up angiography study image after occlusion of vessel showing no blood flow to tumor.
when preoperative embolization was not performed. The amount may be even greater when dealing with metastatic vertebral lesions.\(^8,22,56\)

Eight of the 11 surgically treated patients in this series underwent preoperative embolization (Cases 1, 3, 8, 10–12, 15, and 16). The mean estimated blood loss in these eight patients was approximately 305 ml. This value is much lower than the aforementioned amount; the reduction is probably due to a combination of the extent of embolization, surgical technique, pathological characteristics of the lesion, and the anatomical location of tumor. When possible, the tumor was excised in a circumferential manner with normal margins.

Provocative testing prior to definitive occlusion was performed when feasible to reduce complications related to the embolization procedure. In this series one patient (Case 3) underwent temporary balloon occlusion of the left VA because the vessel was encased by a cervical chordoma; the patient did not exhibit any transient deficit. Permanent occlusion with the placement of coils was then performed, allowing extensive en bloc tumor resection and producing an excellent outcome. Many authors prefer permanent rather than temporary occlusion for tumor resection.\(^24,46,68,71\)

Coils are safer than balloons because they provide a more complete occlusion of blood flow over a longer arterial segment and minimize the risk of spontaneous deflation or migration.\(^24,46,68,71\) The evolution of endovascular techniques during the past decade has allowed coils to become a useful adjunct to surgery, and as these techniques continue to be developed in terms of safety and durability, it is likely that coils will continue to be widely used.

**Types of Primary Bone Tumors and Options for Treatment**

Primary bone tumors can involve any portion of the spinal column and extend in any direction. Accordingly, a variety of surgical approaches may be used for resection. In posterior approaches, spinal deformity and instability are known possible consequences of a multilevel laminectomy in children, so consideration should be given to primary reconstruction or stabilization at time of resection. The incidence of postlaminectomy deformity in children has been reported to be as high as 25 to 46%\(^66,73\) and has been attributed to a loss of posterior supporting structures, which places greater biomechanical loads on the largely cartilaginous anterior VB and the remaining capsular facet ligaments.\(^73\)

Prophylactic osteoplastic laminotomy and reconstruction instead of laminectomy, followed by spine immobilization, has resulted in greater stability\(^32\) and improved outcome in some series.\(^1,44,54\) Although originally intended for lumbar disc herniation repair,\(^33\) this procedure in which posterior structures are removed en bloc allows wide exposure of the spinal canal and facilitates tumor excision; these elements are then replaced and fixed with screws to restore structural integrity.\(^32\) Complications, such as postoperative wound infection\(^65\) or pseudomeningocele,\(^25\) are similar to those that occur with laminectomy alone. Such techniques are considered motion sparing because they provide stability, yet they do not greatly compromise flexion and extension once fusion is complete. It has been the practice of the senior author (A.H.M.) to perform osteoplastic laminectomies in children to maintain anatomical alignment.

**Eosinophilic Granuloma.** Eosinophilic granuloma is known...
to be a local or systemic disease and as a result, there is a wide range of treatment regimens. These options also exist when the entity is restricted to the spine. Management options include aggressive resection, biopsy alone, combined chemotherapy and radiotherapy, and conservative treatment. Such treatments are chosen depending on symptoms and objective findings. No patient in the current series with EG was treated surgically. This is in contrast to the earlier series in which two of the five patients identified as having EG underwent gross-total resection and fusion for the treatment of cervical lesions. Both patients presented with pain, spinal instability, and neurological deficits. The four patients with spinal EG in this series (Cases 4 and 5–7) all presented without neurological deficits and were treated nonsurgically, attaining excellent outcomes at the last follow-up visit. In one patient (Case 6) persistent disease was noted on images but the patient is asymptomatic and prepubertal. Researchers have reported that resolution of symptoms in young patients with EG may precede radiographic reconstitution, and that the pubertal growth spurt may be required to reconstruct lytic bone.

Chordoma. Chordomas are rare, slow-growing neoplasms that are thought to arise from remnants of the primitive notochord and are predominantly located either in the sacrococcygeal region or in the clivus. Less than 5% of these tumors occur in children, making them extremely rare. Two patients in the earlier series had this pathological entity in the upper cervical spine (Cases 2 and 3), an uncommon location accounting for only 15% of chordomas. Skull base chordomas typically grow along lines of least resistance, posteriorly toward the craniovertebral junction. This growth pattern can result in an insidious onset of neurological symptoms due to slow encasement and compression of the brainstem and upper spinal cord, causing progressive bulbar motor and sensory deficits as well as the classic cruciate paresis. Chordomas are benign tumors but because of their critical location, local invasion, recurrence rate, and occasional metastatic spread, their prognosis is similar to that of malignant tumors. Radical resection is imperative; recurrences are treated with repeated operation followed by PBRT; traditional radiation therapy is not beneficial. Of the few pediatric cases reported in the literature, the 5-year disease-free survival rate is 63% following PBRT.

Ewing Sarcoma. Primary ES of vertebrae accounts for only 3.5 to 10% of total cases of osseous ES, yet it is the most common primary malignant lesion of the spine. Other sarcomas, such as mesenchymal chondrosarcoma and osteosarcoma, much more rarely have primary origins in the spinal column; all sarcomas more frequently metastasize to the spine from other locations. In cases of ES the principal presenting symptom is pain, and neurological deficits are seen in more than two thirds of patients on presentation, chiefly due to nerve compression. A short duration of symptoms prior to presentation is commonly observed caused by frequent spinal cord compression. Treatment is directed toward resection and aggressive chemotherapy, usually prior to radiation therapy.

In our current series, two of 16 patients were treated for ES; at the 2-year follow-up examination, one patient is disease free following surgery, chemotherapy, and radiotherapy (Case 8), whereas the other with disseminated disease who only underwent chemotherapy and radiotherapy had persistent tumor and ultimately died of widespread metastases (Case 9). Authors of the few extant series in the literature have cited a mean 5-year survival for vertebral ES of less than 50%. This prognosis is similar to that of patients with ES originating in other bones. Patients who present with disseminated disease have poorer outcomes, but tumor size is a less reliable prognostic indicator with the advent of newer chemotherapeutic agents.

Mesenchymal Chondrosarcoma. Mesenchymal chondrosarcomas are rare malignant tumors of the bone and soft tissue; of those occurring in the central nervous system, most are located intracranially, and intraspinal lesions are even rarer. Mesenchymal chondrosarcomas accounted for one of the pathological diagnoses in our series. These lesions frequently arise in the meninges and due to their vascularity may initially be misdiagnosed as angiofibroma or hemangiopericytoma, although destruction of the vertebral elements is seen and their histological features are distinct. Clinically, neurological symptoms of pain, weakness, sensory loss, and incontinence progress over a protracted period of time. In Case 14, and other similar patients, treatment was directed toward radical tumor resection with adjuvant chemotherapy and radiotherapy. After more than 5 years of follow up, there is no detectable tumor, although local recurrences long after the initial resection have been noted.

Osteosarcoma. Osteosarcoma is the most common primary malignant bone tumor, but less than 2% originate in the spine. Multiple metastatic lesions are usually found at presentation and have a shorter duration of neurological symptoms; they are treated using aggressive resection and adjuvant chemo- and radiation therapy. Their prognosis is traditionally poor, with reported median survival rates less than 10 months after diagnosis. In the present series we have not reported on any patients who received this diagnosis.

Osteoblastoma. Osteoblastoma was present in two patients in the present series (Cases 15 and 16) and six in the former. Like osteoid osteomas, which are histologically identical but several centimeters smaller, osteoblastomas are benign lesions that most commonly affect the posterior elements of the spine. Unlike osteoid osteomas, however, osteoblastomas are expansile lesions with less sclerosis and may extend beyond the vertebral arch, causing neurological symptoms due to compression of the spinal column; back pain and scoliosis on presentation are less stereotypical and the pain is not as easily relieved by aspirin. Osteoblastomas are rare, accounting for approximately 1% of primary bone tumors; fortunately complete resection is curative. The tumors can recur, and there is a risk of local invasion; in these cases radiotherapy and chemotherapy can be of use if complete resection is not possible.

Aneurysmal Bone Cyst. Aneurysmal bone cyst was one C-2 VB lesion in a 6-year-old girl (Case 1) in the present series. Although this benign pseudotumoral hyperemic lesion accounts for only 1% of all primary bone tumors, it accounts for nearly 15% of benign bone spinal lesions, and the majority appear before the patient reaches the age of 20. This disease entity is usually successfully treated via intralesional surgery, selective vertebral embolization, and/or radiotherapy. Our patient underwent selective arterio- venous embolization followed by complete curettage and autograft arthrodesis; at the 1-year follow-up examination, the patient is pain free and has not experienced a recurrence. In
Primary bone tumors of the spine in children

the series by Boriani, et al., the authors advocated that selective embolization may be sufficient, and that partial curettage of the cyst wall reduces intraoperative morbidity and maintains stability, obviating the need for an arthrodesis. However, two patients in this series were found to have recurrences after treatment with selective arterial embolization and partial curettage; indeed, recurrence has been noted after complete removal of cystic walls and typically is noted within 1 year following treatment. Repeated surgery or radiotherapy is then the option, and the prognosis is uniformly good.

Fibrous Dysplasia. Similarly, fibrous dysplasia can be successfully treated by resection, as was seen in the two patients in our current series (Cases 11 and 12). This entity that occurs in the spine of patients with monostotic disease is extremely rare; fewer than 25 cases have been reported in patients of all ages. With these few numbers, a variety of management options is reported in the literature including conservative and aggressive treatment, although most authors recommend excision. Outcomes are generally favorable although there are a few reports of malignant transformation.

Conclusions

The management of primary bone tumors arising from the vertebral column in pediatric patients has benefited from technological advances in both diagnosis and treatment. These tumors are being diagnosed in children earlier in the course of disease, and as a result the outcomes have improved. Aggressive resection and stabilization, aided by the use of preoperative embolization, is the treatment most likely to achieve long-term clinical success. However, evidence is mounting that for EG, nonsurgical management is the preferred option to effect symptomatic improvement.

References
