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Pediatric Tumors Involving the Spinal Column

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The most common extradural spine tumors involving the spine in children are those masses that grow into the epidural space and vertebral body either locally or from metastatic spread, followed by primary bone tumors that originate in the bony spine. Because approximately 3% to 5% of children who have systemic cancer develop spinal cord compression, primarily as a result of mass in the epidural space [1,2], there is a large differential of histology regarding extradural childhood spinal tumors. Children who have solid malignant tumors outside the spine have been shown to develop spinal metastases in up to 5% of patients [1], and tumors of the hematopoietic system, including leukemias and lymphomas, not uncommonly seed the epidural space [3]. Tumors that originate in the paraspinal soft tissues and locally invade the spine include ganglioneuromas and neuroblastomas, which are both derived from the paraspinal autonomic system. Soft-tissue sarcomas likewise can invade the epidural space by way of direct extension or metastatic spread. Finally, tumors originating from the bony spine itself consist of a wide range of pathologies, including osteoid osteomas, osteoblastomas, osteochondromas, aneurysmal bone cysts (ABC), Langerhans cell histiocytosis, and Ewing' sarcoma.

Epidemiology

Cancer is the second largest cause of death in children younger than 15 years of age in North America [4]. Although approximately 50% of childhood malignancies are hemopoietic in origin, 20% of childhood cancers involve the central nervous system (CNS) [4]. Compared with adults, the overall incidence of tumors of the spine and spinal cord in children is less, and the incidence of tumors at specific locations in the spine of children differs from the distribution in adults. Specifically, in children approximately 40% of spinal tumors are intramedullary, 20% are intradural, and 40% are extramedullary [5]. In contrast, in the adult population 20% are intramedullary, 60% are intradural, and 20% are extradural [6,7]. The most common histopathologies of childhood spine tumors are astrocytoma, followed by sarcoma, neuroblastoma, ependymoma, dermoid, and teratoma. Medulloblastoma, neurofibroma, metastatic carcinoma, and lipoma each account for 5% of tumors [3]. Primary tumors of bone are even less common.

Clinical presentation

The most common presenting symptom of extramedullary spinal tumors in children is pain, reaching 96% in some series [8]. In reviewing the history of the patient presentation, certain features of spine pain may suggest an underlying tumor rather than a non-neoplastic diagnosis. Tumor-related pain more often (1) is focal, (2) occurs at night, (3) wakes the child from sleep, (4) increases in intensity over time, and (5) is unrelated temporally to activity. A warning sign for locally aggressive or malignant tumors is rapid increase in intensity of pain over a short period or any pain of severe intensity [9]. If the patient is very

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young, however, especially in the preverbal age group, pain may not be effectively expressed. As a result, pain may manifest as irritability or refusal to ambulate [3]. Because of such potential limitations in communication, diagnosis is often delayed in young children until more obvious neurologic deficits appear, and thus tumor growth may be more advanced at the time of diagnosis [10–12]. In a review of pediatric intraspinal tumors, Raffel and Edwards [7] found that weakness was a predominant presenting symptom in more the 67% of cases of spine tumor in children.

Other signs attributable to a spine mass may also be common, including reflex changes (49%), sensory changes (30%), abnormal spinal curvature (28%), muscle atrophy (19%), tenderness (14%), spasms (13%), paraspinal mass (12%), and torticollis (8%) [7]. Not surprisingly, children who have regional pain syndromes often have significant clinical testing that does not include the spine. For instance, thoracic spine pathology leading to referred pain from nerve root compression not uncommonly leads to misdirected investigations for abdominal complaints [13]. For this reason, cervical tumors should also be considered in the differential for childhood neck stiffness and head tilt.

Spinal deformity develops in up to 40% of children who have spine tumors, likely as a result of a muscular response to pain or asymmetric vertebral destruction [9]. In patients who have osteoid osteoma and osteoblastoma, scoliosis is believed to be a muscular response to pain [14]. The apex of the curve is generally at the level of the tumor, and the lesion is usually located on the convex side of the deformity [9]. Deformity caused by asymmetric vertebral destruction is more common with Langerhans cell histiocytosis (eosinophilic granuloma), osteosarcoma, or other lytic bone tumors.

Neuroblastic tumors

Neuroblastic tumors are embryonal tumors derived from neural crest cell precursors and include neuroblastoma, ganglioneuroblastoma, and ganglioneuroma. Neuroblastomas are considered the most poorly differentiated and most malignant, whereas ganglioneuromas are the most differentiated and most benign. Including all pediatric malignancies, neuroblastomas are the fourth most common pediatric tumor, accounting for 8% of all pediatric cancers, and they are the most common cause of extradural spinal cord compression in childhood [15]. Most neuroblastomas occur in children younger than 3 years of age (75%) and are rare after the age of 10 years [16]. Because they are believed to originate from embryonic neural crest precursor cells that differentiate into sympathoblasts, most masses are located within the adrenal gland or the paraspinal sympathetic chain [3]. Location is primarily abdominal (40% adrenal, 25% paraspinal), followed by thoracic (15%), pelvic (5%), cervical (3%), and elsewhere (12%). Infants are more likely to present with thoracic or cervical tumors, whereas older children present more frequently with abdominal masses [16].

Histologically, neuroblastomas are composed of densely packed small round cells in a rich fibrovascular stroma. Using electron microscopy, they possess a characteristic of dense neurosecretory core granule in the cytoplasm. These granules contain the catecholamines vanillylmandelic acid (VMA) and homovanillic acid (HVA), which are elevated in the serum and urine of at least 65% of patients [3]. The differential diagnosis based on histology includes Ewing' sarcoma, lymphoma, rhabdomyosarcoma, and Askin' tumor. In very young children, neuroblastomas may regress into more histologically mature ganglioneuromas [16,17].

Clinical presentation in children often involves development of an abdominal or thoracic paraspinal mass with or without accompanying scoliosis. Paraplegia secondary to cord compression is not uncommon, occurring in approximately 10% of patients [18–20]. In 11 patients who had spinal canal involvement, Armstrong and colleagues [21] found neural symptoms in only 55%. Children who have neuroblastomas may also exhibit paraneoplastic syndromes with an incidence of 2% to 3%, the most notable being opsoclonus-myoclonus-ataxia syndrome, believed to result from the production of antineuronal antibodies [22,23].

On imaging, lesions can be variable in diameter (1–10 cm), and their morphology is often of a dumbbell shape because of the combined paraspinal and intraspinal location. CT scans often reveal an enhancing paraspinal mass with or without epidural extension and finely stippled calcifications present in 85% of patients [24]. The neural foramina and intercostal spaces may be widened by pedicle erosion, and the associated ribs may be splayed. MRI usually reveals a hypo- or isointense paraspinal mass on T1-weighted imaging (T1WI) and a hypo- to hyperintense paraspinal mass on T2-weighted imaging (T2WI)

(Fig. 1). Although MRI is considered the best imaging tool for diagnosis and presurgical planning, nuclear medicine techniques are important for staging and post-treatment surveillance. Specifically, metaiodobenzylguanidine (MIBG) uptake by sympathetic catecholaminergic cells can be sensitive [25]. Bone marrow involvement is common, and bone marrow aspirate may be recommended to confirm the diagnosis and for staging.

The treatment of spinal neuroblastoma involves a combination of modalities, including chemotherapy, surgery, radiation, and steroids. For strictly localized tumors, surgery may be the optimal treatment [26,27]. If disease is disseminated at presentation, however, initial treatment with chemotherapy is used and has shown success [28]. De Bernardi and colleagues [20] treated 77 patients who had spinal cord compression from a neuroblastoma using radiation therapy in 11, surgery in 32, and chemotherapy in 33. Surgery was used as the primary treatment modality, consisting of laminectomy, mainly for patients who had a favorable prognosis and in those who had severe motor deficits. Unfortunately none of the patients who had severe motor deficits (paraplegia) recovered or improved. In all other patients who did not possess serious motor deficits, neurologic outcome was similar, regardless of modality (surgery, chemotherapy, radiation therapy). Nonetheless, complete surgical resection should be attempted in cases of localized disease in an effort to cure the patient. In such cases, laminectomy alone is seldom adequate to achieve complete resection, and thus lateral, anterior, or

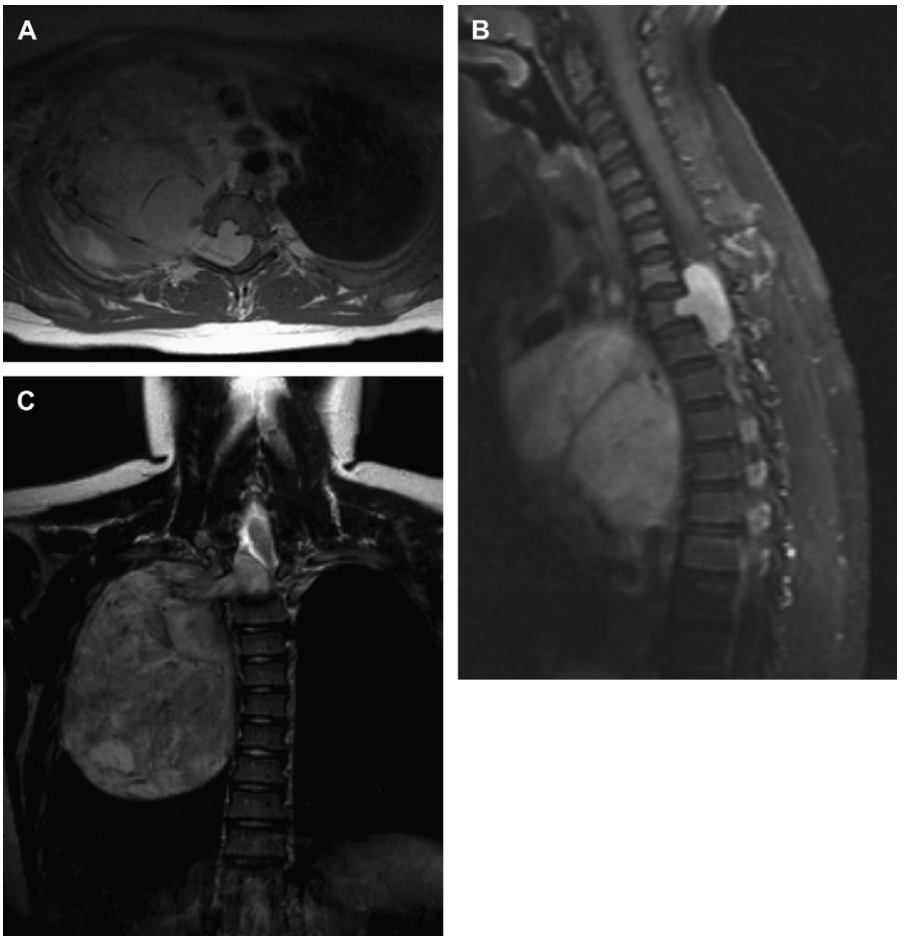

Fig. 1. T1-weighted MR images with gadolinium of a classic dumb-bell–shaped neuroblastoma with significant extension into the chest cavity and spinal canal shown in (*A*) axial, (*B*) sagittal, and (*C*) coronal planes.

combined approaches should be considered to optimize oncologic treatment and to achieve adequate decompression of the cord.

Overall the 5-year survival rate is approximately 83% for infants, 55% for children 1 to 5 years of age, and 40% for children older than 5 years [16]. Favorable prognostic factors include focal disease, younger age, histologic maturity with predominance of ganglion cells, low n-myc amplification, and hyperdiploid DNA [3,16]. Unfavorable prognostic indicators include stage 4 disease, ocular involvement, HER2/neu oncogene overexpression, increased NSE/LDH/serum ferritin, increase urine HVA/VMA, and 1p chromosomal deletion [20,29–31].

Sarcomas

Ewing' sarcoma

Of the sarcomas that may involve the pediatric spine, Ewing' sarcoma is the most common. They account for 6% of bone tumors, of which 5% to 10% originate in the spine, affecting approximately 3 in 1,000,000 Caucasian children younger than 15 years of age [32–35]. These tumors classically involve the vertebral body before the neural arch but may originate in soft tissues, and more than half of these tumors occur in the sacrum, followed by the lumbar region [34]. Spine involvement most often occurs in the second decade of life with a predilection for males, such that approximately 90% of all patients who have Ewing' sarcoma present before 20 years of age [35]. They are rare in patients of Black African and Chinese descent [36]. Pathologically these tumors are considered to be undifferentiated mesenchymal cells with a slight differentiation toward neuroectodermal cells [35]. On histologic preparation, these tumors are the prototypical nonhematologic small round cell tumor, not dissimilar to primitive neuroectodermal tumor (PNET) histology. Macroscopically these tumors are gelatinous, cystic, necrotic, and often hemorrhagic. The transposition chromosomal abnormality t(11:22) is also typical for Ewing' sarcomas [3].

The most common clinical presentation of a spinal Ewing' sarcoma is localized pain and neurologic symptoms ranging from radiculopathy to paraplegia. Systemic manifestations, such as fever and weight loss, which may be accompanied by leukocytosis and an elevated erythrocyte sedimentation rate (ESR), occur with these lesions, leading some to the erroneous early diagnosis of osteomyelitis. Imaging classically shows "moth-eaten" destruction of bone centered in the vertebral body or sacrum, and up to 50% of patients may possess an extraosseous, noncalcified, soft-tissue mass. Such lytic bone destruction may lead to vertebra plana, but unlike osteomyelitis disc height is generally preserved [35]. With MRI the paraspinal mass can be more easily delineated, characteristically exhibiting low intensity on T1WI, high intensity on T2WI, and moderate enhancement on contrasted studies (Fig. 2) [35].

Surgery is often necessary for diagnosis and decompression of the neural elements. Fifteen percent to 50% of patients have evidence of microscopic metastatic spread at presentation, however, involving lung, lymph nodes, and other skeletal sites [32,37]. In addition, prognosis is worse with spinal Ewing' sarcoma compared with peripheral disease, likely because of the difficulty of complete resection in this area [35]. Surgery or radiotherapy without chemotherapy has been shown to be universally fatal. The introduction of multimodality treatment has thus largely contributed to the improved survival rates in patients in the past 3 decades [32,37,38], and recent studies have shown disease-free survival rates of 49% and 36% at 5 and 10 years, respectively [39]. Because most patients who have spinal Ewing' sarcoma exhibit evidence of spinal cord compression, decompressive surgery is often required.

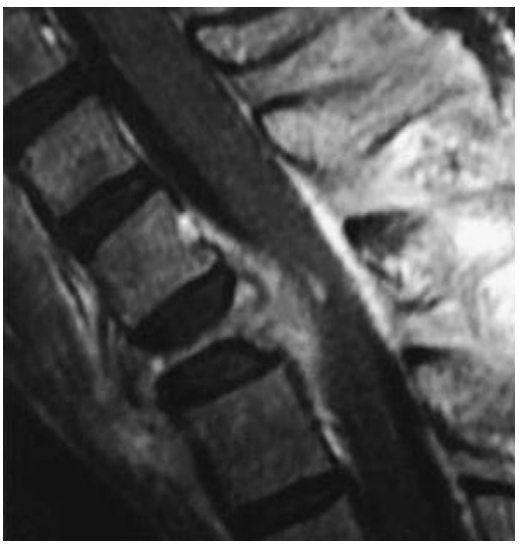

Fig. 2. Contrast-enhanced T1-weighted MRI of the thoracic spine in the sagittal plane showing vertebra plana and epidural extension of tumor.

If possible, however, a chemotherapeutic protocol should be started before surgery. The goal of surgery should be to obtain a wide, radical resection. Radiation therapy is then used for surgically inaccessible lesions, residual disease, or a poor response to chemotherapy. Patients who have localized disease at presentation, axial tumors, and age younger than 15 years have the best prognosis [38]. Local recurrence, metastatic spread, treatment complications, and secondary malignancies are not uncommon [35].

Rhabdomyosarcoma and other sarcomas

Another sarcoma involving the trunk and retroperitoneal space in children that can involve the spine is rhabdomyosarcoma [40,41]. Like other sarcomas, spinal involvement may result from direct extension or from metastatic spread. When feasible, aggressive surgical resection is recommended, because local control seems to be the most important prognostic factor in children [40]. Nerve sheath tumors such as neurofibrosarcomas account for approximately 10% of non-rhabdomyosarcomas in children. Although the primary goal of treatment for these aggressive tumors is aimed at complete excision, adjuvant chemotherapy may be of benefit. Other sarcomas include synovial sarcomas, hemangiosarcomas, leiomyosarcomas, and malignant fibrous histiocytomas [3]. Patients who had previous radiation for pediatric tumors can also develop radiation-induced sarcomas that may involve the spine and sacrum. Previous irradiation of the femur and pelvis can lead to significant lumbosacral sarcomas and should not be mistaken for recurrence of the original tumor.

Primary tumors of bone

Primary bone tumors are uncommon in children. They include osteoid osteomas, osteoblastomas, osteochondromas, ABC, and Langerhans cell histiocytosis (LCH, eosinophilic granulomas).

Osteoid osteoma and osteoblastoma

Osteoid osteomas and osteoblastomas are benign tumors, and they comprise approximately 3% and 1%, respectively, of all benign bone tumors [42,43]. They are histologically identical, and differentiation is based on size, with osteoid osteomas defined by size smaller than 1.5 cm. They overwhelmingly involve the neural arch and only rarely involve the vertebral body. Most of these tumors are encountered in the second decade of life. Approximately 10% of all osteoid osteomas occur in the spine [44]. Location is primarily lumbar (59%), followed by cervical (27%), thoracic (12%), and sacral (2%) [44]. Patients who have osteoid osteomas in the spine usually present with a painful scoliosis (70%) with the concavity of the curvature contralateral to the tumor. Approximately half of patients have pain at the site of the lesion and half have radicular pain [3]. Gait disturbance, muscle atrophy, and torticollis can also be seen. A nocturnal increase in pain is typical and is classically relieved by aspirin or nonsteroidal anti-inflammatory medications (NSAIDs). Because smaller lesions can be missed, ideal imaging is bone CT with 1-mm helical sections. Findings include an isolated radiolucent area, termed the central nidus, surrounded by reactive sclerosis. MRI reveals a hypo- or isointense nidus and a surrounding zone on T1WI and T2WI. Radionucleotide bone scan is also a reliable screening method, revealing a well-demarcated area of increased uptake [45].

Although less common than osteoid osteomas, a higher proportion of osteoblastomas occurs in the spine (40%) compared with lesions elsewhere in the skeleton. They originate in the posterior spinal elements with a predilection for the base of the transverse process [3]. They are most commonly found in the cervical spine (40%), followed by the lumbar (25%), thoracic (20%), and sacral regions (15%–20%) (Fig. 3) [44]. Like osteoid osteomas, osteoblastomas are composed of prominent osteoblasts and osteoclasts in a fibrovascular stroma of woven bone. Osteoid osteomas, however, tend to have a more benign course, whereas osteoblastomas tend to progress if left untreated [42,45–47]. Surgery for decompression or to relieve pain consists of local removal by intralesional resection. CT-guidance may be beneficial for small osteoid osteomas, and preoperative embolization may be helpful for large osteoblastomas. Partial removal often suffices, but if the entire nidus is not removed, recurrence is possible for both entities. For typical osteoblastomas, recurrence is approximately 10% to 15%, and for aggressive osteoblastomas, recurrence can be as high as 50% [44].

Osteochondroma

Osteochondromas are cartilage-covered osseous protuberances that can occur anywhere in the skeleton. Less than 5% occur in the spine, but when present, they classically arise from the

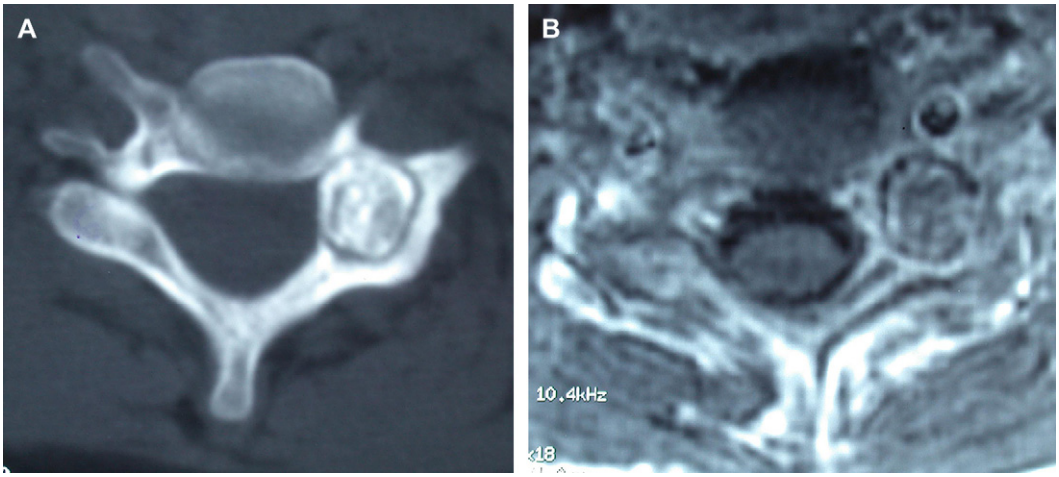

Fig. 3. Axial images of a cervical osteoblastoma showing central nidus and surrounding sclerotic rim on (*A*) CT and (*B*) MRI.

spinous or transverse processes. Most occur in the cervical spine, with C2 as the most often involved level. The peak age of such lesion is between 10 and 30 years [48]. Most patients present with a painless, palpable mass; however, radiculopathy, scoliosis, and dysphagia or hoarseness secondary to a pharyngeal mass may also occur. Patients who have hereditary multiple exostoses (HME), also known as familial osteochondromatosis, are prone to the development of such lesions. Most patients who have HME are diagnosed by 5 years of age, and virtually all are diagnosed by 12 years of age [48]. The ideal imaging modality is MRI, which shows a hyperintense cartilaginous cap on T2WI that forms a mushroom-like shape over an osseous pedicle (Fig. 4). Lesions can be removed if symptomatic, with a local recurrence rate of less than 2%. Although malignant transformation to chondrosarcoma occurs in less than 1% of solitary lesions, the incidence increases to 3% to 5% in patients who have HME [48].

Aneurysmal bone cyst

Aneurysmal bone cysts are benign expansile lesions containing thin-walled, blood-filled cavities of unknown origin, accounting for approximately 1% to 2% of primary bone tumors [49]. Approximately 80% of such lesions occur in patients younger than 20 years of age [49]. They frequently present as expanding solitary cystic lesions that are composed of a fibro-osseous network and giant cells. On CT and MRI these lesions appear as expansile, multiloculated, neural arch masses with fluid–fluid levels, the latter produced as a result of intralesional hemorrhages (Fig. 5). Local pain, most severe at night, has been reported as the initial symptom in more than 70% of patients with or without scoliosis, whereas approximately 20% report a palpable mass [50]. Spinal cord and nerve root compression are not uncommon, with 29% and 18% incidences, respectively [3]. Furthermore, neurologic symptoms or deficits can suddenly worsen with acute expansion of the lesion [51].

Treatment consists of embolization or surgical excision. Because of the prominent vascularity, profuse hemorrhage can be encountered in surgery or even during percutaneous biopsy. To prevent hemorrhage, selective angiography and embolization have been advocated as preoperative treatment [52–54]. Recurrence rates can approach 30%, especially with incomplete surgical resection [50,52]. In addition, spinal instrumentation may be required, depending on the extent of the bony resection. Radiation therapy is reserved for lesions that cannot be resected completely [50–52], but it may predispose to radiation-induced sarcoma [49].

Langerhans cell histiocytosis/eosinophilic granuloma

Although considered an inflammatory, granulomatous condition that may affect the spine, LCH should be considered in the differential diagnosis in children who have spine lesions.

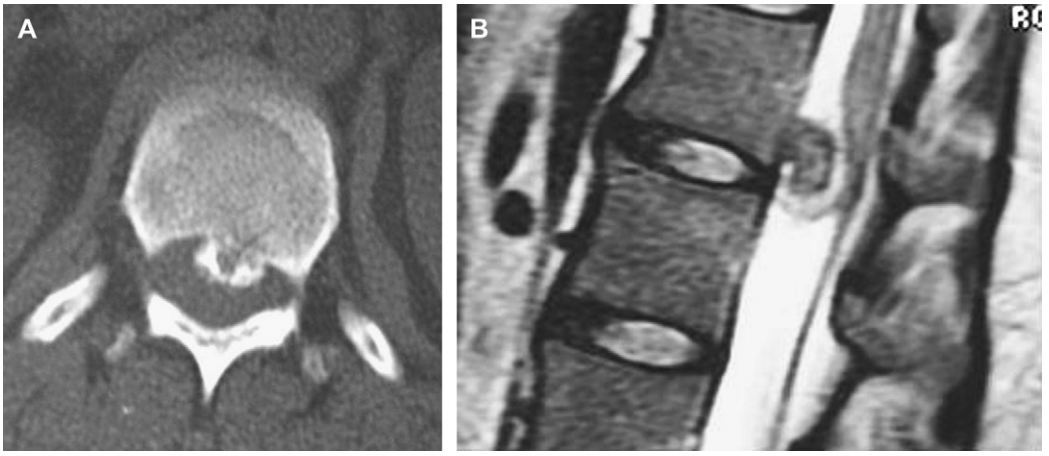

Fig. 4. Vertebral osteochondroma shown on axial (*A*) CT and (*B*) sagittal T2-weighted MRI.

They occur far more commonly in children compared with adults, and the clinical presentation can be dramatic. These lesions are destructive, lytic masses that may create pathologic fractures of the bony spine with resulting vertebra plana or may lead to soft-tissue extension into the spinal canal. The ideal imaging tools include MRI, which can evaluate epidural extension, and CT, which can assess bony destruction. Treatment is generally conservative with initial bracing, because lesions usually regress after 3 months and resolve by 2 years [55]. In patients

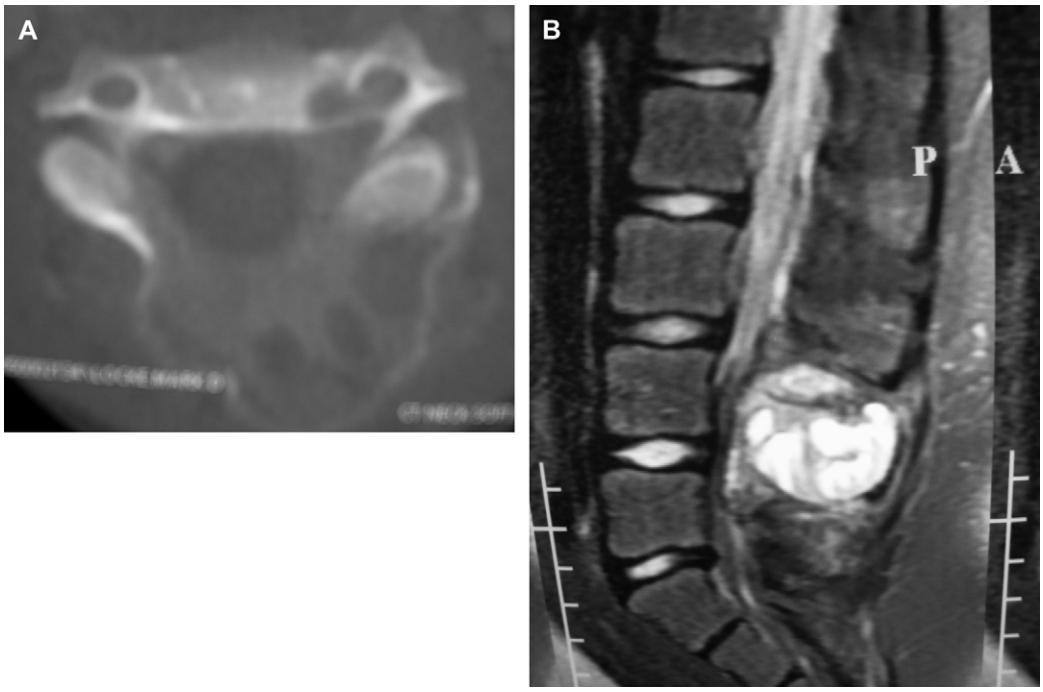

Fig. 5. Images of aneurysmal bone cysts (ABC). (*A*) CT image of cervical ABC showing expansile lesion with characteristic "egg-shell" of remaining cortical bone. (*B*) Sagittal T2-weighted MRI image of lumbar ABC showing large cystic spaces within lesion.

who have conservative treatment failure regarded as significant bone destruction or extensive growth of lesion, however, multimodality treatment can be instituted, involving intralesional surgical resection with or without fusion, radiation therapy, and chemotherapy [55].

Chordomas

Chordomas are tumors believed to arise from notochordal remnants in the vertebral body. Such lesions are rare in children, especially those younger than 10 years of age. In patients younger than 25 years, approximately one third of lesions are located at the skull base (clivus), one third in the mobile spine, and one third in the sacrum [56]. In their review of chordomas in children, Coffin and colleagues [57] found half the tumors had atypical features, possibly explaining the aggressive local growth sometimes seen in this population [3].

Imaging typically shows lytic destruction on CT with extraspinal extension of a soft-tissue mass on MRI. The physaliphorous cells that constitute most chordoma tissue appear hyperintense on T2WI. Neurologic deficits are related to progressive epidural compression of the spinal cord or cauda equina. The recommended treatment consists of radical surgical excision because of the high rate of local recurrence [58–61]. If complete resection is infeasible, however, radiation therapy may be used as adjuvant therapy. Although standard photon radiation therapy has limited efficacy because of the radioresistant properties of this tumor [62,63], improvements in intensity-modulated radiation therapy (IMRT) photon and proton therapy have shown moderate benefit [64,65].

Congenital spinal teratomas

Teratomas are benign tumors that arise from remnants of all three germ cell layers. Spinal teratomas are most commonly located in the sacrococcygeal region. These tumors can be isolated or they can be associated with anorectal malformations and caudal agenesis in Currarino Triad. Spinal teratoma in other regions is rare and can be associated with spinal malformations, including spina bifida, partial sacral agenesis, hemivertebra, and diastematomyelia.

Sacrococcygeal teratoma is the most frequent congenital tumor, and most of these tumors are diagnosed during the pregnancy or at birth. These tumors are histopathologically classified into mature, immature, and malignant teratomas. Mature teratomas are associated with parenchymal tissue, fat, and calcifications. Immature forms lack the adipose tissues, have increased growth rate, and may metastasize. Most teratomas in infancy and early childhood are benign. There is, however, a tendency for malignant transformation over time. Treatment of choice in the management of the sacrococcygeal teratomas is radical excision. With early diagnosis and surgical excision, newborns who have sacrococcygeal teratomas often have excellent prognosis with long-term survival and rare recurrence. Fetuses diagnosed with sacrococcygeal teratomas, however, are at high risk for perinatal complications and death related to hemodynamic changes associated with tumor vascular steal and maternal obstetric complications. Several studies have used fetal surgery, tumor embolization, radiofrequency, or laser ablation for the management these fetal teratomas.

Atypical teratoid/rhabdoid (AT/RT) tumor is an uncommon congenital central nervous system neoplasm that is highly aggressive and malignant (Fig. 6). Histologically these tumors are characterized by rhabdoid cells with or without a mixture of epithelial, neuroepithelial, or mesenchymal components. The rhabdoid cells exhibit round nuclei with prominent nucleoli and eccentrically located cytoplasm. Cytogenic studies have shown that these tumors are characterized by monosomy or partial deletions of chromosome 22q in 60% to 87.5% of cases. Molecular studies have also demonstrated homozygous deletions or mutations of the INI1 gene at chromosome 22q11.2 locus in 75% of AT/RT and rhabdoid tumors of other organs. The tumor is most commonly located in the posterior fossa and other regions of the intracranial cavity. Spinal atypical teratoid/rhabdoid tumors are extremely rare, however; 12 cases have been reported in the English literature thus far. Most of these tumors were located in the spinal cord, with several cases reporting extramedullary location. Multimodality treatment, including surgical excision, chemotherapy, and radiotherapy, is typically required for this highly malignant neoplasm. Despite aggressive treatment, patients who have spinal AT/RT have a life expectancy of less than 12 months, similar to those who have intracranial diseases.

Instrumentation

When operating on the spine of a pediatric patient, long-term spinal balance and stability

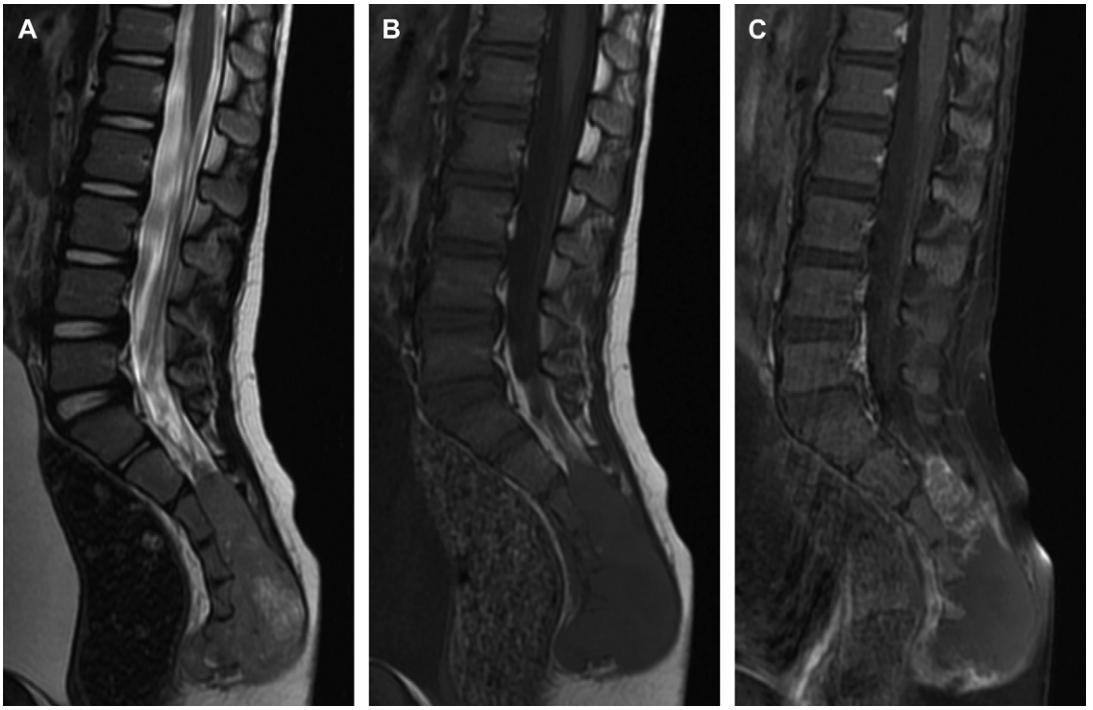

Fig. 6. MRI of sacral atypical rhabdoid tumor in a 4-year-old girl. (*A*) Sagittal T2, (*B*) sagittal T1, and (*C*) sagittal T1 with gadolinium.

must be considered. If the child is at risk for development of postsurgical deformity, a spinal fusion, instrumented or noninstrumented, should be done at the same time as the tumor excision. Although stainless steel has long been used as an internal fixator in the treatment of idiopathic scoliosis of children, titanium is recommended following tumor resection. The use of titanium instrumentation allows MRI scans to be obtained for postsurgical surveillance with less metal-associated artifact [66].

Summary

Treatment of pediatric spine tumors is based on location, extent of involvement, and biologic behavior. Patients who have benign tumors that possess a favorable natural history may not require biopsy or surgical excision if discovered as an incidental finding. The natural history of some of these tumors is natural resolution over a couple of years. Such patients should be closely followed, however, to ensure that further progression does not occur. In some cases, surgery may be considered to relieve symptoms or to confirm the diagnosis. Pediatric patients who have benign tumors that are locally aggressive, namely osteoblastomas and ABCs, require open surgery to achieve cure. Although marginal excision is more likely to prevent recurrence, the associated morbidities of such radical surgery in the pediatric patient may outweigh the benefits.

Children who have malignant tumors of the spine are extremely challenging to treat and are best managed with multidisciplinary care. Biopsy of suspected malignancies should be done with the assumption that the lesion is locally aggressive. For this reason, transcavitary approaches (transoral, transrectal) should be avoided, and the biopsy site should be clearly identifiable on the skin for subsequent en bloc removal with the specimen if needed. Children who have confirmed malignancy usually begin chemotherapy before definitive tumor surgery to reduce the size of the tumor mass, allowing en bloc resection of the tumor with negative margins to be more feasible. Postoperative chemotherapy is often required to eradicate micrometastases, and postoperative radiation therapy may also be necessary for patients who

have positive margins on final pathology or those who have unresectable tumors following chemotherapy. Finally, current instrumentation allows effective stabilization of a compromised spinal column and may be used even in very young children following spine tumor excision.

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