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Spinal stenosis frequent in children with multiple hereditary exostoses

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Abstract
Purpose Children with multiple hereditary exostoses (MHE) have numerous osteochondromas, with the most prominent lesions typically over the appendicular skeleton. A recent report noted a high rate of intracanal lesions in this patient population and recommended preventative spinal screening with magnetic resonance imaging (MRI) or computed tomography (CT). We sought to evaluate the prevalence of spinal stenosis from intracanal osteochondromas at our pediatric orthopedic center in order to evaluate if routine screening is warranted.
Methods All pediatric patients treated for MHE were retrospectively identified. Records were reviewed to determine demographics, previous orthopedic surgery, and indication and results of axial spine imaging (CT or MRI). Imaging studies were reviewed to evaluate the presence of intracanal and compressive spinal lesions.
Results Between 1990 and 2011, axial imaging was performed in nine patients with MHE due to concerns of pain, weakness, and/or dizziness. These patients had moderate disease involvement, with a mean of 4.9 previous orthopedic surgeries to address skeletal osteochondromas. Two patients with MHE had cervical spinal stenosis secondary to intracanal osteochondromas. Both children successfully underwent spinal decompression. Thus, of our MHE population undergoing axial imaging, 22 % were noted to have intracanal lesions.
Conclusions Our experience reveals a >20 % rate of compressive intracanal osteochondromas in MHE patients undergoing spinal imaging. These two patients represent 5 % of the MHE patients treated at our center. These lesions may be slow growing, and significant consequences can occur if not identified promptly. Thus, we confer that routine axial screening of the spinal canal may be warranted in these children.

Keywords Pediatric · Multiple hereditary exostoses · Neurologic symptoms · Spinal impingement · Imaging · MRI · CT

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Introduction

Multiple hereditary exostoses (MHE) is a skeletal dysplasia resulting in multiple cartilage-capped lesions. It is a genetic disorder inherited in an autosomal dominant fashion with incomplete penetrance in females [1, 2]. Although osteochondromas have a predilection for the metaphyses of long bones, the axial skeleton and vertebral column can also be involved [3–8].

There are numerous published reports of spinal cord impingement and neurologic compromise from intracanal osteochondromas [6, 9]. In most series, patients present with pain, weakness, or other significant neurologic findings [4, 8–23]. Roach et al. found that 12 out of 44 patients screened had lesions encroaching into the spinal canal and, thus, recommended routine magnetic resonance imaging (MRI) or computed tomography (CT) screening of the neuroaxis in patients with MHE to detect compressive lesions prior to the onset of neurologic deficits [6, 9]. However, the prevalence of asymptomatic compressive intracanal lesions in patients with MHE has not been confirmed at other centers [6]. Thus, we undertook a retrospective review to determine the prevalence of intracanal osteochondromas in children with MHE imaged at our center.

Materials and methods

A retrospective chart review was undertaken at a dedicated pediatric orthopedic hospital. The diagnostic registry was searched to identify all patients treated for a diagnosis of MHE between 1990 and 2011. Patients with solitary osteochondromas were excluded from this study, as were patients greater than 20 years of age. Records were reviewed for demographics, previous surgeries, and indication for axial imaging. A search of diagnostic codes yielded 67 possible patients. Of those, 44 were found to have a confirmed diagnosis of MHE. Nine patients had undergone axial imaging. All imaging studies were reviewed to determine the presence of an intracanal osteochondroma, associated neurologic symptoms and physical examination findings, and treatment course. Institutional review board approval was obtained for all aspects of this study.

Results

During the study period, axial imaging was performed in nine patients with MHE for a variety of indications, including back pain, routine screening, leg pain, spasticity, and headache (Table 1). The mean age at the time of scanning was 16.1 years (range 9–20 years). During the study period, the nine patients had undergone a mean of 4.9 orthopedic surgeries (range 2–10) under general anesthesia at our center.

Two patients with MHE with minimal symptoms were found to have cervical spine stenosis secondary to intracanal osteochondromas. Both children successfully underwent spinal decompression. Thus, of our MHE population, 20.5 % (9 of 44 patients) had symptoms that warranted axial imaging and 4.5 % (2 of 44 patients) were noted to have intracanal lesions.

The first patient was a 15-year-old female with known history of MHE who presented with right hip pain and sudden give-way on her right lower extremity, resulting in frequent falls. She had had multiple previous symptomatic osteochondromas resected from her lower extremities. She had missed a significant amount of school. She had no history of headaches or neck or back pain. On examination, she walked with a stable gait with no limp. She had 3+ knee jerk and ankle jerk, and downgoing toes bilaterally with no clonus. Sensation was intact to light touch throughout. She had 5/5 strength in her bilateral upper and lower extremities. While prone, right hip internal rotation was to 10°, compared to 35° on the left side. External rotation was 60° bilaterally. The patient had no pain with flexion internal rotation on the right hip. Hip flexion was to 120° bilaterally. Pain in the right hip was reproduced with extension and external rotation, with pain radiating down the leg. The spine appeared straight, with no evidence of

Table 1 Summary of patients undergoing axial imaging

Patient number	Age at imaging	Gender	Reason for imaging	Area imaged	Findings
1	20	F	Back pain	MRI—spine	Normal
2	19	F	Osteochondroma on chest wall	MRI—spine	Normal
3	16	F	Back pain and MHE	MRI—spine	Normal
4	9	M	MHE	MRI—spine	Normal
5	15	F	Leg weakness, pain, lesion in right hip	MRI—spine	Osteochondroma on lamina of C5 compressing the spinal cord
6	19	M	Pain	MRI—spine	Normal
7	18	M	Spasticity, pain, and fasciculations	MRI—brain and spine	Normal
8	14	F	Rib and back pain	CT—spine	Normal
9	15	F	Dizziness, headaches needing ER visits	MRI—spine	Osteochondroma on C2 compressing the spinal cord

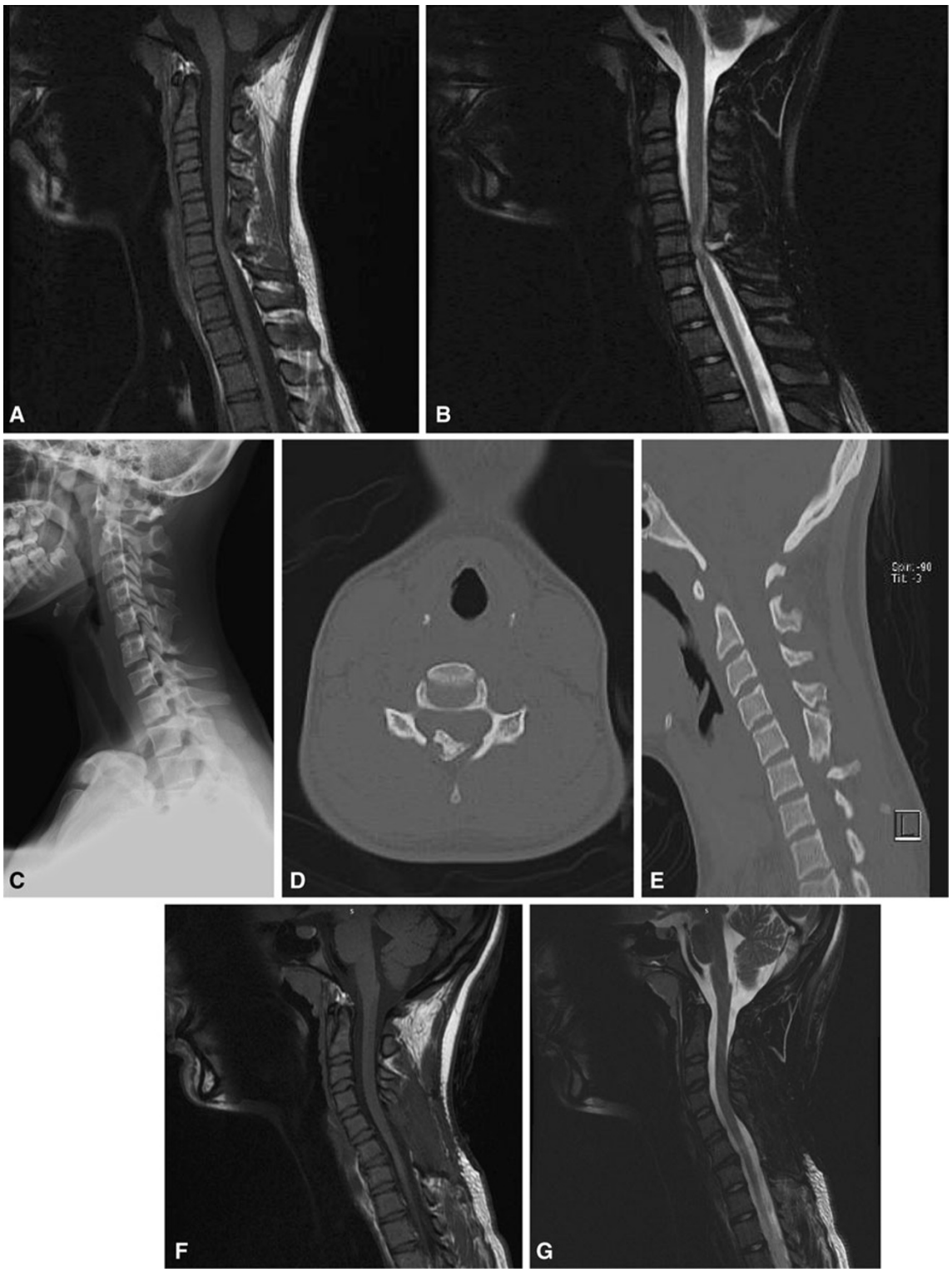

Fig. 1 A 15-year-old female with multiple hereditary exostoses (MHE) presented with hip and posterior leg pain. Magnetic resonance imaging (MRI) screening of the entire spine was undertaken, given concern for radicular-type symptoms. **a**, **b** Sagittal T1 and T2 imaging of the cervical spine revealed a large posteriorly based osteochondroma compressing the spinal cord, with signal change on T2 imaging. **c** The lesion is not evident in plain radiographs. **d**, **e** Computed tomography (CT) underestimates the size of the lesion, given the large cartilage cap. Nevertheless, the appearance of the osteochondroma on computed tomography (CT) is concerning for spinal stenosis. This patient underwent surgical decompression, with no improvement in her hip and leg symptoms. **f**, **g** Sagittal T1 and T2 imaging of the cervical spine status post laminectomy. Note the resolution of the previous signal change on T2

scoliosis. There were no limitations in cervical, thoracic, or lumbar spine range of motion and no tenderness to palpation throughout her cervical, thoracic, or lumbar spine. Radiographs revealed two large osteochondromas in the right proximal femur, which were thought to be impinging on soft tissues or neurovascular structures. Due to concern about the atypical presentation and her history of MHE, she underwent MRI screening of her entire spine. This revealed several osteochondromas, including a large compressive lesion extending from the lamina at C5. This resulted in significant cord compression, with increased T2 signal on MRI (Fig. 1). She underwent emergent C5–C6 laminectomy. She recovered well with no complications, although her hip symptoms persisted. She underwent surgical hip dislocation with removal of two proximal femoral osteochondromas. She tolerated this procedure well, and her hip and leg symptoms have subsequently resolved.

The second patient underwent distal femoral osteotomy at age 15 years. Postoperatively, she complained of headaches. She subsequently did well in her postoperative course. She returned six months later with worsening headaches. The recorded physical examination at that visit was unremarkable, with no evidence of neurologic deficit. CT of her brain was obtained, which was unremarkable. MRI screening of her cervical spine was subsequently obtained, which revealed a compressive lesion at C2 (Fig. 2). She underwent urgent laminectomy with complete resolution of her headaches and no change in her neurologic function.

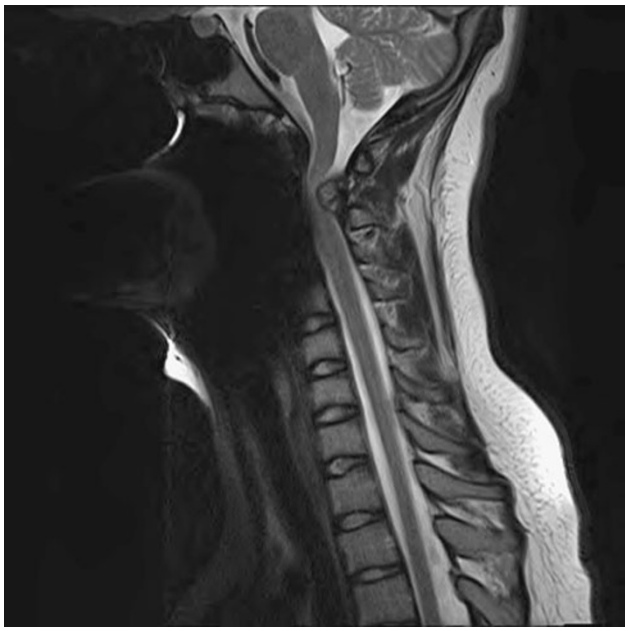

Fig. 2 Another 15-year-old female presenting with severe headaches, who initially underwent CT head imaging, which was unremarkable. Eventually, MRI screening was obtained of her cervical spine, revealing a large compressive lesion at C2. She underwent surgical decompression with good relief of her symptoms

Discussion

Osteochondromas are among the most common musculoskeletal tumors. First described in 1814 by Boyer, MHE is due to a mutation in the tumor suppressor genes EXT1 (18q), EXT2 (11p), and EXT3 (19p), which codes for a protein that participates in the biosynthesis of heparan sulfate. In patients with MHE, heparan sulfate stays inside the cell and does not go through the membrane. This causes a disturbance in the negative feedback regulatory system, thus, leading to undifferentiated proliferation of chondrocytes in the metaphyseal region [23–27]. Their composition consists of healthy, lamellar bone and a cartilaginous cap, growing by endochondral bone formation at the metaphysis of long bones [4, 6, 28, 29]. Malignant transformation to a chondrosarcoma has been reported in both the appendicular and axial skeleton, primarily in adults [30–32].

The majority of the vertebral column lesions occur in the cervical (50–80 %) and thoracic (20–36 %) regions, and rarely in the lumbar spine (Table 2). Overall, C2 is the most commonly affected vertebrae [1, 8]. It has been hypothesized that the microtrauma from cervical spine mobility leads to the displacement and subsequent growth of small cartilaginous remains, thus, resulting in a predominance of cervical lesions [5, 7].

Extra-canal cervical osteochondromas may present with dysphagia, sleep apnea, or a palpable mass [33–37]. Intracanal lesions may cause pain, paresthesias, myelopathy, weakness, or gait disturbance [4, 6, 8–10, 23]. However, spinal cord compression can be difficult to detect in children. Neurologic findings such as pain or weakness are frequently a late manifestation of cord compression in children [6, 8]. Although spinal lesions tend to progress slowly, they can lead to acute neurological symptoms following minor trauma [5, 6, 8, 38, 39]. In Roach et al.'s series, two of the three patients with symptomatic lesions had permanent neurologic sequelae (presenting with paraplegia and quadriplegia), strengthening the case for screening and aggressive surgical management of asymptomatic lesions [6].

Our series is noteworthy in that the first patient had no symptoms attributable to the cervical spine lesion, and the second patient only had headaches. More concerning, the second patient had headaches following orthopedic surgery, then her symptoms resolved, only to recur 6 month later, finally resulting in a diagnosis. It is possible that hyperextension of the neck at the time of intubation resulted in worsening symptoms due to her C2 spinal

Table 2 Summary of reported spinal osteochondromas in young patients (≤25 years of age)

References	Number of patients	Mean age, years (range)	MHE or solitary	Number symptomatic	Number asymptomatic	Type of symptoms	Location of lesion
Tahasildar et al. [40]	1	5 (5)	MHE	1	0	Restricted range of motion of the neck	Spinous process and lamina of C2 without spinal encroachment
Bonic and Kettner [33]	1	21 (21)	Solitary	1	0	Neck pain and discomfort, tender palpable mass, limited range of motion of the neck	Right bifid tip of spinous process of C5
Eap et al. [17]	1	23 (23)	Solitary	1	0	Worsening of pre-existing right hemiparesis, appearance of left hemiparesis, bladder dysfunction and urgency	Posterior arch of C4 with spinal cord encroachment
Tian et al. [4]	1	16 (16)	MHE	1	0	Difficulty walking, spastic paraparesis, hypesthesia of lower extremities, bilateral hyperactive deep tendon reflexes	Posterosuperior endplate of T6 vertebral body with spinal cord encroachment
Patel and Thacker [41]	1	14 (14)	MHE	1	0	Painful restriction of neck movement	Posterior elements of C2 with spinal cord encroachment
Ezra et al. [11]	1	4 (4)	MHE	1	0	Difficulty walking postfall, mildly reduced sensation to touch, decreased deep tendon reflexes in biceps, triceps, and brachioradialis, left Babinski sign, absent cremasteric reflex	Posterior elements of C7 and T1 with spinal cord encroachment
Gunay et al. [42]	2	14 (9–19)	Solitary	2	0	Painful swelling on the neck, restricted neck range of motion	Spinous process of C5–C6, C5 spinous process, no spinal cord encroachment
Hassankhani [43]	1	16 (16)	Solitary	0	1	Lumbar mass	Spinous process of L3
Roach et al. [6]	44	12.7 (4.2–19)	MHE	27	3	Neurological symptoms, acute quadriplegia following a minor trauma	30 out of 44 patients had spinal lesions, 12 of the 44 had spinal canal encroaching lesions, 18 of 44 patients had lesions that did not encroach the spinal canal
Han and Kuh [44]	1	7 (7)	MHE	1	0	Brown-Séquard syndrome development following mild trauma	Lamina of C7 with spinal cord encroachment
Rao and Jakheria [35]	1	8 (8)	MHE	0	1	Swollen neck mass, no neurologic symptoms	Spinous process of C2–C6, no spinal canal encroachment
Chatzidakis et al. [45]	1	22 (22)	Solitary	0	1	Asymptomatic, incidentally found on CT scan of the brain	Dens of C2

Table 2 continued

References	Number of patients	Mean age, years (range)	MHE or solitary	Number symptomatic	Number asymptomatic	Type of symptoms	Location of lesion
Song and Lee [46]	1	11 (11)	Solitary	1	0	Gait disturbance	Left superior articular process of T4 with spinal cord encroachment
Samartzis and Marco [47]	1	11 (11)	Solitary	1	0	Right posterior thigh pain for 28 months, unresponsive to conservative treatment	Right anterior surface of S2 lamina, compressing the S2 root
Maheshwari et al. [48]	1	20 (20)	Non-hereditary multiple exostoses	1	0	Increased muscle tone, exaggerated deep tendon reflexes and extensor plantar response, bilateral weakness below C6 myotome, motor loss below T2	Left pedicle of C7 with spinal cord encroachment
McCall et al. [49]	1	13 (13)	Solitary	0	1	Posterior neck mass	Lamina of C3, mild cord encroachment
Giudicissi-Filho et al. [10]	1	18 (18)	MHE	1	0	Progressive weakness in lower limbs, quadriparesis, bilateral hyperactive deep tendon reflexes, bilateral Babinski sign	Lamina of C7 with spinal cord encroachment
Aldea et al. [50]	1	24 (24)	MHE	1	0	Acute onset of difficulty in ambulating, bilateral hyperesthesia below C7	Right C7 lamina with spinal cord encroachment
Faik et al. [18]	2	18 (17–19)	1 MHE and 1 solitary	2	0	Spastic paraparesis and a pyramidal syndrome in bilateral lower extremities, weakness and fatigue of right lower limb, pyramidal signs in right lower limb	Anterior and lateral part of the spinal canal at the T2–T3 level with spinal cord encroachment, costovertebral angle at the T4 level with spinal cord encroachment
Miyamoto et al. [51]	1	23 (23)	MHE	1	0	Progressing tetraparesis	Pedicle of C2 on the left side, severe spinal cord encroachment
Chooi et al. [29]	1	23 (23)	MHE	1	0	Weakness of right arm and both legs, difficulty ambulating, shock-like sensation radiating down right arm when head turned to right, urinary and bowel incontinence	Right anterior aspect of the posterior arch of C1 with spinal cord compression
Korinth et al. [52]	1	12 (12)	MHE	1	0	Right-sided weakness and gait disturbance	C2 lamina with spinal cord encroachment
Fiechtl et al. [53]	1	8 (8)	MHE	1	0	Difficulty with ambulation	L4–L5 facet joint with neural canal encroachment
Oga et al. [54]	1	13 (13)	MHE	1	0	Slowly progressive gait disturbance	Lamina of C3 with spinal cord encroachment
Govender and Parbhoo [55]	1	14	MHE	1	0	Weakness in both lower extremities and urinary incontinence	Neural arch of T8 with spinal cord encroachment

Table 2 continued

References	Number of patients	Mean age, years (range)	MHE or solitary	Number symptomatic	Number asymptomatic	Type of symptoms	Location of lesion
Khosla et al. [56]	2	11 (5–17)	Solitary	2	0	Weakness and atrophy of right arm, hyperreflexia of both lower extremities, difficulty ambulating, spastic gait	Right pedicle of C7 with encroachment, right posterior arch of T8 with encroachment
Ergün et al. [57]	1	16 (16)	MHE	1	0	Quadriparesis	Left pedicles and intervertebral joints at the level of C5–C6 with spinal cord encroachment
Mikawa et al. [58]	1	17 (17)	MHE	1	0	Progressive difficulty walking, spastic quadriparesis	Lamina and pedicle of C7 with spinal cord encroachment
Atabay et al. [15]	1	17 (17)	MHE	1	0	Weakness in all extremities, neck pain and urinary dysfunction	Right C2 lamina with spinal cord encroachment and posterior arch of C4
Labram and Mohan [59]	1	9 (9)	MHE	1	0	Difficulty with ambulation and balance, nocturnal enuresis, quadriparesis	Lamina of C2 with encroachment and spinous process of T9 with no encroachment
Robbins et al. [60]	1	15 (15)	MHE	1	0	Right arm weakness and reduced lateral rotation of the neck following fall from trampoline	Posterior elements of C3 with spinal cord encroachment
Morikawa et al. [61]	1	21 (21)	Solitary	1	0	Intermittent numbness of both hands	Posterior arch of C1 with cord encroachment
Barros Filho et al. [62]	1	16 (16)	MHE	1	0	Dysphagia	Anterior arch of C1
Eder et al. [63]	1	7 (7)	MHE	1	0	Gait disorder	Posterior arch of C2 with spinal cord encroachment
Albrecht et al. [7]	1	16 (16)	Solitary	0	1	Asymptomatic	Right pedicle of T12
Shapiro et al. [64]	1	11 (11)	MHE	1	0	Difficulty with ambulation	Lamina of C2 with spinal cord encroachment
Moriwaka et al. [65]	1	9 (9)	MHE	1	0	Progressive transverse myelopathy	C7–T1 vertebral bodies with spinal cord encroachment
Wen et al. [66]	1	23 (23)	MHE	1	0	Symptoms following a fall from chair: apnea, tetraplegia, reduced sensation on right side and left arm	Lamina of C1 with spinal cord encroachment
Tully et al. [67]	1	12 (12)	MHE	1	0	Spastic quadriplegia	Vertebral body of C5 with spinal cord encroachment
Scher and Panje [68]	1	10 (10)	Solitary	1	0	Hoarseness, neck swelling	Vertebral body of C2–C5 on the left posterior side
Kozlowski et al. [69]	2	9.5 (6–13)	Solitary	1	1	Tenderness over coccyx, asymptomatic	Second segment of the coccyx, right pedicle of L5 with slight indentation of the dural sac

Table 2 continued

References	Number of patients	Mean age, years (range)	MHE or solitary	Number symptomatic	Number asymptomatic	Type of symptoms	Location of lesion
Cohn and Fielding [70]	1	9 (9)	Solitary	0	1	Enlarged neck mass	Spinous process and lamina of C2, with no spinal canal encroachment
Karakaş and Patiroğlu [71]	1	7 (7)	Solitary	0	1	Non-tender neck mass, limited cervical range of motion, and slight torticollis	Spinous process and lamina of C5
O'Connor and Roberts [72]	1	24 (24)	MHE	1	0	Numbness and weakness in right arm and leg	Lamina of C5 with cord encroachment
Misra et al. [73]	1	25 (25)	MHE	1	0	Difficulty walking, weakness in arms and legs	C4 lamina with spinal cord encroachment
Novick et al. [74]	2	8.5 (8–9)	Solitary	0	2	Painless neck mass	Spinous process of C5, left transverse process of C4
Palmer and Blum[75]	2	18 (14–22)	MHE and solitary	2	0	Incontinence, spastic gait, pain radiating down left arm	Body of C6 with encroachment, C7 with no encroachment
MacGee [76]	1	16 (16)	Solitary	1	0	Transient quadriplegia for 30 min following trauma to chin	Posterior atlas of C2 with spinal cord encroachment
Ferrari et al. [77]	1	23 (23)	MHE	1	0	Spastic paraparesis, gait disturbance, sphincter dysfunction	C2 right hemilamina with spinal cord encroachment
Glasauer [78]	1	19 (19)	Solitary	1	0	Inverted supinator reflex, depressed right biceps reflex	Right lateral mass of C4–C5
Urso et al. [79]	1	9	MHE	1	0	Bladder disturbance, lumbar lordosis, abnormal gait	Right vertebral lamina of L4 with spinal canal encroachment
Inglis et al. [80]	1	8 (8)	Solitary	0	1	Painless neck mass	Spinous process of C5
Twersky et al. [81]	1	13 (13)	Solitary	1	0	Lumbar pain, bilateral sciatica	L4 with spinal canal encroachment
Madigan et al. [14]	1	7 (7)	MHE	1	0	Paresis, gait disturbance, frequent falls, urinary incontinence	C2 lamina with anterior spinal cord encroachment
Fielding and Ratzan [82]	1	14 (14)	Solitary	0	1	Painless neck mass	Spinous process of C3
Crowell and Wepsic [83]	2	15.5 (13–18)	MHE (1)/low-grade chondrosarcoma (1)	2	0	Weakness in both legs, impaired position sense in toes, ankles, and knees, episodic tingling in low back	Pathology revealed low-grade chondrosarcoma, T2 with spinal encroachment, T8 with spinal canal encroachment
Vinstein and Franken [84]	1	14 (14)	MHE	1	0	Gait disturbance, left hemiparesis with clasp-knife spasticity	Arch of C2 with spinal cord encroachment
Hickey [85]	3	16 (11–20)	Solitary and MHE	2	1	Large palpable mass at lumbosacral region, wasting and atrophy of left hand, bilateral Babinski, asymptomatic	Spinous process of L4, C6, and C7, pedicle of T7

Table 2 continued

References	Number of patients	Mean age, years (range)	MHE or solitary	Number symptomatic	Number asymptomatic	Type of symptoms	Location of lesion
Decker and Wei [86]	1	15 (15)	MHE	1	0	Bilateral spastic paraparesis, numbness and weakness of right leg	Right-sided paraspinal lesion at T10–T11 with spinal cord encroachment
Carmel and Cramer [87]	1	13 (13)	MHE	1	0	Progressive weakness in right leg and right arm, loss of sensation in the left arm, neck pain on turning head to the right	Under the right hemilamina of C2 with spinal cord encroachment
Gokay and Bucy [88]	1	24 (24)	MHE	1	0	Numbness and weakness of the left lower extremity, increased urgency and frequency of urine, incontinence	Lamina of L3
Cannon [89]	1	23 (23)	MHE	1	0	Diminished temperature sensation, numbness and tingling of legs, bilateral positive Babinski	T10

stenosis, which, at the time, was undiagnosed. Thus, routine preoperative spinal screening may be warranted for MHE patients in anticipation of neck manipulation during the process of intubation, particularly given the high rate of surgical treatment in this population. According to a recent report, 77 % of children with MHE may be expected to undergo an orthopedic surgery related to their condition [32]. Children with MHE who had had axial imaging had undergone nearly five surgeries on average, typically for the removal of symptomatic osteochondromas or placement of guided growth plates for angular deformity.

In contrast to the rheumatoid population, routine plain radiographs will not necessarily show the pathology [6]. Roach et al. [6] demonstrated that only 17 % of lesions were identifiable on radiographs. Osteochondromas that occur in long bones tend to be pedunculated lesions, while in the spine, these lesions are sessile [4, 29]. Identifying spinal osteochondromas on plain films is difficult due to the complex anatomy of the spine [7]. Thus, axial imaging of MHE children would be required, which is a potentially costly undertaking, particularly if the study were to be obtained in young children who require sedation. Both patients in our series were adolescents. However, the youngest patient in Roach et al.'s series with spinal cord compromise was 5.8 years old, suggesting that imaging is necessary even in young patients.

Roach et al.'s series was remarkable in that they noted that 9 out of 12 patients with lesions penetrating into the canal were asymptomatic. Three of the nine asymptomatic patients underwent prophylactic surgical decompression [6, 9]. Our series adds to these findings, reporting two additional children, one asymptomatic and one complaining only of headaches, who had compressive cervical spine lesions with T2 signal change requiring urgent decompression. It is important for caregivers to be familiar with the potential for spinal stenosis in this patient population, and for providers to have a low threshold for obtaining axial imaging. The primary weakness of our study is that not all 44 MHE patients in our cohort underwent axial imaging. This indicates that the prevalence of compression lesions may be higher than our reported rate of 4.5 % (2 of 44 patients). Given the retrospective nature of this study, we have not yet undertaken routine MRI screening of the neuroaxis in this patient population.

Thus, our series lends further support to routine axial imaging in children with MHE in order to detect compressive lesions and prevent neurologic compromise. Further work remains to determine whether a particular mutation or phenotype holds a greater propensity for large intracanal lesions. This may help refine the indications for screening. For now, we have begun screening all children with MHE at our center at one point during their childhood. Unless symptoms warrant it, screening is typically performed after the child is able to undergo an MRI scan without sedation in order to minimize the risk to the child. We prefer MRI over CT for screening children with MHE. With CT, the osseous center can be clearly visualized, but this often underestimates the size of the lesion. With MRI, the cartilaginous cap of the lesion is clearly visualized. It has been found that MRI and CT are comparable with respect to reliability in detecting an osteochondromas

compressing the spinal cord. MRI has a greater advantage in growing children in that there is no radiation exposure, particularly in children with MHE, who have multiple lesions with potential for malignant transformation in adulthood.

In summary, the experience at our center reveals that at least 4.5 % of patients with MHE had compressive osteochondromas (2 of 44 patients). One patient was asymptomatic. Neurologic deficit in similar patients has been reported following minor trauma. Unlike patients with skeletal dysplasia and rheumatoids who undergo preoperative cervical evaluations, neck pathology in MHE patients may not be readily detectible in children with plain imaging. Thus, we confer with the recommendations of Roach et al. for the routine axial screening of these children. Further study is required in order to determine the true prevalence of these lesions and to formalize the indications and frequency of imaging, including preoperative and, potentially, preparticipation sports screening for MHE patients.

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