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Primary Tumors of the Spine in Children: A Review from the Pediatric Musculoskeletal Tumor Program at the Children's Hospital of Philadelphia

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Abstract: Although musculoskeletal tumors are the third most common type of tumor in the pediatric population, primary tumors of the spine are rarely encountered. Despite their low incidence, pediatric orthopaedic surgeons must always consider the diagnosis in any evaluation of a child with back pain. Although the vast majority of these tumors are benign, most require surgical intervention to achieve diagnosis and cure. Unfortunately, malignant tumors of the spine remain frequently fatal and require coordinated care from surgeons and oncologists in the attempt to achieve cure. A comprehensive review of the pediatric musculoskeletal tumor database for the presentation, treatment, and outcomes in children with primary tumors of the spine has begun at our institution to try and improve clinical characterization of these tumors as well as identify optimal treatment methods. This review will discuss the clinical presentation, diagnosis, natural history, and treatment of the most common primary tumors of the spine in children.

Introduction

Back pain is one of the most common reasons for patients to visit their physicians. In comparison to adults, the workup for back pain in children is extensive and aggressive at a much earlier stage in the evolution of symptoms. Children frequently have skeletal pathology as a cause of back pain. The most frequent causes of back pain in children are trauma, spondylolisis, spondylolisthesis, and infection. Tumors of the spine also invariably present with back pain (approximately 90% of cases) [1,2,3,4]. While tumors are an infrequent cause of back pain in children, they can be devastating and must be considered as a possible diagnosis in any child with back pain.

Thankfully, in children the vast majority of spine tumors behave in a benign manner and are readily curable with modern management. Three to nine percent of all skeletal tumors (pediatric and adult) involve the spine [2,5,6]. Using the pediatric musculoskeletal tumor database we retrieved the records of 112 children with tumors or tumor-like conditions of the spine (excluding metastatic disease) treated at our institution over the past thirty years. Of these, 78 (70%) were benign and 34 (30%) were malignant (Table 1). Tumor-like conditions with unknown etiology such as aneurysmal bone cyst (ABC) and eosinophilic granuloma are included since their behavior mimics benign tumors and oncologic principles of diagnosis, staging, and treatment are used. We have begun a comprehensive retrospective review of our experience with each specific tumor to shed insight on clinical presentation and outcomes following modern management of these infrequently encountered lesions.

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Clinical Presentation

As previously mentioned, the most common presenting symptom of spine tumors is pain. The pain can often is quite intense and is particularly suggestive of tumor if it awakes the child from sleep. Nevertheless, these are not absolutes and many children, particularly with benign tumors may not present with stereotypical pain. The pain from spinal tumors is usually localized to the affected level and can often be elicited with firm palpation. In comparison with benign tumors, the pain of malignant tumors usually increases in intensity more rapidly and can become debilitating in the course of a few weeks. On examination, pain with forward flexion of the spine suggests an anterior process while pain with extension suggests involvement of the posterior elements of the spine.

Neurologic symptoms are found to varying extent among the different spine tumors. Children with benign tumors of the spine will present with neurologic symptoms less commonly than those with malignancies. Of the benign tumors, only giant cell tumors and ABCs have a prevalence of neurologic symptoms at presentation greater than 25% [7,8,9,10,11]. For the most part, the neurologic symptoms consist of radicular pain, however, there are occasional reported cases of patients presenting with complete paraplegia [2,12,13]. Aggressive lesions low in the spinal column, especially the sacrum, may lead to bowel or bladder dysfunction [4,14]. Lee laid out four possible causes of neurologic symptoms from tumors of the spine: 1) direct compression of nerves by the tumor 2) osseus compression of nerves due to pathologic fracture 3) compression of vascular structures serving the nerves by tumor 4) direct tumor invasion of nerves [15]. In the workup of a child with back pain it is essential to perform and document a thorough neurologic

Table 1. Number of patients with primary tumors of the spine treated at the Children's Hospital of Philadelphia from 1970-2002.
(Malignant in BOLD)

Tumor	Number
Aneurysmal Bone Cyst	13
Chondrosarcoma	**1**
Chordoma	3
Ewing's Sarcoma/PNET	**23**
Fibrosarcoma	**2**
Giant Cell Tumor	1
Hamartoma	1
Hemangioma	16
Langerhans' Cell Histiocytosis	26
Leukemia*	**2**
Lymphoma*	**3**
Osteoblastoma	2
Osteochondroma	3
Osteoid Osteoma	7
Osteosarcoma	**3**
Teratoma	6
Total Malignant	***34***
Total Benign	*78*
OVERALL TOTAL	***112***

* Primary presentation in the spine

exam including checking for subtle findings of neurologic compromise such asymmetric abdominal reflex, clonus, and hyper-reflexia.

Other physical findings that are especially important to look for in the clinical presentation of a suspected spine tumor are scoliosis, kyphosis, and a palpable mass. Palpable masses have been appreciated in about 10–20% of all patients with spine tumors; with painless masses almost always a sign of a malignant tumor or neurofibromatosis [6,16,17]. Scoliosis is also frequently found to result from tumors of the spine. The deformity can be due to structural damage to vertebrae in the case of benign aggressive tumors or malignant tumors. In less aggressive tumors, however, the scoliosis is often due to paraspinal muscle spasm due to inflammation [18].

Diagnosis

Following a thorough history and physical examination targeted to elicit the particular features described in the clinical presentation of spine tumors, the next step in making a diagnosis is imaging. Any child who comes to the orthopaedic surgeon with a complaint of back pain should be sent for an AP and lateral plain x-ray of the affected area of the spine. Oblique views of the lumbosacral spine are useful also to look for spondylolisis and spondylisthesis. If scoliosis is detected on physical examination a standing three foot PA and lateral x-ray of the entire spine should be obtained. Two large case series of spine tumors, with 45 and 82 patients, found that greater than 98% of initial spine x-rays were abnormal [16,17]. These findings are biased, however, since the films were being looked at retrospectively following tissue diagnosis. Nevertheless, plain x-rays remain the first line imaging modality in the evaluation of back pain.

In most orthopaedic centers advanced imaging modalities are much more readily available than has been the case in the past. Computerized tomography (CT) scanning is a rapid and highly sensitive imaging technique to evaluate the structural integrity of the skeleton. Magnetic resonance imaging (MRI) offers highly detailed views of the skeleton and soft-tissues, with an added advantage of giving the physician views in several different planes. Bone scans with spect are also heavily utilized in the work-up of children with suspected tumors of the spine. They help localize disease when x-rays are unrevealing (especially useful for osteoid osteoma and osteoblastoma) as well as find multi-focal disease (especially useful in Langerhans' cell histiocytosis).

In adjunct to the plain x-rays that should always be obtained in the child with back pain, we advocate obtaining an MRI if there are any abnormal physical findings on neurologic exam or concerning points in the patient's history (e.g. complaints of numbness, paresthesias, weakness, etc.). Consideration should also be given to obtaining a bone scan if the x-rays do not reveal any abnormality, there are multiple sites of pain, to rule out multi-focal disease, and if the location of pain on exam is discordant with findings on x-ray. Along with high sensitivity for finding tumors, bone scan can also be useful when looking for spondylolisis, which is a very frequent cause of back pain in children. CT scan or MRI should be obtained prior to any attempt at biopsy in order to localize the disease process and plan the biopsy.

Obtaining a biopsy is an essential part of the work-up of a child with a spine tumor. Although advanced imaging and sampling techniques have made percutaneous biopsy of the spine possible, in many cases an open surgical biopsy is advised for spine tumors. Percutaneous biopsy often (40% of attempts) will not give sufficient tissue to do a comprehensive pathology evaluation. It can be used for initial biopsy, with the caveat that if the sample obtained is not diagnostic an open procedure will be done. A major advantage of an open surgical procedure is that definitive treatment of benign tumors can be done in one operation following preliminary intraoperative frozen section diagnosis. Surgical biopsy for a suspected tumor of the spine should only be performed by an orthopaedic tumor specialist, ideally the same one who will perform the definitive operation in the future if necessary.

Finally, many spine tumors have a predilection for a particular anatomic area of the spine and/or a particular age group. Knowledge of these tendencies can be helpful in narrowing the differential diagnosis of a spine tumor. Table 2 displays the anatomic distribution of common spinal tumors while Table 3 displays the breakdown of common skeletal tumors based on age at diagnosis.

Oncologic and Surgical Staging

Musculoskeletal tumors are usually staged based on the simple classification developed by Enneking. Benign tumors are graded either 1 (latent, inactive), 2 (active), or 3 (aggressive). Stage 1 benign lesions will burn out without treatment and require only close follow-up for signs of

Table 2. Common Locations of Pediatric Spine Tumors (Modified from Dormans J, Pill S. Benign and Malignant Tumors of the Spine. *Spine: State of the Art Reviews*. 2000;14:263-279, with permission)

Anterior (Vertebral Body)	Posterior (Spinous Process and Laminae)
Chordoma	Aneurysmal Bone Cyst
Giant Cell Tumor	Osteoblastoma
Hemangioma	Osteochondroma
Langerhans' Cell Histiocytosis	Osteoid Osteoma
Leukemia	
Lymphoma	
Metastatic Tumors	

change. Certain cases have accelerated healing following biopsy. Stage 2 benign lesions can be treated with intralesional curettage with good results while stage 3 benign (but locally aggressive) lesions require marginal excision to achieve good results. Malignant tumors are staged I-III based on size and spread of tumor. Stage I have low-grade histology, stage II have high-grade histology, and stage III have distant spread. An A or B is added to the classification based on intracompartmental location (A) or extarcompartmental location (B) [19]. This system is appropriate and effective in the staging of primary tumors of the spine.

Recently, two new surgical staging systems have been created that take into account the unique structural features of the vertebrae. The Weinstein-Boriani-Biagini system divides the vertebrae into a clock-face and divides the vertebrae into twelve radial segments. Compartmental location is noted from A-E (Fig. 1) [20]. Tomita's system marks lesions from 1-5 based on location in the vertebrae (1 – body, 2 – pedicles, 3 – posterior elements, 4 – spinal canal, 5 – intervertebral space) and into seven types based on number of affected areas and intra or extra-compartmental status (Fig. 2) [21]. These systems are useful for the orthopaedic tumor surgeon in planning complex resections of aggressive benign and malignant tumors of the spine.

Tumor surgery of the pediatric spine is extremely challenging. The orthopaedic surgeon must not only achieve appropriate margins, but also be cognizant of the effects of his surgery on the stability and future growth of the spine. Spinal stabilization and fusion procedures with both bone graft and instrumentation are often an integral part of tumor surgery in the pediatric spine. The use of titanium instrumentation systems has been an important advance in allowing spinal stabilization at the time of tumor resection while still allowing high quality MRI images to be obtained following instrumentation (unlike standard steel systems which produce a large amount of scatter artifact on MRI) [22].

Table 3. Peak Age of Common Pediatric Skeletal Tumors (Modified from Dormans J, Pill S. Benign and Malignant Tumors of the Spine. *Spine: State of the Art Reviews*. 2000;14:263-279, with permission)

Age (years)	Benign	Malignant
0-5	Langerhans' cell histiocytosis	Ewing's sarcoma Leukemia Neuroblastoma (metastatic) Wilm's tumor (metastatic)
5-10	Aneurysmal bone cyst Langerhans' cell histiocytosis Nonossifying fibroma Osteoblastoma Osteoid osteoma Unicameral bone cyst	Ewing's sarcoma Osteosarcoma
10-20	Aneurysmal bone cyst Chondroblastoma Osteochondroma Osteoid osteoma	Chondrosarcoma Ewing's sarcoma Osteosarcoma

Specific Primary Tumors of the Spine

Aneurysmal Bone Cyst

Aneurysmal bone cysts (which are neither aneurysms nor cysts) are highly vascular space-consuming lesions that aggressively lead to bone destruction. The etiology remains unknown, though most believe it is related to a disruption of normal vascular flow. They also frequently arise secondary to adjacent tumors, perhaps spurred by abnormal new vasculature feeding the primary tumor. Dahlin's series of 289 primary ABCs found 14% to affect the spine with predominance of lumbosacral lesions [5]. Of the benign tumors, this is one of the few that can span adjacent vertebrae though it does not violate the intervertebral disc. They almost always involve the posterior elements of the spine and can sometimes expand anteriorly into the vertebral body [7,9,10].

Of the primary tumor and tumor-like lesions of the spine, ABCs have among the highest rates of neurologic symptoms at presentation. Based on large series of ABCs of the spine, between 10–50% of patients will have neurologic symptoms on presentation ranging from radicular pain all the way to complete paraplegia [7,9,10,12,13,23]. As with most spine tumors, the main symptom is usually back pain that is progressive and unrelenting. X-rays usually reveal a large, expansile lesion that appears to be multi-loculated, with many septations, and often a thin blown-out cortical shell; MRI almost always will show multiple fluid-fluid levels. Other MRI characteristics that differentiate ABCs from unicameral bone cysts are: 1) double density fluid levels 2) septations 3) low signal on T1 images and high signal on T2 images [24]. Often the radiographic appearance is diagnostic (Fig. 3).

The treatment of aneurysmal bone cysts over the years has been highly varied. Surgical approaches have ranged from wide surgical resections to intralesional curettage. Medical treatments have focused on radiation therapy while recent advances in imaging have brought selective arterial embolization as a pre-operative adjunct and even as primary treatment for these lesions [7,25]. Cure without any treatment has also been observed [26,27]. Most commonly, extended intralesional curettage followed by bone grafting has been the surgery of choice for these lesions. Local recurrence rates are reported between 10–30%, frequently skewed due to higher rates of recurrence in cases where there was incomplete curettage [7,10,13,23,26,28]. Radiation treatment, while used in the past as primary treatment, or as post-operative adjunct, is no longer generally recom-

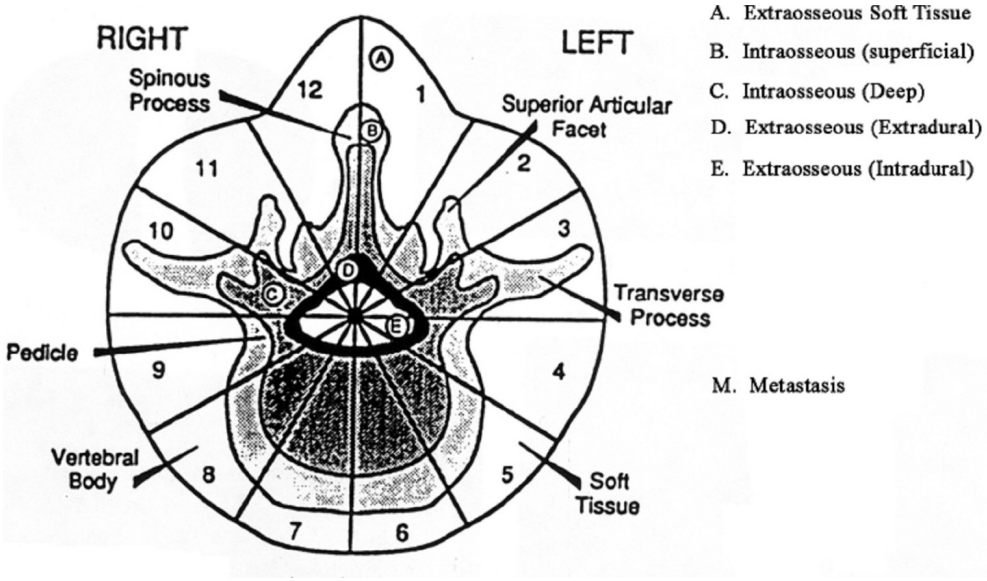

Fig. 1. Weinstein-Boriani-Biagini surgical staging system for spine tumors (From Boriani S, Weinstein J, Biagini R. Primary bone tumors of the spine: terminology and surgical staging. Spine. 1997;22:1036-1044, permission pending)

mended due to adverse affects on long-term spine stability and risk of post-radiation sarcoma [7,13,29].

Although ABCs are frequently classified as Enneking stage 2 or 3 benign tumors, given modern technologies and adjuncts, we now believe that wide or even marginal lesions are not usually necessary to achieve long-term cure. At our institution we have had excellent results following a four-step surgical procedure for removal of aneurysmal bone cysts. Following pre-operative selective arterial embolization (when appropriate) and confirmation of diagnosis on intraoperative frozen section, our surgery includes: 1) curettage 2) high-speed burr 3) electrocautery 4) phenolization, followed by bone grafting of the defect. Furthermore, due to the large and aggressive nature of these lesions, spinal stability is often compromised with surgical treatment of the primary lesion. In any case where spinal stability is felt to be at risk we advocate the use of instrumented spinal fusion with titanium systems to prevent development of future spinal deformity while allowing for good MRI surveillance for recurrence [22].

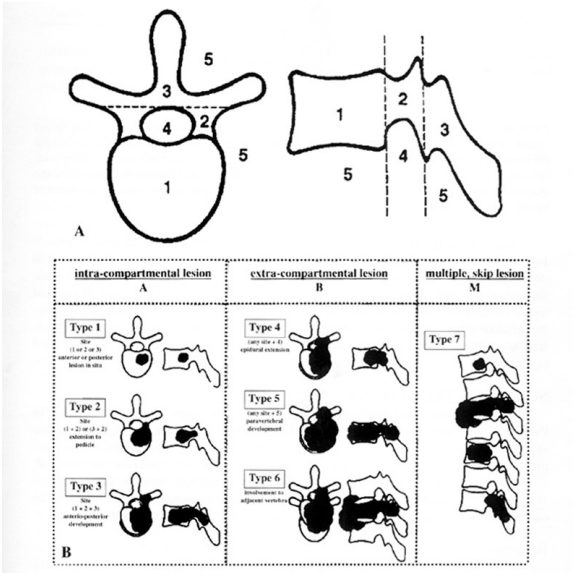

Fig. 2. Tomita surgical staging system for spine tumors (From Tomita K, Kawahara N, Baba H, et al. Total en bloc spondylectomy. A new surgical technique for primary malignant vertebral tumors. Spine. 1997;22:324-333, permission pending)

Langerhans' Cell Histiocytosis (Eosinophilic Granuloma)

The initial description of vertebral Langerhans' cell histiocytosis (LCH) was by Calve in 1925 [30]. The radiographic appearance described, with vertebral body collapse, preservation of disc space, and lack of soft tissue extension, is now termed vertebra plana (Fig. 4). The vertebral column is affected in approximately 30% of children with LCH involving the skeleton [31,32,33]. Dull, aching back pain is normally the only symptom of LCH at presentation [34,35,36,37,38,39,40,41,42]. Although pain is found in nearly 100% of patients with LCH of the spine, neurologic symptoms are rarely elicited at presentation. When present, the symptoms are generally radiculopathies that resolve promptly following treatment of the lesion.

The diagnosis of LCH is usually presumed following the finding of characteristic vertebral destruction on x-ray. The finding of vertebral collapse (regardless of extent) should prompt the treating physician to obtain a bone scan to look for multifocal disease since up to 50% of cases will present at multiple non-adjacent vertebral levels [35,38]. Despite a highly characteristic appearance on x-ray, pathologic diagnosis remains necessary to confirm the diagnosis of LCH. Malignancies such as Ewing's sarcoma, lymphoma, leukemia, and neuroblastoma also may present with a vertebra plana on x-ray. Because of this, we consider tissue diagnosis an essential part of LCH management. Juvenile adolescent idiopathic scoliosis can also mimic LCH on imaging and should be a diagnostic consideration.

The natural history of isolated skeletal LCH has been shown to be spontaneous healing of the lesion with reconstitution of destroyed bone.31,33,36,37,38,43 Various modali-

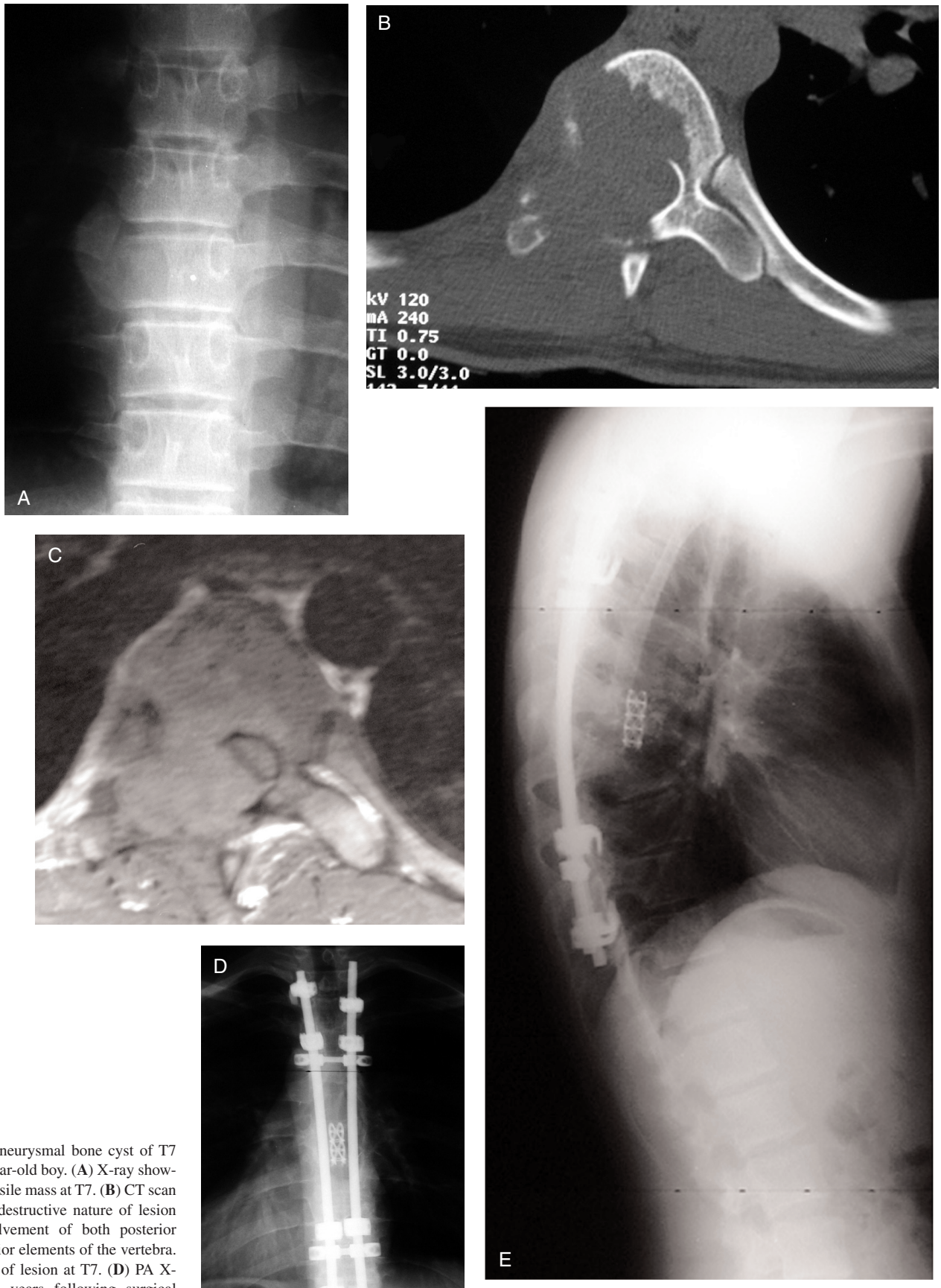

Fig. 3. Aneurysmal bone cyst of T7 in a 17 year-old boy. (**A**) X-ray showing expansile mass at T7. (**B**) CT scan showing destructive nature of lesion and involvement of both posterior and anterior elements of the vertebra. (**C**) MRI of lesion at T7. (**D**) PA X-ray three years following surgical treatment of ABC and stabilization of spine (**E**) Lateal x-ray three years following surgical treatment of ABC and stabilization of spine

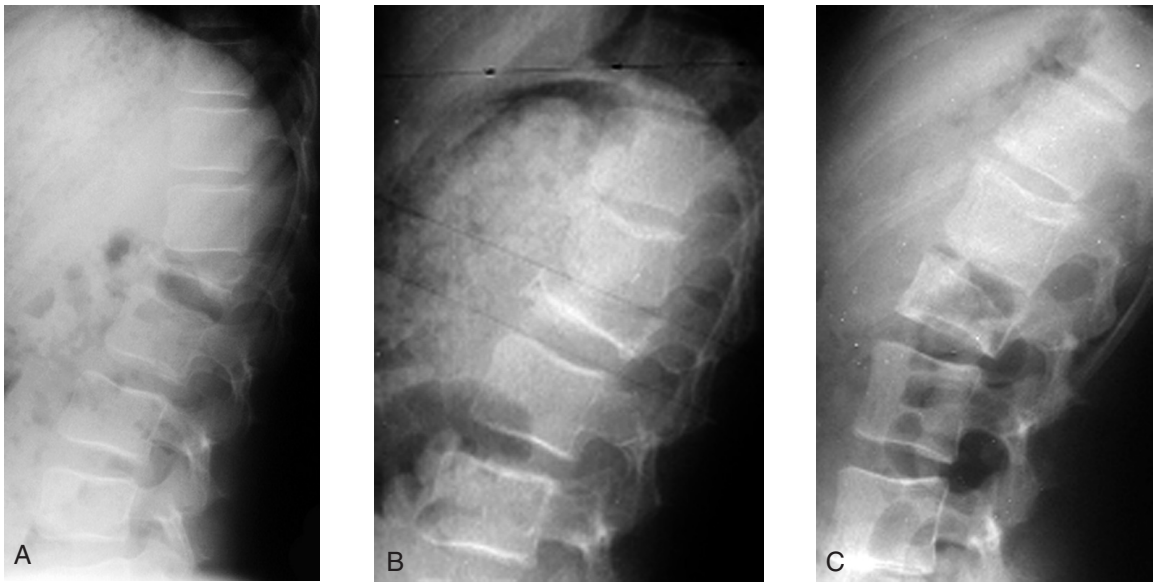

Fig. 4. Langerhans' cell histiocytosis of L2 in a 12 year-old girl. (**A**) X-ray showing characteristic vertebra plana. (**B**) X-ray two years following biopsy showing mild reconstitution of vertebral height. (**C**) X-ray three years following biopsy showing marked reconstitution of vertebral height.

ties have been used over the years to treat LCH of the spine including bracing, surgery, chemotherapy, radiation therapy, steroid injection, no therapy, and a combination of these methods. Single agent chemotherapy with prednisone or methotrexate is often utilized for multi-focal skeletal LCH or systemic LCH [44,45,46,47]. Radiation therapy is contraindicated due to the benign natural history of the disease and risk of post-radiation sarcoma.

We recommend biopsy (either percutaneous CT-guided or open with intraoperative frozen section) of the lesion if it is found to be LCH. In cases of multi-focal disease, only the most easily accessible site needs to be biopsied to make a diagnosis. Biopsy usually leads to rapid pain relief and regression of disease in nearly all cases. Furthermore, reconstitution of vertebral height (often to nearly normal levels) during the healing process makes the need for spinal fusion exceedingly rare [37,38,41]. Following biopsy most patients only need to be braced for a short period of time to prevent collapse or deformity while healing occurs. The treating surgeon needs to also be aware that additional late-onset lesions may appear. Long-term outcomes in LCH of the spine, regardless of treatment, are excellent [34,35,36,37,38,39, 40,41,42]. Therefore, following tissue diagnosis of this benign lesion and appropriate studies to identify multi-focal disease, only conservative management with close follow-up is needed to care for these patients appropriately.

Osteoid Osteoma/Osteoblastoma

Osteoid osteoma and osteoblastoma are felt to be on two ends of a continuum of disease and are histologically similar lesions. Traditionally, the dividing line between the two diagnoses has been an arbitrary measurement of size, with lesions greater than 15 mm (some use 20mm) in diameter called osteoblastoma, and smaller lesions osteoid osteomas. Although they have similar histology, osteoid osteomas tend to evoke a more sclerotic reaction while osteoblastomas are usually more expansile lesions [48]. These lesions have a very characteristic pattern of pain, with night pain being a very significant feature (especially in osteoid osteoma). Another diagnostic clue is that aspirin and other nonsteroidal anti-inflammatory agents provide very rapid and effective pain relief (this is more prominent for osteoid osteoma than osteoblastoma). Many times small lesions are not readily identified on x-ray, however, with the characteristic clinical picture and a negative x-ray, a bone scan should be obtained (Fig. 5). Bone scan is nearly 100% reliable for picking up osteoid osteoma and osteoblastoma [49]. In cases of osteoblastoma, chest x-ray is recommended to rule out pulmonary metastasis that will occasionally be present subclinically at diagnosis of the spine tumor.

Both tumors have a heavy predilection for the posterior elements of the spine. Overall, 10% of osteoid osteomas and 40% of osteoblastomas occur in the spine [5,48]. Scoliosis is often a significant component of the disease process; 40% of children with spinal osteoblastoma have scoliosis at presentation (Fig. 6). The curve usually has its apex at the level of the lesion, which is almost always on the concavity of the curve [48,50,51]. Unlike the scoliosis caused by destructive tumors, in osteoid osteoma and osteoblastoma, the scoliosis is felt to be caused by inflammation, irritation, and spasm of paraspinal muscles. These findings have been identified on advanced imaging of the spine and paraspinal region in patients with these tumors [18]. Pettine demonstrated that scoliosis present for less than 15 months before diagnosis usually resolves with treatment of the underlying osteoid osteoma or osteoblastoma while scoliosis present for longer than 15 months does not usually resolve on its own.

In preparation for surgery, CT scans with fine-cuts (3mm) are extremely valuable in determining the extent of the

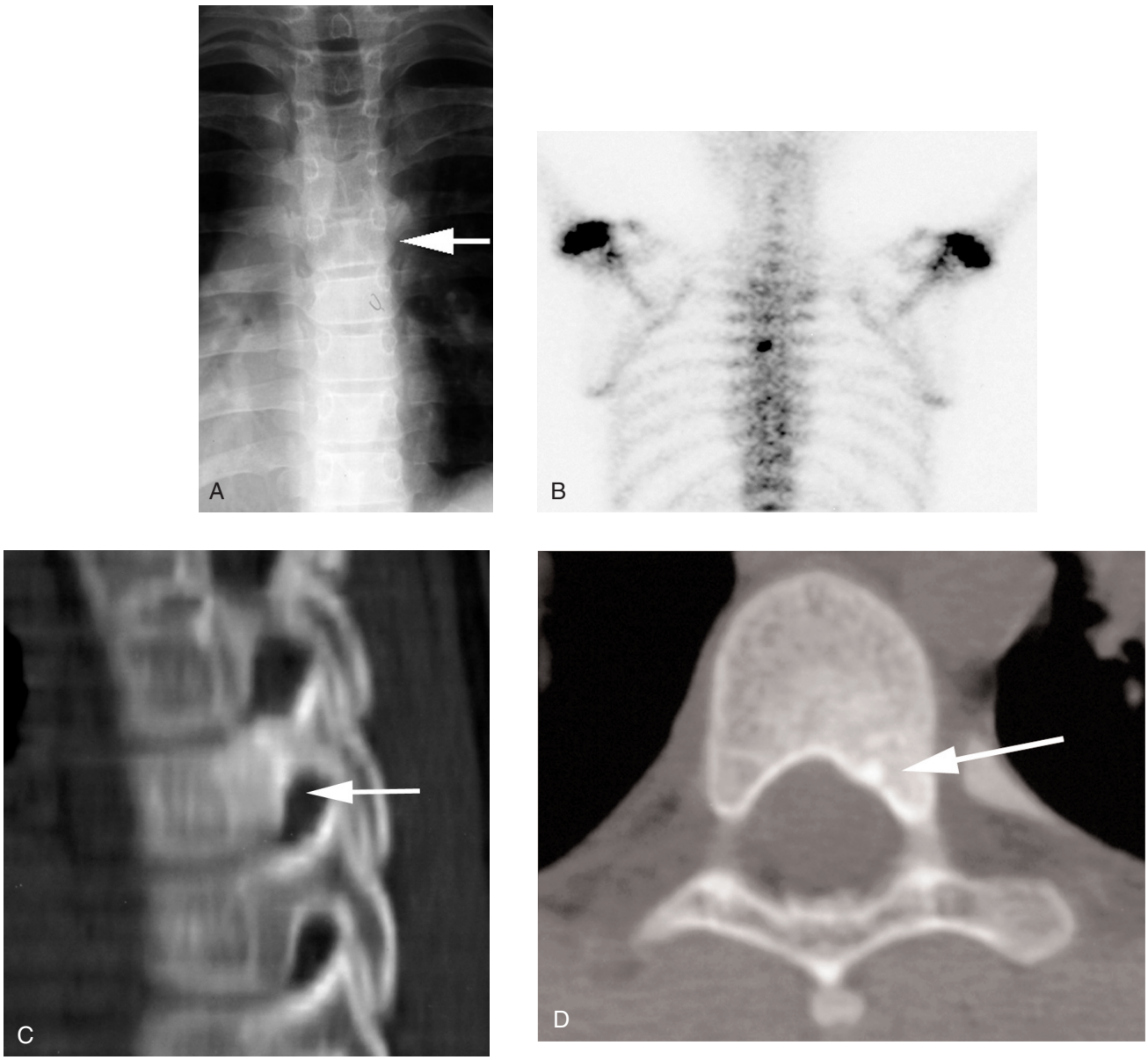

Fig. 5. Osteoid osteoma of T6 in a 12 year-old boy. (**A**) Normal appearance of T6 (arrow) on x-ray obtaining following the complaint of back pain. (**B**) Bone scan showing increased uptake of tracer at T6. (**C**) Sagittal CT revealing nidus (arrow) in posterior elements of T6. (**D**) Axial CT showing small nidus (arrow) in left lateral mass of T6.

lesion. In cases of osteoid osteoma, although the natural history of the tumor is to burn out and resolve on its own, most patients are unwilling to simply wait and take an extended course of non-steroidal anti-inflammatory agents for pain relief. With biopsy and curettage (which often can be performed percutaneously under CT guidance) there is usually almost total immediate pain relief if the nidus of disease is removed. Osteoblastomas are usually larger lesions which require open procedures for adequate excision. Intralesional margins have been shown to have recurrence rates of 10% [52]. In Enneking stage 3 lesions extended curettage and adjuvant phenolization is recommended while intralesional curettage of the nidus is acceptable for stage 2 lesions. Radiation has in the past been used for cases in which complete surgical excision of the nidus is not possible, however, is now contraindicated [4]. Some authors argue that radiation is not even effective, and in our practice, we discourage use of adjunct radiation therapy for osteoid osteoma and osteoblastoma.

Osteochondroma

Osteochondromas, while very common benign skeletal tumors, rarely appear in the spinal column. The frequency of isolated spinal lesions is estimated at 1–4% of all osteochondromas; spinal lesions as part of multiple hereditary exostoses are slightly greater in number [5,53]. Osteochondromas are thought to be caused by continued endochondral ossification of misplaced cells from the growing epiphyses of bone. Pathologically they appear as hard bony lesions with cartilage caps, often giving a “cauliflower” appearance.

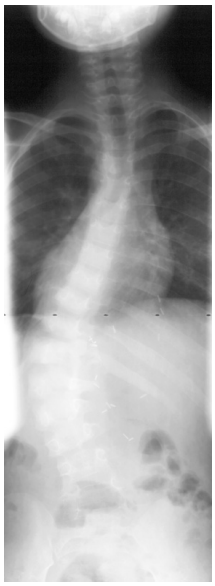

Fig. 6. Presentation of osteoblastoma of T11 in a 7 year-old girl as a 48 degree scoliosis

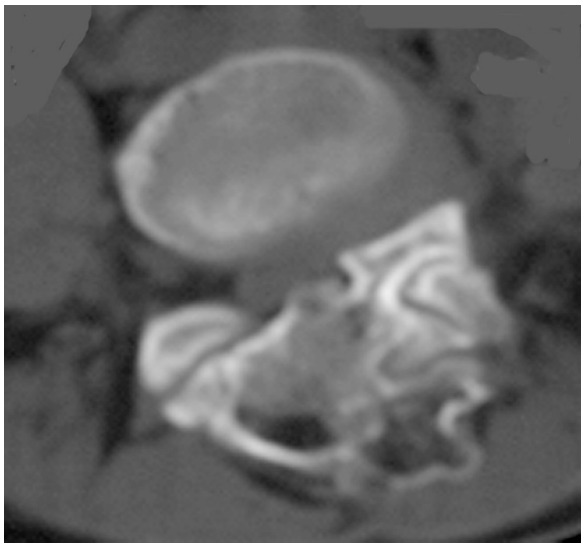

Fig. 7. MRI revealing an osteochondroma of L5 in an 8 year-old girl showing invasion of the canal and compromise of L5 nerve roots.

They are benign lesions which grow in tandem with the growth plate—aggressive growth or growth beyond skeletal maturity are concerning signs for chondrosarcoma. In multiple hereditary exostoses (MHE), there is an autosomal dominant gene mutation that leads to osteochondromas throughout the skeleton that are usually too numerous to count. Lesions in MHE also may have malignant degeneration into a chondrosarcoma, a process seen particularly in axial locations.

In the spine, osteochondromas are usually found in the posterior elements and can compress the spinal cord (Fig. 7). They usually are painless until they increase in size enough to compress the spinal cord and nerve roots to cause neurologic symptoms. Indeed, neurologic symptoms are often what bring these lesions to medical attention. Barring neurologic symptoms, osteochondromas will only become clinically apparent if they become large enough to form a palpable mass. These lesions usually have a highly characteristic appearance on x-ray, CT, and MRI—revealing the bony exostosis covered by a cartilage cap. Biopsy is not necessary if imaging clearly points to osteochondroma, however, lesions growing rapidly, growing after spinal growth has ceased, or which cannot be confidently diagnosed on imaging should be biopsied. Indications to remove an osteochondroma of the spine are similar to those for the skeleton in general. We recommend surgical excision for lesions that are painful, compromising neurovascular structures, or are found to be malignant following biopsy. There is a 5–10% recurrence rate over the first twelve months for osteochondromas following excision; therefore, patients should be followed closely with serial radiographs following surgery.

Leukemia and Lymphoma

These are primary malignancies of the hematologic and immune system that will occasionally sprout up in the spinal column. Their radiographic appearance will include vertebral fracture and vertebral destruction (at times with vertebra plana) (Fig. 8). Leukemia is the most common type of malignant tumor encountered in the pediatric population. Symptoms are generally non-specific and include fatigue, malaise, fever. Pain usually is found only after pathologic fracture. Biopsy is not usually necessary in the diagnosis of leukemia and lymphoma since blood work (lymphocytosis, abundant abnormal blasts on blood smear) usually points to the diagnosis. Occasionally with lymphoma biopsy is needed to make the diagnosis.

Orthopaedic surgeons are sometimes asked to be involved in the primary management of children with leukemia or lymphoma involving the spine. A pediatric oncologist usually manages treatment with the orthopaedic surgeon's role mostly confined to treating pathologic fractures of the spine and preventing spinal deformity during the course of treatment. Survival is varied and depends on the specific nature of the underlying tumor.

Ewing's Sarcoma

Ewing's sarcoma is one of the small round blue cell tumors of unknown origin. Unique features of Ewing's sarcoma are a characteristic chromosome 11:22 translocation as well as specific immunohistochemistry staining with *HBA-71*. Recently it has been suggested that primitive neuroectodermal tumors and Ewing's are one in the same. Ewing's sarcoma is highly aggressive and frequently fatal. It makes up approximately 10% of all malignant musculoskeletal tumors and frequently involves the spine. Most patients are diagnosed before the age of twenty. Dahlin's series found about 10% of all Ewing's sarcomas to be located in the spine with a heavy predominance in the lumbosacral region [5]. The main clinical symptoms are usually pain; 60% will have neurologic symptoms, 25% of patients have a tender palpable soft-tissue mass [4,54]. ESR may be elevated in 50% of cases, but is very non-specific. They can often grow quite large in size before diagnosis. Although sometimes confused with neuroblastoma on imaging, the

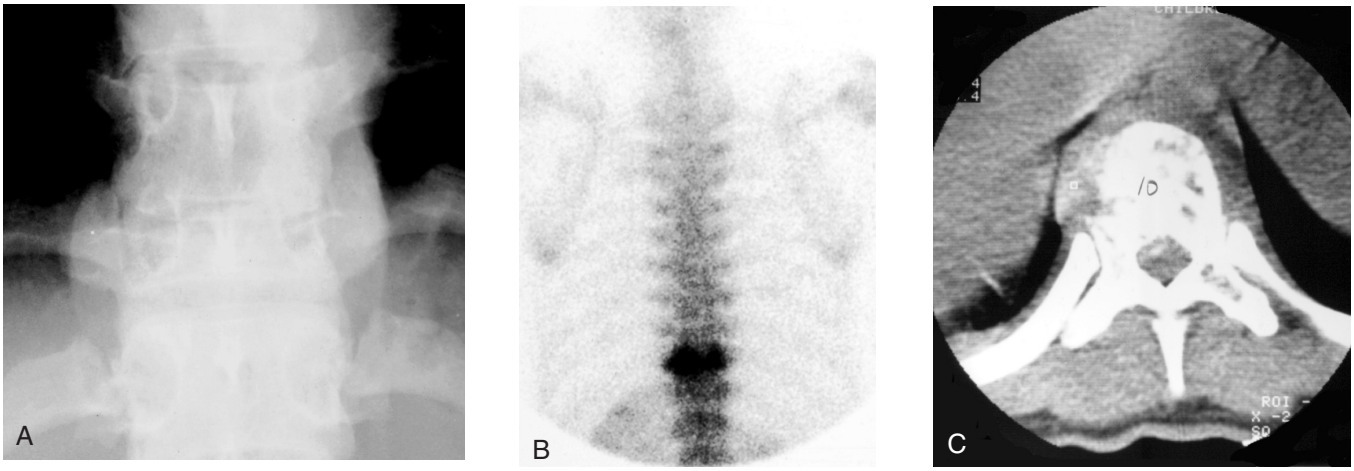

Fig. 8. Lymphoma of T10 in a 17 year-old girl. (**A**) X-ray showing vertebral collapse. (**B**) Bone scan showing increased uptake of tracer at T10. (**C**) CT scan showing destruction of vertebral body by tumor.

distinction is easily made following urinary analysis that can confirm or reject neuroblastoma as diagnosis. A more difficult distinction often occurs between Ewing's sarcoma and other lesions that lead to vertebral destruction. Often a plain x-ray will show only vertebral destruction; MRI is necessary to further classify the lesion (Fig. 9). The differential diagnosis of an aggressive appearing spinal lesion includes metastatic disease and osteosarcoma. Unlike long bone Ewing's, "onion-skinning" periosteal reaction is not usually seen in the spine. As has been emphasized, however, tissue diagnosis is the only definitive way to make a correct diagnosis. Frequently Ewing's will be suspected with an aggressive appearing lesion on MRI, and as such the biopsy must be planned with definitive surgical resection in mind. The biopsy tract must be removed with the tumor specimen for a successful surgery [4,5,55].

Treatment of Ewing's sarcoma is very difficult and requires the coordination of multiple medical specialties.

Fig. 9. Ewing's sarcoma of T10 in an 8 year-old boy. (**A**) X-ray showing collapse of vertebral body. (**B**) CT scan showing destruction of vertebral body and large soft-tissue mass. (**C**) MRI revealing extent of tumor into the spinal canal.

Given the often large size at diagnosis, chemotherapy and radiation therapy are usually first-line therapy. The goal of chemotherapy is cytoreduction to make surgical resection with wide margins possible. Post-operative chemotherapy is universal. Metastasis can complicate the picture (25% of patients have metastases at presentation), as Ewing's often will spread to the lungs; CT scan of the chest is recommended for all children with suspected Ewing's sarcoma. Survival for Ewing's has improved recently due to the advent of modern surgical technologies that allow spondylectomy and en bloc resection of these large lesions while still preserving spine stability. Two large series of Ewing's sarcoma of the spine, with 33 and 36 patients, had five-year survivals of 48.1% and 33% [54,56]. Greatest tumor diameter less than 8 cm was a favorable risk factor with five-year survival of 57.7% compared to only 10% for tumors greater than 8 cm [54].

Osteosarcoma

Although it is the most common primary malignant tumor of the musculoskeletal system (excluding multiple myeloma), osteosarcoma only rarely affects the spine. Approximately 2–3% of all osteosarcomas arise in the spine, with a slight predilection for the lumbosacral region. Pulmonary metastases are common (10-20%) in osteosarcoma [4,57]. Pain is uniformly present at diagnosis and 70% have neurologic symptoms [58,59]. A soft-tissue mass may also be appreciated. The medical history is important as a history of retinoblastoma leads to a greater than 2000 fold increased risk of later developing osteosarcoma [60]. Plain x-rays usually will reveal vertebral destruction and a soft-tissue mass. The expansile mass will often have hazy and diffuse calcification indicative of the ossifying nature of the tumor. This is often sufficient to make a confident diagnosis of osteosarcoma. Advanced imaging is useful to determine anatomic extent of disease and plan for biopsy and surgery.

As with all regions, survival with spine disease is poor, with five-year rates between 3–10% [58,59]. The advent of modern chemotherapy regimens along with advances in surgical technology is beginning to increase survival. In a similar fashion to treatment of Ewing's sarcoma, medical management is usually first-line therapy with the goal of cytoreduction. Surgery is usually deferred until the lesion has reduced in size to allow resection with appropriate margins. The biopsy tract must be removed en bloc to avoid leaving behind residual tumor. Surgical treatment of pulmonary metastases has been shown to be feasible and effective in improving survival [57]. Chemotherapy regimens often continue following surgery and bone scans can be used as surveillance for recurrence of disease.

Conclusion

Thankfully most primary tumors of the spine in children are benign, often with a self-resolving natural history. Malignant tumors, on the other hand are extremely challenging to manage and require close coordination between general pediatric, oncology, general surgery, and orthopaedic surgery teams. Modern surgical technologies and techniques in tandem with advances in medical oncology have dramatically changed the prognosis for children with malignant spine tumors. Since they are a rare occurrence, most experience in managing primary tumors of the spine comes from specialized musculoskeletal centers such as ours. By collecting and sharing our experience with these tumors we hope to advance the care of children with tumors of the spine throughout the world.

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