I. Lotfinia MD[1]
A. Baradaran MD[2]
M. Ghavami MD[3]

Lumbar Spine Osteochondroma Causing Sciatalgia: An Unexpected Presentation in Hereditary Multiple Exostoses

Osteochondroma, the most common benign tumor of the bone also known as exostosis, is a rarity in the spine. It occurs in solitary or multiple forms. The multiple form is often hereditary and is usually called hereditary multiple exostoses (HME). The precise diagnosis of spinal osteochondroma is performed by CT and MRI.

We report a case of HME in a 17-year-old man with a positive family history. The tumor was in continuity with L4 vertebra and the spinal canal stenosis was evident. The full recovery was successful after tumor resection.

Key words: Osteochondroma, Hereditary Multiple Exostoses (HME), Lumbar Vertebrae, Sciatal Neuralgia

Introduction

Osteochondroma (osteocartilaginous exostosis) according to the 2002 WHO definition is a cartilage capped benign bony neoplasm on the outer surface of the bone performed by endochondral ossification.[1] These tumors known as exostosis, are the most common benign tumors of the bone, accounting for 36% of all benign bone tumors and 85% of all bone tumors.[2] These lesions may be solitary or multiple.[3] The multiple forms or the hereditary nature of this disorder is usually transmitted as an autosomal dominant trait with a variable penetrance.[4] Radiculopathy and/or myelopathy caused by osteochondromas are rarer because the majority of these lesions grow out of the spinal canal.[4-6] Simultaneous spinal cord compression is an extremely serious and uncommon complication of osteochondromatosis.[4] In the cases published in the literature, the incidence of osteochondroma of the lumbar region is definitely rarer than other regions.[7,8]

Case Presentation

A 17-year-old man with a two-month history of intermittent sciatalgia involving the right lower extremity, was admitted to our department on August 19, 2007. He had a history of hereditary multiple exostoses (HME) in his lower limbs, upper limbs and ribs since childhood and a positive family history. His past medical history was remarkable for excisions of exostoses from his right femur at the age of 15. He had a normal intelligence level and his father also had numerous exostoses.

On examination, palpable exostoses were discovered over the bones of his arms and legs. A neurological examination revealed limited straight-leg-rising (Lasegue's sign) on the right side and no motor weakness. Sensory examination

1. Associate Professor, Department of Neurosurgery, Shohada Hospital, Tabriz University of Medical Sciences, Tabriz, Iran.
2. Department of Neurosurgery, Shohada Hospital, Tabriz University of Medical Sciences, Tabriz, Iran.
3. Assistant Professor, Department of Radiology, Shohada Hospital, Tabriz University of Medical Sciences, Tabriz, Iran.

Corresponding Author:
Iraj Lotfinia
Address: Department of Neurosurgery, Shohada Hospital, Ellgolle St., Tabriz, Iran.
Tel: +98411-540-8016
Fax: +98411-3340-830
Email: lotfiniai@yahoo.com

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showed hypalgesia on the lateral aspect of the lower right leg. Laboratory examinations including electrolytes and complete blood count were normal. Plain lumbar spine radiographs appeared normal, but other preoperative radiographs revealed multiple osteochondromas in the femur, humerus, ribs and tibia (Fig. 1.A&B). CT scan showed a bony tumor arising from the right pedicle of the L4 vertebra. The cortex and medullary cavity of the tumor were in continuity with the pedicle of the L4 vertebra. The spinal canal was narrowed by the tumor. The central portion of the tumor showed an increased intensity signal on MRI and was surrounded by peripheral ossification and a cartilage cap of low signal intensity on T1-and T2 weighted MRI. MRI indicated compression of the right L5 nerve root (Fig. 1.C-E). The tumor was surgically resected via a right interlaminar approach. There was no adhesion observed between the tumor and the nerve root or dura mater. After resecting the tumor, we confirmed that the L5 nerve root was fully decompressed. The gross appearance of the resected tumor was composed of cartilaginous and osseous components. Histological study confirmed the diagnosis of benign osteochondroma (Figs. 2A&B). The patient became asymptomatic immediately after surgery and the postoperative course was uneventful. The 4-month post-operative radiograph and CT scan showed complete tumor resection without recurrence (Fig. 2C).

Discussion

Osteochondroma occurs as sporadic (solitary) or multiple. The multiple form is usually in the context of the hereditary syndrome.[1] Conflicting reports exist as to whether solitary exostosis or lesions associated with HME more commonly affect the spine. However, according to the reported study by Albrecht et al., 75% of the spinal exostoses are solitary.[9]

Interestingly in patients with HME, spinal lesions are usually solitary despite presence of multiple exostoses.[7] HME is usually transmitted as an autosomal dominant gene with a variable penetrance.[4] The family history is positive in only 60% of the patients. A mutated gene is the possible cause of the disease in those cases in which the familial history is not positive.[4]

Some authors describe a predilection for men in hereditary cases[4,8] and Albrecht et al. defined a male predominance of 2.5:1 in a separate review.[9]

Although the incidence of spinal involvement is 7%-9%,[4,8] the real occurrence of the lesions is unknown because asymptomatic lesions are not usually diagnosed.[5,] Only 0.5%-1% of the spinal osteochondromas may cause neurological symptoms.[10] The main reason that the lesions are asymptomatic is because they almost always grow out of the spinal canal.[8]

Although osteochondromas originate from all parts of the spinal column,[2] the cervical region is the most common location in the spinal column while the lower thoracic and lumbar region rarely get in-

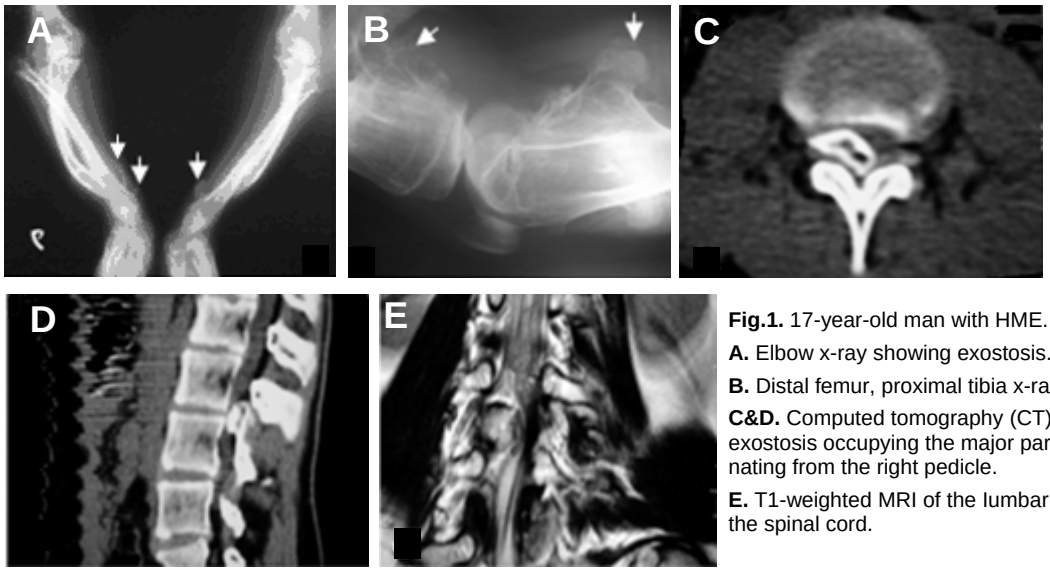

Fig.1. 17-year-old man with HME.

A. Elbow x-ray showing exostosis.

B. Distal femur, proximal tibia x-ray displaying exostoses.

C&D. Computed tomography (CT) scan through L4, showing exostosis occupying the major part of the spinal canal and originating from the right pedicle.

E. T1-weighted MRI of the lumbar spine; showing compression of the spinal cord.

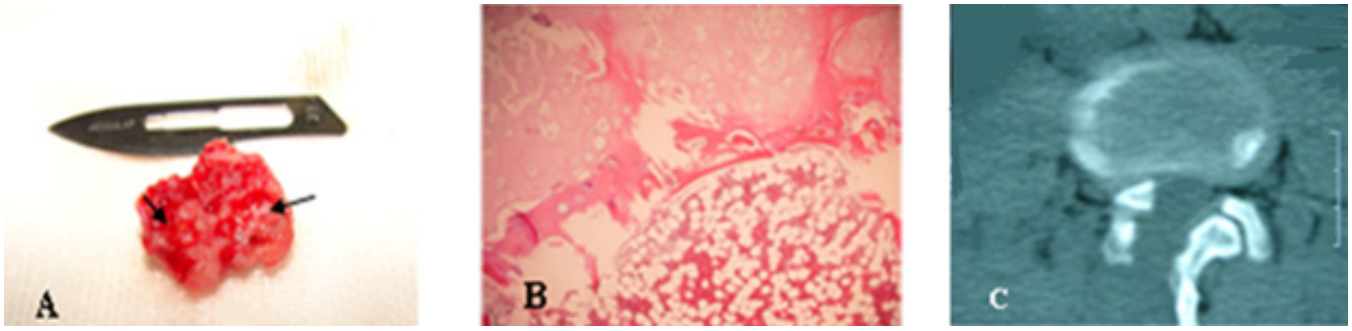

Fig. 2. A. Photograph of the en bloc resected tumor cartilage cap (arrows)

B. Micrograph of histologic section of the tumor demonstrating a cartilage cap (pink) and enchondreal ossification in the central part (purple). The base of the tumor contains bone marrow (red). This picture is suggestive of an osteochondroma

C. Postoperative lumbar CT scan, defect of partial laminectomy

volved.[8]

The reported cases of osteochondroma of the lumbar region mentioned in the English literature are summarized in Table 1. Osteochondromas are not present at birth and appear gradually.[11] The neurological symptoms resulted from progressive compression on the neurological elements are due to the osteochondromas that enlarge gradually. Although in some cases initiation of the symptoms is acute, this occurs as a result of sudden hyperextension of the spinal column or maybe after falling down.[12]

The duration of the symptoms ranged from 8 days to 33 years (average 2 years and 6 months),[4] and usually became symptomatic during the second or third decades of life.

In the vertebral column the posterior elements are where exostosis usually originates, as it is the location of secondary centers of ossification.[13] Absence of an epiphyseal plate is the reason osteochondroma is rare within the vertebral body.[14] The pathogenesis of multiple exostoses has been controversial with many theories mentioned. One of these theories includes the structural/mechanical theory, which emphasizes that the osteochondroma arises in the displaced growth plate cartilage, penetrating a defective periosteum[15] or in other words originates through lateral displacement of a portion of the epiphyseal growth cartilage.

It can be speculated that the more rapidly the ossification process of these centers develops, the greater the probability of aberrant cartilage formation will be. Therefore, the fact that osteochondroma is more frequently located in the upper segments of the vertebral column could be explained by different durations of ossification processes in these centers.[8]

Albrecht et al.[9] believe that the other reason for the higher occurance of the lesions in the cervical region is related to the greater mobility of the vertebra which causes micro trauma inflicted on the epiphyseal cartilage and displacement of a portion of this cartilage leading to exostosis formation.

According to Fiumara et al.[8] the incidence of osteochondroma found in the lumbar region is defiantly more rare. Therefore, the relationship between the incidence and the location of this tumor in the vertebral column remains unclear.

Spinal osteochondromas are more difficult to detect on plain radiographs because of the complex image formed by the spine.[3,5] In our patient's plain radiograph, no abnormality was detected. Myelography

Table 1. Reported Cases of Lumbar Osteochondroma in Hereditary Multiple Exostoses

Author (Reference)	Year	Age	Gender	Family History	Level	Origin	Duration	Surgery	Outcome
Gokay [20]	1955	24	F	+	L3	Lamina	5 years	Yes	Good
Urso[4]	1977	9	M	+	L4	Lamina	7 mon	Yes	Good
El Quessar[21]	1998	13	M	+	L1	Body	_____	Yes	Good
Fiechtl[2]	2003	8	F	+	L4	Posterior elements	2 years	Yes	Good
Bess[10]	2005	34	F	+	L5-S1	Facet	_____	Yes	Good
Present Case	2008	17	M	+	L4	Pedicle	2 mon	Yes	Good

will show evidence of extradural compression in almost all patients with neurological signs and symptoms,[3] but because it is an invasive modality, it is not used very often. Diagnosis is performed with CT scan and MRI, of which CT scan is the diagnostic imaging modality of choice.[3,5] It shows the extent of the cartilaginous and osseous components and MRI is more useful than CT in defining the extradural component and cord compression.[5] MRI displays the spinal canal mass. The lesions show a prominent peripheral rim of low intensity corresponding to ossification and a small central core of intermediate signal similar to that of bone marrow, giving the mass a "bull's eye" appearance.[16] Also the cartilaginous cap is best visualized on MRI.[10] Malignancy which is an important complication of osteochondroma may be determined by the thickness of the cartilaginous cap.[14] In spinal lesions the cartilaginous part is thin, so this part cannot be seen on MRI. Although those lesions generally do not manifest contrast uptake, cases of unusual gadolinium uptake have been detected.[17] Sometimes mild contrast enhancement may be present within the marrow.[16] Other lesions in the skeleton can be identified by nuclear medicine bone scan.[18]

From the treatment point of view, asymptomatic lesions can be followed without surgical intervention,[10,14] whereas symptomatic lesions are treated surgically.[14] Delay in diagnosis and treatment may cause irreversible neurological impairment.[19]

Standard surgical approaches are used for various spinal levels. This may necessitate wide laminectomy, partial facetectomy and sometimes stabilization.[10] Where feasible, complete excision is recommended as a treatment option, incomplete removal of the cartilage cap may lead to tumor recurrence.[3] Intra-lesional excision of the exostosis is associated with an unacceptably high recurrence rate and should be avoided.[10] The recovery of neurological function after surgical treatment is excellent.[10] In all cases of osteochondroma of the lumbar region in which surgical treatment has been performed, the results have been satisfactory.

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