Hip Flexion Contracture Caused by an Intraspinal Osteochondroma of the Lumbar Spine

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abstract

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Osteochondroma (or osteocartilaginous exostosis) is the most common bone tumor of childhood, with an incidence ranging from 1 to 1.4 per 1,000,000. In the lumbar spine, osteochondromata usually arise from the posterior column at the secondary ossification center and grow away from the spinal canal without causing neurologic deficits. This article reports a rare intraspinal lumbar osteochondroma that compressed the thecal sac, resulting in a hip flexion contracture in an 11-year-old boy. This lumbar, intraspinal, extradural exostosis was confluent with the L3 inferior articular process and compressed the L3 nerve root and thecal sac severely. The patient underwent an en bloc resection of the tumor with a right-sided hemilaminectomy of L3 and L4, a right-sided partial facetectomy at L3 to L4, and an extended resection from the pars intra-articularis of the L2 to the L5 vertebrae. The tumor specimen measured 4.8×3.7×2.5 cm with clear margins. Instrumented posterolateral fusion was completed from L2 to L5 due to iatrogenic instability from the resection. The patient had an uneventful recovery and returned to his normal activities of daily living, including sports. He remains asymptomatic at 54-month follow-up. A solitary lumbar osteochondroma that compresses the spinal cord, resulting in a motor neurological deficit, has not been reported in a pediatric patient. Orthopedic surgeons should be aware of potential intraspinal presentation of osteochondromas. Magnetic resonance imaging is the modality of choice in diagnosing and screening for spinal osteochondromas. These cases can be treated with resection surgery.

Figure: Sagittal computed tomography scan of the lumbar spine demonstrating a large osseous structure that is confluent with the right L3 inferior articular process. Note the characteristic features for an osteochondroma, including a well-delineated cortex continuous with the host vertebrae and sclerotic changes in the neighboring bone.

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Osteochondromas (or osteocartilaginous exostoses) are the most common bone tumor of childhood. These tumors may be solitary or multiple, as seen in hereditary multiple exostoses, an autosomal dominant genetic disorder. Spinal osteochondromas are rare, with a frequency of 1% to 4%, and occur most often in the cervical and upper thoracic vertebrae. Lumbar spinal osteochondromas are even more rare, comprising only 9% of all spinal osteochondromas and 0.09% to 0.36% of all osteochondromas. In hereditary multiple exostoses, 3 specific genotypes may determine the severity of the phenotype: EXT1 (18q), EXT2 (11p), and EXT3 (19p). Approximately 60% to 70% of these patients have a mutation in EXT1 gene, which is associated with the most severe form of the disease and the highest probability of malignant transformation.

In the spinal column, osteochondromas usually arise from the posterior elements as secondary ossification centers. The tumor typically grows posteriorly away from the spinal canal, rarely causing neurological deficits. The current study reviews a rare case of hip flexion contracture caused by an intraspinal osteochondroma of the lumbar spine and the literature on this topic.

**Case Report**

An 11-year-old boy presented with a 2-year history of vague groin pain (right greater than left). Developmental milestones and growth had been appropriate throughout his life. Neither the patient nor his family had a remarkable medical history. There was no associated back pain. He was the lead linebacker for his middle school. One year prior to presentation, the coach had observed deterioration in the patient’s performance and recommended physical therapy. Over the course of physical therapy, the therapist observed progressive hamstring tightness, bilateral hip stiffness (right greater than left), and poor gait refractory to therapy. The patient’s pediatrician noted a hip flexion contracture and scoliosis on examination after an unremarkable history and examination 3 years earlier. The patient was referred to a pediatric orthopedist.

At presentation to the senior author (A.E.), on physical examination, palpation of the lumbar spine revealed a midline nontender mass with diminished lumbar lordosis in a standing position. Thompson test of the right hip was 50° with a 15° hip flexion contracture. Thompson test of the left hip was 40° with an 8° flexion contracture. The patient had a popliteal angle of 45° on the right and 35° on the left. The patient had an antalgic gait with reduced motion at the hips and knees.

Neurological examination revealed hypoesthesia of the right anterior thigh, a positive straight leg test on the right, and grade 3/5 strength for the right quadriceps. The right thigh showed muscle atrophy with a circumference 3 cm less than the left side. Babinski and deep tendon reflexes were unremarkable bilaterally.

Plain radiographs showed a right thoracolumbar congenital scoliosis centered at the L3-L4 junction measuring 20°. Loss of the normal lumbar lordosis was seen with a lordosis angle (L1-L5) of 17°. A mass was noted in the posterior elements of the lumbar spine (Figure 1).

Computed tomography (CT) revealed a right-sided osseous structure measuring approximately 3.7 cm in length and confluent with the right L3 inferior articular process. Note the characteristic features for an osteochondroma, including a well-delineated cortex continuous with the host vertebrae and sclerotic changes in the neighboring bone. The mass extended posteriorly through the posterior elements past the spinous process and spanned multiple vertebral levels from L2 to L4. There was an extradural component inside the canal pressing on the thecal sac posteriorly at the L2-3 intervertebral disk space level, which created a focal area of severe spinal stenosis and compression of the L3 nerve root (Figure 2).

T2-weighted magnetic resonance imaging (MRI) of the lumbar spine revealed...
Case Report

Severe spinal dura mater compression by the intracanicular, extradural exostosis at the L2-L3 level (Figure 3). This lumbar osteochondroma occupied 80% of the spinal canal. The central portion of the tumor had an increased signal intensity on MRI that was surrounded by peripheral ossification and a cartilage cap of low-signal intensity on T1- and T2-weighted MRI. Whole-body technetium-99m-methylene diphosphonate bone scan showed increased concentration activity of the posterior elements of L3, but no abnormal uptake elsewhere in the body. Supplemental full-length MRI of the spine also showed no other lesions. A diagnosis of a solitary lumbar osteochondroma was made.

The patient underwent an en bloc resection of the tumor with a right-sided hemilaminectomy of L3 and L4, right-sided partial facetectomy at L3 to L4, and extended resection from the pars intra-articularis of the L2 to L5 vertebrae (Figure 4). The tumor specimen measured 4.8×3.7×2.5 cm with clear margins. Instrumented posterolateral fusion was completed from L2 to L5 due to iatrogenic instability from the resection. No changes in neuromonitoring were noted throughout the case. Histopathologic examination confirmed a lumbar osteochondroma diagnosis: typical cartilage cap and endochondral ossification (Figure 5). There were multiple islands of cartilage that had not yet ossified. There was no histological evidence of malignant transformation.

Immediately postoperatively, the right groin pain resolved and the patient had no new focal neurological deficits. He was discharged on postoperative day 1. On follow-up visits, the patient reported no complaints or neurological deficits. Eight months postoperatively, CT showed fusion, maintenance of coronal and sagittal alignment, and no tumor recurrence. At 54-month follow-up, the patient was asymptomatic and fully participating in football with no clinical or radiographic signs of tumor recurrence.

Discussion

Osteochondromas are epiphyseal growth disorders originating from the periosteum. They are caused by aberrant epiphyseal growth when part of the physis is displaced laterally through a ring of perichondrium. Most solitary and hereditary osteochondromas occur in the cervical spine (57%), with the C-2 vertebra being the most frequent site, followed by the thoracic spine (33%) and lumbar spine (9%). In children, solitary exostoses comprise roughly 52% of tumors, whereas 48% are associated with hereditary multiple exostoses.14 Spinal exostoses grow gradually during these years, and often become symptomatic during the 2nd to 3rd decade of life.8,15,16 Although reports show that up to 27% of spinal exostoses are intraspinal, only 0.5% to 1% of patients present with neurological dysfunction.11-13 Spinal cord compression is unusual because the majority of lesions grow outside of the spinal canal.9 As found in the current case, spinal cord compression may be associated with a delay in diagnosis because the lesion grows slowly and may be inconspicuous on radiographs.18

Only 1 case of a motor deficit caused by intraspinal lumbar osteochondroma has been described; no study to date has described this phenomenon in a pediatric patient. Kahveci et al19 described a 48-year old man who had a right foot drop due to acute compression of the right L4 nerve root by a solitary lumbar osteochondroma. Similar to the current patient, the symptoms of this patient also improved after surgical removal of the lesion.19

In the current patient, hip flexion contracture was caused by the limited lumbar lordosis. The patient favored a kyphotic position due to the lumbar stenosis to increase the canal diameter. In the office, the patient favored a markedly kyphotic posi-
tion that was not as apparent on the radiographs. Furthermore, radiographs showed a loss of the normal lordosis from the congenital kyphosis and from the autofusion of the posterior elements above that level from the large exostosis. The chronic postural and structural kyphosis led to a contracture of the hip flexor muscles.

The literature suggests that neurologic findings in hereditary multiple exostoses usually present between 22 and 25 years of age. Prophylactic screening of the spine has not been routinely performed in these patients because intraspinal lesions are thought to be rare. Roach et al, however, used MRI to screen for spinal exostoses in 44 patients with hereditary multiple exostoses and reported that 68% had spinal osteochondromas and 27% had intraspinal lesions. This is clinically relevant because these silent intraspinal osteochondromas can be dangerous and life-threatening. Reports in the literature describe symptoms that present acutely and lead to quadraplegia and death. Due to the potential consequences of such lesions, Roach et al used a protocol whereby all patients with hereditary multiple exostoses were screened with full-spine MRI beginning at 4 years of age. This age was chosen because they found 2 patients younger than 4.5 years who had asymptomatic intraspinal exostoses.

The imaging modality of choice in detecting osteochondromas of the spine is controversial. Computed tomography, post-myelography CT, and MRI are useful for evaluating the origin, size, and extent of spinal cord compression. Nuclear medicine bone scans are useful in identifying additional lesions throughout the skeleton, and can be used in patients with hereditary multiple exostoses. Plain radiographs are limited in detecting spinal osteochondromas because of the complex image formed by the spine, which can lead to inconclusive interpretations in up to 79% of cases. Roach et al found that MRI has an adequate sensitivity for detecting osteochondromas of the spine. Although CT gives excellent visualization of the bony cortex, MRI may be the imaging modality of choice due to adequate sensitivity, visualization of the cartilaginous cap, effects on the neural tissue, and the risk of radiation exposure from CT. This is especially true for screening purposes in which the entire spinal axis can be imaged without radiation exposure in a pediatric patient.

The current patient’s radiographs demonstrated congenital scoliosis (L4-L5 unilateral segmentation) and a lateral curvature of 20°. A literature review found only 2 case reports of an osteochondroma presenting as scoliosis, and both cases were associated with hereditary multiple exostoses. In 1 case, the lesion was located near the apex of the curve on the concave side, similar to the location of the more common osteoid osteoma and osteoblastoma when they typically cause scoliosis. In the other case, the patient’s curve was centered at the thoracolumbar junction, but the lesions were located at T4 and L4. In both cases, the curvature was believed to be due to spasm secondary to lumbar nerve root traction or pain, similar to the proposed mechanisms of scoliosis associated with an osteoid osteoma and osteoblastoma. These are the only cases of a osteochondroma presenting as congenital scoliosis. The congenital scoliosis can alter the biomechanics of the lumbar spine, but there is no known association between osteochondromas and congenital scoliosis.

Surgical planning requires precise localization of the tumor origin by full-spine imaging workup. Asymptomatic lesions can be followed nonoperatively due to the low rate of malignant transformation. A sudden increase in lesion size or new onset of pain may suggest malignant transformation in these cases. The risk of malignant degeneration into chondrosarcoma is 1% for solitary lesions and 10% to 25% in patients with hereditary multiple exostoses. Radiological findings consistent with malignant transformation include tumor growth after closure of the growth plate, changes in the tumor’s border, internal lytic areas, destruction of adjacent bones, and the presence of soft tissue masses containing calcifications. On MRI, malignancy should be suspected when the thickness of the cap is greater than 2 cm in adults and greater than 3 cm in children.

Surgical indications include pain, neurologic deficit, or if the tumor appears malignant. Complete excision is recommended due to high recurrence rates with incomplete removal. Bess et al reported that the mean recurrence time was 4.2 years, and recurrence was seen exclusively in patients with intraläsional excision. In patients with neurologic compromise, surgical resection by decompressive laminectomy is advised and often results in excellent functional recovery. Albrecht et al found that 89% of symptomatic patients treated operatively reported improvement of symptoms. In the current patient, a decompressive hemilaminectomy was performed with complete neurological recovery without recurrence at 4.5-year follow-up.

Conclusion

A solitary lumbar osteochondroma that compressed the spinal cord resulted in a motor neurological deficit that was successfully treated with surgical excision in a pediatric patient. The authors advocate screening these patients with full-spine MRI starting at 4 years of age to locate intraspinal exostoses in order to avoid potentially unfavorable outcomes. Future studies should focus on determining the true incidence of intraspinal osteochondroma, comparing the sensitivity and specificity of MRI to CT in locating intraspinal osteochondromas and further validating the use of MRI as a screening tool for these patients.

References
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