Case Report

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Osteochondroma of Upper Dorsal Spine Causing Spastic Paraparesis in Hereditary Multiple Exostosis: A Case Report

Orthopaedics Section

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ABSTRACT

Osteochondroma of the spine is rare. It may present in solitary or multiple form (hereditary multiple exostoses). Herein, we report a case of an 18-year-old male who was diagnosed with thoracic osteochondroma, originating from the D4 vertebra with intraspinal extension and spinal cord compression in hereditary multiple exostosis. The patient was managed with surgery. Complete tumour excision was done to relieve cord compression and recurrence. Postoperatively the patient's symptoms were improved. At 2.5 year follow-up patient is doing well without any recurrence.

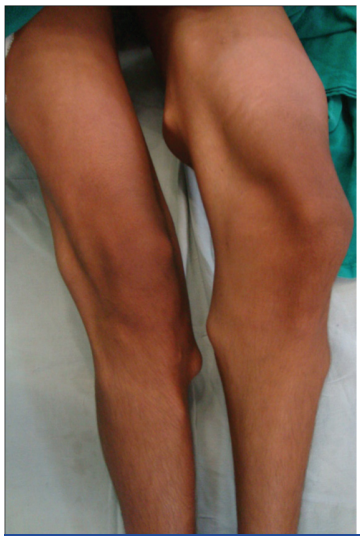
Keywords: Myelopathy, Spine, Thoracic

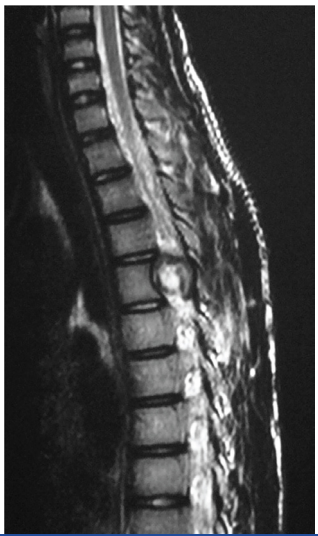
CASE REPORT

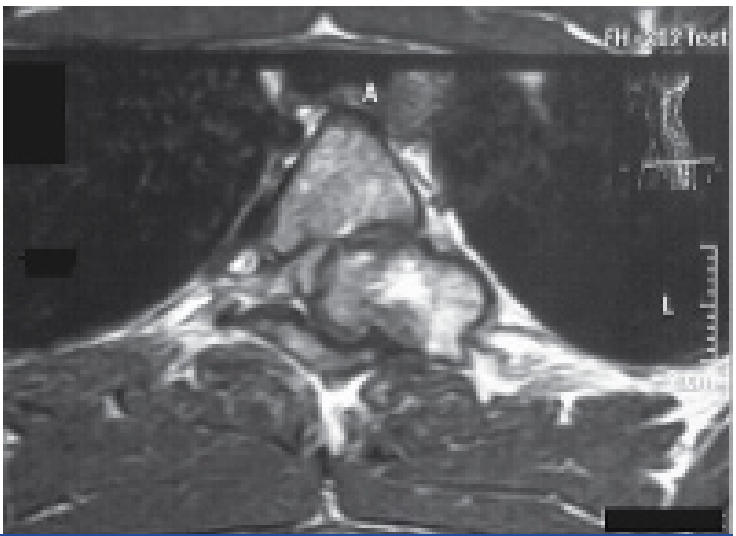
An 18-year-old male patient presented in outpatient department with complaints of multiple bony hard swellings in the arms, thighs and legs since the past 5 years and inability to walk due to weakness of both the lower limbs since the past 5 months. The complaint of weakness in both the lower limbs was gradually progressive over a period of 5 months. There was no history suggestive of bladder or bowel involvement. Patient had a positive family history with multiple osteochondromas in his younger sibling. There was no history of trauma, fever, and weight loss. On physical examination multiple, non-tender, fixed, bony hard swellings of varying sizes were present in the extremities [Table/Fig-1]. The local examination of spine was normal. The neurological examination revealed spastic paraparesis with reduced sensation below D6 dermatome on both sides. There was weakness, decreased pinprick sensation, and hyperreflexia of his lower extremity. Positive Babinski response was also elicited. Bladder and bowel functions were intact. Radiograph of the upper dorsal spine revealed exophytic bony mass overlying D4 vertebra more on the right side. Magnetic resonance imaging of the whole spine showed 3cm x 1cm bony mass arising from the right pedicle of the 4th thoracic vertebra involving the contiguous lamina, spinous process and extending from vertebral foramen to vertebral canal compressing the spinal cord. There was marked cord compression seen at this level without change in signal intensity [Table/Fig-2,3]. Radiographs of the extremities too revealed multiple mass arising from the ends of the involved bones. Excision of mass in the D4 vertebra was undertaken by posterior approach. Tumour mass was removed including lamina and pedicle of the 4th thoracic vertebrae on the right side along with facetectomy on the right side. The excised mass was sent for histopathological examination which confirmed the diagnosis of osteochondroma. The patient was given choice to remove other swellings of the body for cosmetic correction but he had not given consent for any other surgeries. Postoperatively the tone of bilateral lower limbs became normal. The weakness and numbness improved. The patient was able to ambulate on his own without support at 3 months after surgery and there was full neurological recovery after 1 year postoperatively. At 2.5 years follow up period patient is doing well and is walking without support.

DISCUSSION

Osteochondromas are the most common benign bone tumours presented as solitary (90%) or multiple lesions (10%) [1]. They are frequently located in the long and flat bones. The involvement of the spine is very rare and comprises only 1.3%- 4.1% of all osteochondromas of the spine [2]. With the best of our knowledge there are about 27 cases of thoracic vertebral exostosis in







[Table/Fig-1]: Clinical photograph showing multiple bony masses in a patient with hereditary multiple exostoses **[Table/Fig-2]:** MRI upper dorsal spine sagittal image reveals mass in vertebral canal compressing the spinal cord at D4 level **[Table/Fig-3]:** MRI upper dorsal spine axial image exophytic bony mass overlying D4 vertebra more on the right side arising from the right pedicle of the D4 vertebra involving the contiguous lamina, spinous process and extending from vertebral foramen into spinal canal

patients with Hereditary Multiple Exostosis (HME) has been described in the English literature [Table/Fig-4] [1,3-26]. The multiple osteochondromas are present in HME. HME is a genetic disorder with autosomal dominance pattern and are associated with mutations in tumour suppressor genes EXT1or EXT2orEXT3 located on chromosome 8q, 11p and 19p respectively [27]. OC are considered as developmental lesions rather than true neoplasms. Although aetiologically not clear, OCs are originated from the

Author	Year	Age	Sex	Family history	Level	Origin	Presenting complaint	Surgery	Outcome	Follow up	Remark
Cannon [3]	1954	23	F	Yes	D 10	NR	Weakness tingling numbness	laminectomy	Good	NR	No complication
Larson et al., [4]	1957	33	M	No	D 3	CVJ	Paraplegia	yes	Good	NR	_
Decker & Wei [5]	1969	15	M	No	D10	CVJ	Parapareses	yes	Good	NR	Associated Cerebellar astrocytoma
Blaauw [6]	1975	48	M	NR	D 1	CVJ	NR	yes	Good	NR	-
Twersky et al., [7]	1975	53	M	No	D 5	CVJ	NR	yes	Worsened	NR	Associated costal osteochondroma
Becker & Epstein [8]	1978	17	M	NR	D 2	CVJ	NR	yes	Good	NR	-
Ho & Lipton [9]	1979	58	F	Yes	D 1 - D2	Lamina inferoposterior aspect	15-year gradual Progressive weakness, numbness, bladder involve	laminectomy	Poor	12 Mo	No recovery
Old & Triplett [10]	1979	21	F	Yes	D 3	CVJ	NR	yes	Good	NR	_
Buur & Mørch [11]	1983	33	M	Yes	D 4	Pedicle	Spastic paraparesis	Laminectomy	Good	7 Mo	_
Moriwaka et al., [12]	1990	9	M	NR	C 7 - D1	Pedicle and VB	Pain thigh, couln't walk	Laminectomy, Facetectomy	Improved/complete recovery Good	3 Mo	_
O'Brien et al., [13]	1994	14	F	NR	D 9 - D10, D 12	Pedicle D9, D10 Extracanalicular D12 Intracanalicular	Decreased senastion with paresthesisa, spasticparaparesis with bladder involvement	Wide laminectomy T11-l1, decompression and posterior fusin	Good	1 Mo	_
Quirini et al., [1]	1996	24	M	NR	D 8	VB endplate	Difficulty in walking, numbness	yes, Excision	Good	NR	_
Govender & Parbhoo [14]	1999	14	F	NR	D 8	Neural arch	Weakness of both lower limbs and urinary incontinence	Posterior decompression	Good	3 Mo	Misdiagnosed Tuberculosis
Mermer et al., [15]	2002	15	M	Yes	D 5	VB	Weakness of right lower limb	Anterior decompression of T4–5 with anterior and posterior spinal fusion	Good	6 Mo	one or two clonus beats Remained
Faik et al., [16]	2004	17	M	NR	D2 -D3	Pedicle/VB	spastic paraparesis	decompression laminectomy	Good	NR	No complication
Bess et al., [17]	2005	11	F	Yes	D 5	VB	Ataxia, Hyperreflexia	Observation	Good	29Mo	No complication, treated conservative
Roach et al., [18]	2009	NR NR NR	M NR M	NR NR NR	D 9	NR NR VB	Quadriplegia Progressive weakness & Ataxia Paraplegia	Excision Excision No	Partially resolved Good Poor	NR NR NR	Associated cervical lesion
Ezra et al., [19]	2010	4	M	Yes	C 7 - D1	Lamina, spinous process	Pain in neck & B/L Leg Difficulty in walking urinary incontinence	Laminectomy And Excision	Improved	NR	Residual deficits included right arm and hand weakness and a mild limp
Gunay et al., [20]	2010	36	F	YES	D 12	Pedicle	Pain, Weakness, Numbness	Excision Laminectomy with Instrumentation	Good	44 Mo	slight hypoesthesia on T 11 Dermatome
Lotfinia et al., [21]	2010	31	M	YES	D 8	Facet	B/L Paresthesia, Paraperesis	Laminectomy with Instrumentation	Poor	NR	Partial Improvement
Tian et al., [22]	2011	16	M	YES	D 6	VB endplate	progressive weakness and ascending numbness	Laminectomy with fusion	Good	12Mo	_
Zaijun et al., [23]	2013	16	M	YES	D 5 - D6	SP & TP	Paraplegia, Hypesthesia	yes	Good	9 Mo	No complication
Bari et al., [24]	2012	16	M	NO	D 9 - D12	NR	Restricted movements and urinary incontinence	Surgery	Good	NR	No complication
Al Kaissi et al., [25]	2013	9	M	NR	D3-5	Pedicle	NR	yes		NR	_
Calvo et al., [26]	2013	9	M	NR	D 3	Pedicle	Inability to walk	Laminectomy instrumentation	Good	3Mo	_
Present case	2014	18 yr	M	YES	D 4	Pedicle, lamina	Paraperesis	Excision	Good	2.5 years	_

[Table/Fig-4]: Previously reported cases of thoracic vertebral osteochondromas
Abbreviations: * CT-Computerized tomography; CVJ- Costovertebral junction; MRI -Magnetic resonance imaging; HME- hereditary multiple exostoses; NR- not reported; Mo - month; OC-osteochondroma; SP- spinous process; TP- Transverse process; VB- vertebral body

separation of epiphyseal growth plate cells followed by herniation through the periosteum adjacent the growth plate [26]. The common site of OC are long bones, particularly around the knee joint, the upper humerus, flat bones especially pelvis, scapula, and ribs. It rarely occurs in the vertebrae and accounts for 1-4% of cases [2]. The vertebral OC are more common in younger male patients as seen in our case [21].

About 1% and 4% of solitary osteochondromas and 7% to 9% in hereditary multiple exostoses develop a spinal lesion [26]. The spinal involvement and neurological complications in multiple osteochondromas is higher than solitary variety [27]. In HME, thoracic and lumbar vertebrae are more commonly affected, while in solitary type cervical spine is commonly involved [16]. The involvement of sacrum is rare in both the types. A review of English literature revealed about 27 cases of thoracic myelopathy due to spinal exostosis in HME [Table/Fig-4]. Mean age of the patients was 22.5 years. Nineteen patients were male while seven patients were female. D5 vertebrae (19%) were found to be most commonly affected.

Though any part of vertebrae can be involved, the posterior arch is the most commonly affected [23]. In present case pedicle and lamina both were involved. Patients may present with back pain, cosmetic deformity and or a palpable mass. Very rarely vertebral OC may extend into the spinal canal causing cord compression and present with neurological compromise as occurred in our patient. Myelopathy is predominantly seen with multiple OCs [16].

On plain radiographs, vertebral OC is seen as bony outgrowth continuous with cortex of the bone from where it was originated. The bone marrow of the mass is continuous with the normal bone. The vertebral OCs are often small, sessile and easily missed on radiography. Computed tomography (CT) is useful to demonstrate spinal OCs which are small and have narrow stalk. In addition it is the best method to detect marrow, cortical continuity of vertebral OC though it was not done in our case. MRI of the whole spine should be performed in these cases to look for skip lesions or other masses and relation of vertebral OC to the surrounding structures. MRI shows the extent of cord compression as well as also assesses the size of cartilaginous component to rule out malignant degeneration of vertebral osteochondroma. MRI helps in early identification and planning surgery in these cases [16,23]. Similarly early MRI and surgery prevent permanent neurologic deficits as in our case. We suggest that screening MRI of the whole spine should be used in HME patients for early diagnosis of vertebral OC before the development of neurological deficit.

Grossly vertebral OC is seen as irregular bony mass with the gray white cartilaginous cap. Histologically, it has hyaline cartilages and mature bony spurs [6]. Complications of vertebral OC can include adjacent vascular and neural compromise, fracture, osseous deformity, bursa and malignant change. The malignant transformation in HME is more common than solitary osteochondromas [20]. Early diagnosis should be made in these cases if there is neurological compromise for good results. Asymptomatic vertebral OC can be left as such and patients should be followed up. Review of literature showed surgery was done in majority of cases and resulted in good results in most patients [3-6,8,10-20,22-26]. Surgical excision and decompression of spinal canal vertebral OC is required in these cases. Similarly the decompression and excision of the mass was done in our case. Recurrence can occur due to incomplete excision therefore complete "en bloc" resection is recommended. These patients should be followed up for long duration as relapse or malignant transformation may occur after surgery.

CONCLUSION

Though spine is very rare site for osteochondroma, high index of suspicion is required to diagnos it. A vertebral OC should be excluded in all patients with hereditary multiple exostosis who presents with spinal pain and neurological deficit. Surgical intervention generally has good outcome.

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