Arch Orthop Trauma Surg (2006) 126:654–659
DOI 10.1007/s00402-006-0211-9

ORTHOPAEDIC SURGERY

Osteochondroma of C7 vertebra presenting as compressive myelopathy in a patient with nonhereditary (nonfamilial/sporadic) multiple exostoses

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Received: 24 April 2006 / Published online: 12 August 2006

Abstract
Introduction Osteochondromas are most commonly found in the appendicular skeleton. They occur less frequently in the spine and compression of the spinal cord is very rare. To the best of our knowledge, this is the first report of an osteochondroma arising from C7 vertebra presenting with compressive myelopathy in a patient with nonhereditary multiple exostoses. Our purpose is to report this rare presentation and its successful management, and to highlight the clinico-radiological features of this treatable condition.
Materials and methods A 20-year-old male with nonhereditary exostoses presented with gradual onset weakness in bilateral lower limbs, which had rapidly progressed to complete paraplegia over 1 month. The tumor was effectively treated by surgical excision along with spinal decompression. The diagnosis was confirmed by histopathological evidence complemented by clinico-radiological studies.
Results There has been a complete functional recovery without any evidence of recurrence at last follow-up at 2 years.
Conclusion Compressive myelopathy due to an osteochondroma arising from C7 vertebra in a case with nonhereditary multiple exostoses is being reported for the first time. Both CT and MRI demonstrated the origin, size, extent and relationship of the tumor to the vertebral and neural elements. Complete recovery of functions after surgical decompression was achieved in this case. An osteochondroma of spine must always be considered in all patients with multiple exostoses who have spine pain or develop neural deficit.

Keywords Spinal osteochondroma · Cord compression · Compression myelopathy · Cervical spine myelopathy · C7 vertebral osteochondroma · Nonhereditary (sporadic/nonfamilial) osteochondromatosis · C7 exostoses

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Introduction

Osteochondromas are most commonly found in the appendicular skeleton. Their presentation may be solitary or multiple. The multiple form is less common and is most often inherited as an autosomal dominant trait called hereditary multiple exostoses (HME) in up to 74% of cases. However, the multiple form can also occur sporadically (nonhereditary/nonfamilial) [2, 3]. Neurological complications due to exostosis are uncommon and mostly involve the peripheral nervous system. Involvement of the spinal cord from a vertebral exostosis is indeed rare and is usually discovered only after cord compression [2, 3, 4, 7, 10]. To the best of our knowledge, there is no report of an osteochondroma arising from C7 vertebra presenting with

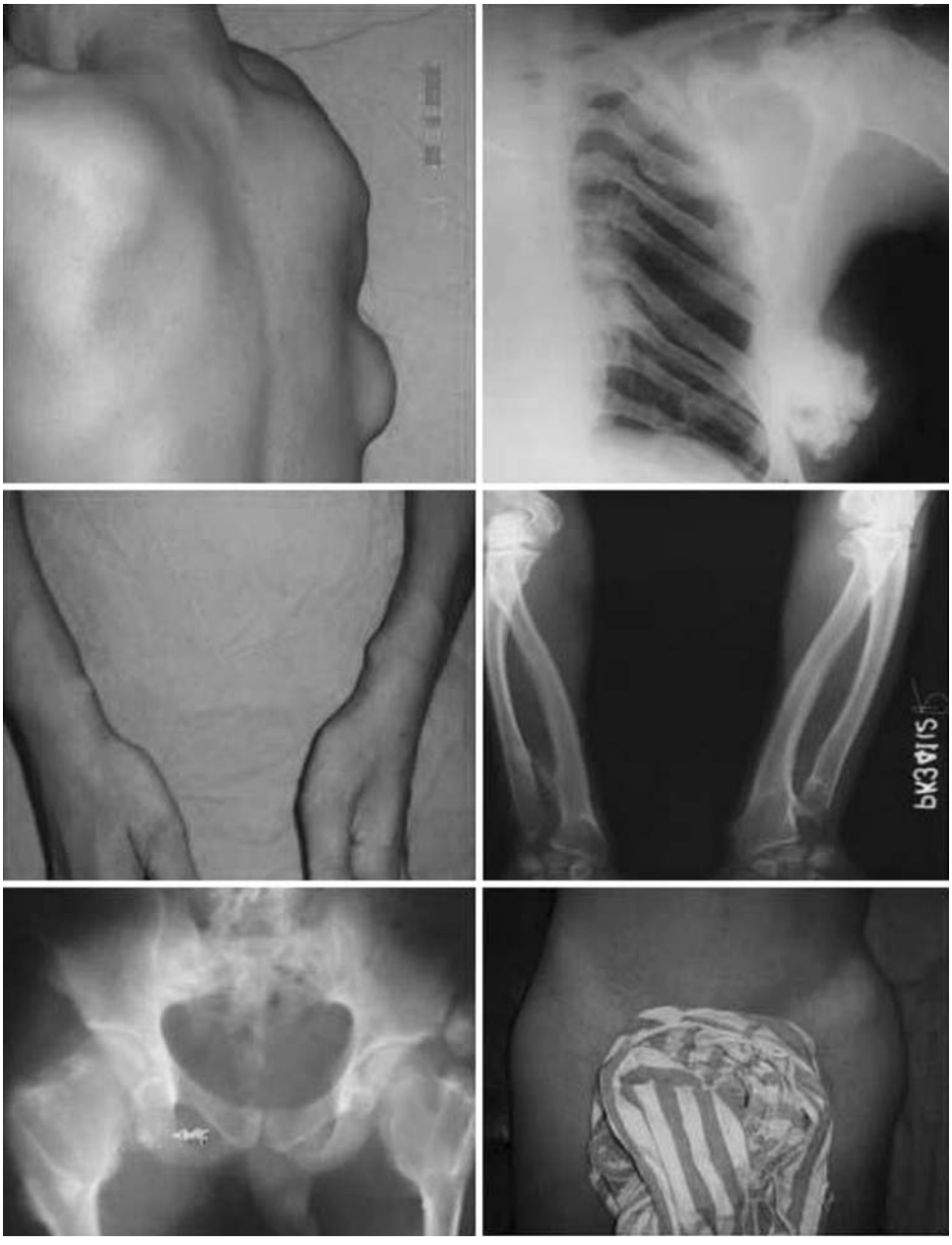

Fig. 1 Clinico-radiological photographs showing some of the multiple osteochondromas present in this patient (arising from the right 7th rib, bilateral radius and ulna, and bilateral proximal femur)

compressive myelopathy in a patient with nonhereditary multiple exostoses. Our purpose is to report this rare presentation and its successful management, and to highlight the clinico-radiological features of this treatable condition.

Case report

A 20-year-old male presented with gradual onset weakness in bilateral lower limbs, which had rapidly progressed to complete paraplegia over the last 1 month. His general health was otherwise good with no other significant past or medical history. There was no family history of tumors, masses or orthopedic related diseases. Physical examination revealed multiple asymptomatic, almost symmetrical bony masses along the flat as well as long bones. These had been present since early childhood, grew with age and had recently stopped growing (Fig. 1). The cervical spine was nontender but the movements were avoided by the patient. Higher mental functions and cranial nerves were clinically unremarkable. Muscle tone was increased in all four limbs with exaggerated deep tendon reflexes and an extensor plantar response. There was bilateral weakness at and below C6 myotome with complete motor loss below T2 level. No sensory deficit was elicitable in lateral or posterior columns. There was no bladder or

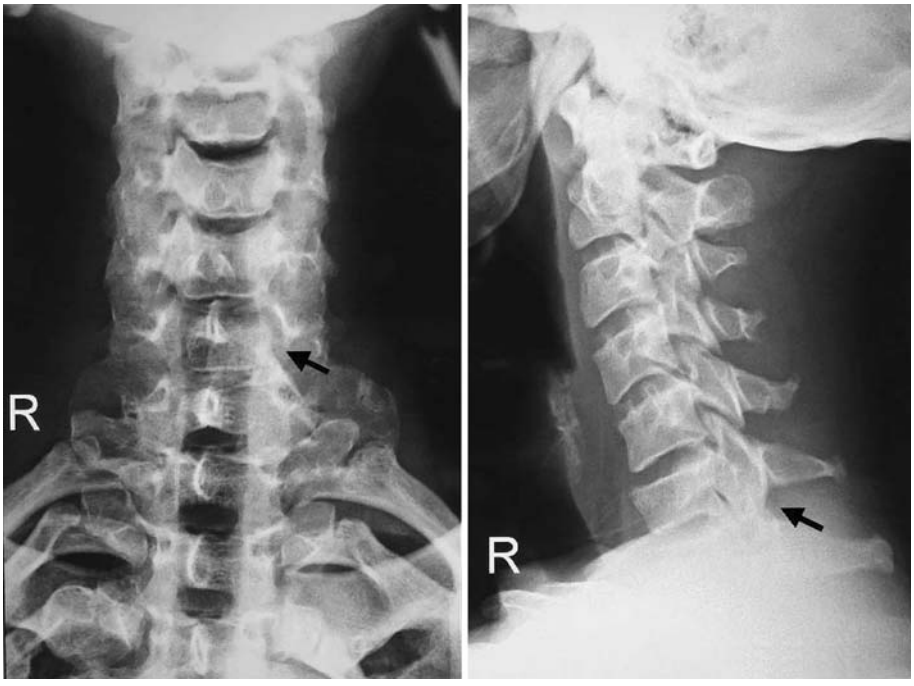

Fig. 2 X-ray *AP* and *lateral* views showing suspicious homogenous ill-defined opacity at left side of C6-7 level

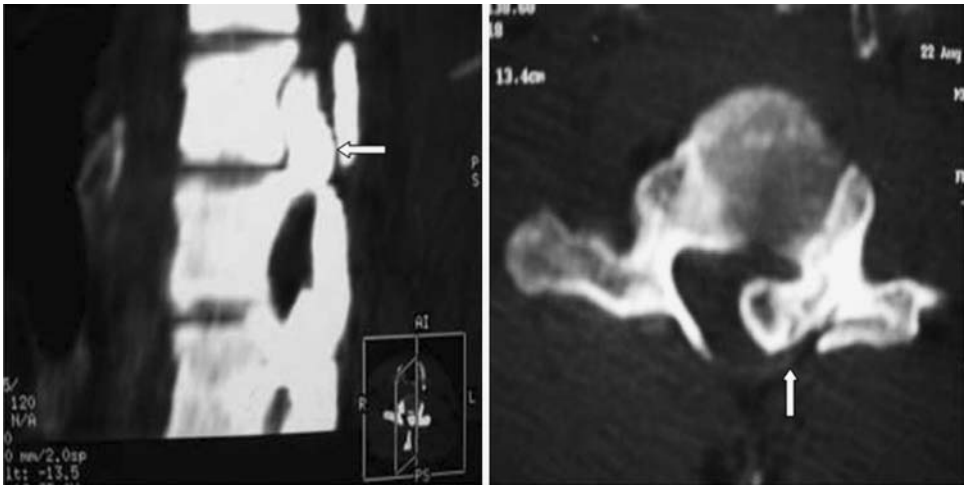

Fig. 3 CT scan (*sagittal* and *transverse*) showing an osseous mass arising from the left pedicle of C7 vertebra with trabecular continuity and growing rostrally in the canal till C6 level

bowel involvement. Plain X-rays showed a suspicious, homogenous, ill-defined opacity at the left side of C6-7 level (Fig. 2). A CT scan demonstrated an osseous mass arising from the left pedicle of C7 vertebra with trabecular continuity and growing rostrally in the canal till C6 level (Fig. 3). An MRI showed an extradural space occupying lesion in the canal at C6-7 level, severely compressing the cord (Fig. 4). Based on the clinico-radiological features a provisional diagnosis of spinal osteochondroma was made.

A decompressive laminectomy was performed at C6–7 level by the senior author (AKJ). The tumor was found to occupy most of the canal displacing the non-pulsatile bluish cord to the right (Fig. 5). Without direct cord handling, the tumor was excised gently, first in piecemeals and then along with its cartilaginous shell. The spinal canal was thoroughly decompressed (Fig. 5). No fusion was performed since the facet joints were preserved. Macroscopically, the tumor consisted of a $2 \times 1 \times 0.5$ cm mass with a grayish blue cartilaginous cap. Microscopically, the lesion was consistent with osteochondroma with no signs of malignant degeneration (Fig. 6). The patient started improving the same evening and was mobilized with a SOMI brace support as his neural improvement continued. By the end of 2 weeks he had regained full power, though still spastic. By the end of 12 weeks he was ambulating independently, but the extensor plantar response was still persisting. There is no recurrence of signs and symptoms at 2 years postoperative and the

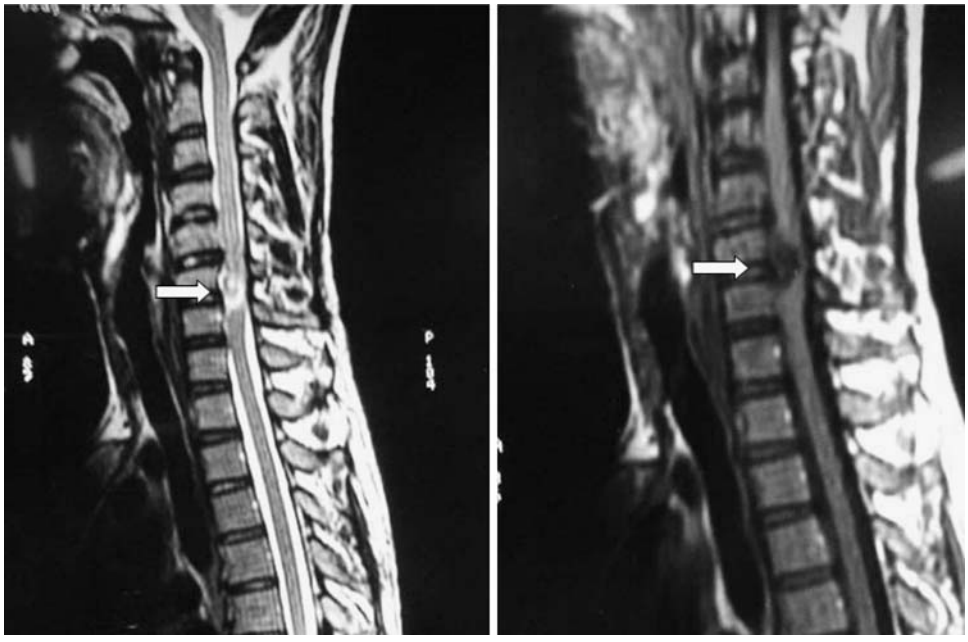

Fig. 4 MRI (T2WI & T1WI) showing an extradural space occupying lesion in the canal at C6-7 level, severely compressing the spinal cord

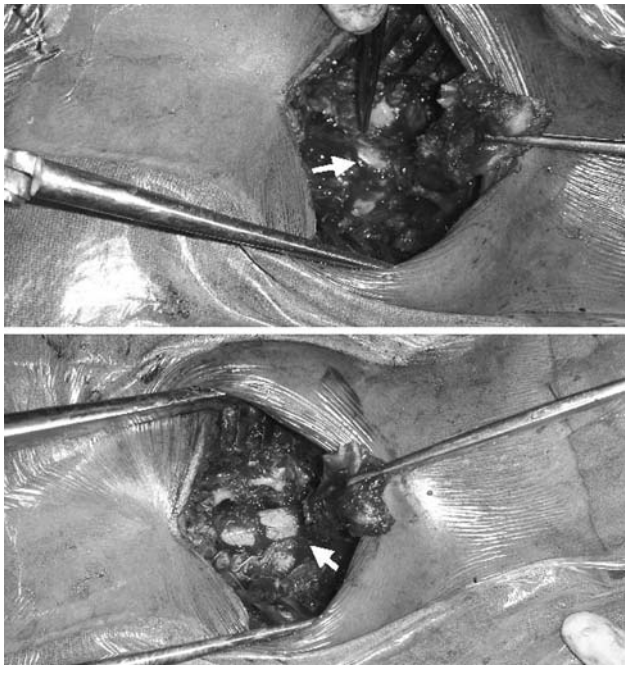

Fig. 5 Peroperative photographs showing the spinal cord compression (*above arrow*) and the decompressed cord after the tumor excision (*below arrow*)

patient is on regular follow-up in lieu of increased susceptibility of malignant changes due to multiple exostoses.

Discussion

Osteochondromas represent the most common bone tumors and account for about 8.5% of all bone tumors and 36–40% of all benign ones [2, 3, 5]. Though described in every bone, they are most frequently found in the humerus (20%) and around the knee (40%) [8]. Only 1–4% of solitary osteochondromas arise in the spine, and 7–9% of patients with HME develop a spinal lesion [4]. Soloman's series of 52 patients with HME had only a 9% incidence of spinal lesions and all were asymptomatic [10]. Thus, compression of the cord is extremely rare and has been reported in only 0–2.5% of patients of HME with spinal lesions, and is still rarer in cases of solitary lesion [7]. However, no data is available for nonhereditary multiple exostoses with spinal cord involvement, and thus, most of the assumptions are based on HME. The exact incidence is still unknown because asymptomatic cases may have never been documented [7]. Patients with multiple exostoses tend to come for treatment at a younger age and have a higher rate of spinal compression. Usually the tumor takes origin at the neural arch and is most frequently localized close to the secondary ossification centers, commonly near the tip of spinous or transverse processes [2, 4, 8]. Nevertheless, they can originate from any part of the vertebrae. About half of spinal osteochondromas are located in cervical spine, the most frequent site being C2 (50%), followed by C3 and C6. This constitutes a significant overrepresentation since the cervical spine contains only 23% of all vertebrae. This uneven distribution may be related to the mobility of the various segments of the spine; the cervical spine being the most flexible. Greater mobility and flexibility are associated with greater stress on the vertebrae, which would lead in turn to microtrauma to the epiphyseal cartilage [7].

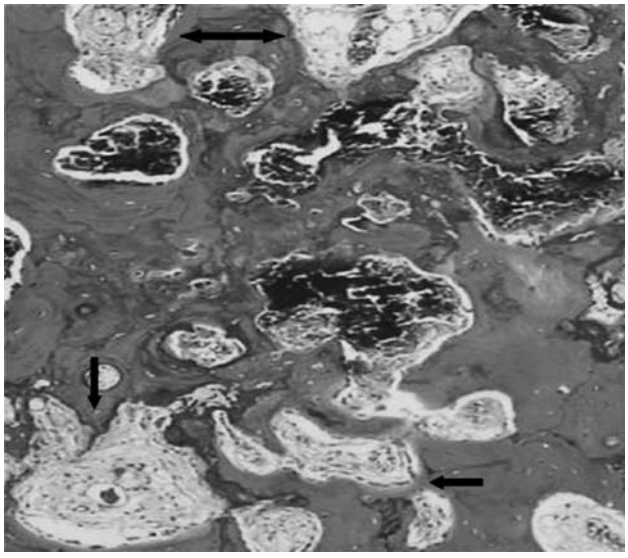

Fig. 6 Histopathological slide (H&E stain ×20) showing enchondrial ossification with no evidence of malignant degeneration

Vertebral osteochondromas usually become symptomatic in the second to third decade of life with a male predominance, although they have been described as late as the eighth decade [2, 4, 6, 8, 9]. The clinical manifestations vary widely [1–4, 6–11] and can take the form of vertebral pain, of a palpable mass that may or may not be painful, and of neurological deficit in form of myelopathy or radiculopathy. Even sudden death due to transection of cord at odontoid level has been reported. Bulky tumors arising from the ribs, if close to the spine can also cause myelopathy. Unusual cases may present with Horner's syndrome, Brown Sequard syndrome, cauda equina, diaphragmatic perforation and intestinal obstruction, sciatica or even Freidreich'c ataxia. Anterior extension can cause anterior or oropharyngeal mass, hoarseness or dysphagia, obstructive sleep apnea and even obstruction of the arteries. The duration of symptoms varies widely from weeks to many years [4]. Recurrence or a sudden acceleration in the growth may herald a malignant transformation, which occurs in about 1% of solitary and 10% of multiple lesions [2, 7, 8]. Differentials include osteoblastoma, giant cell tumor and aneurysmal bone cyst. Special attention must be paid to another familial disorder, the Gardner's syndrome, which is associated with multiple colonic adenomas, soft tissue hamartomas, and osteomas or osteochondromas, because of a high probability of malignant transformation in the colonic adenomas.

Because of the complex image of the spine on plain radiographs, prospective interpretation of these lesions has been inconclusive in up to 79% of the cases [2]. Plain radiographs may show calcification but provide limited information. Computerized tomography, preferably with myelography, is the imaging modality of choice. It demonstrates a bony mass with a sharply defined periphery, a more lucent but organized center with cortex and medullary cavity continuous with the bone of origin and a thin cartilaginous cap. MRI is useful to show the level and the extent of neural compression along with the marrow content and the cartilaginous cap. It usually shows 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 [8]. A bone scan is helpful to screen for other lesions as approximately 50% of patients with osteochondroma of cervical spine will have multiple lesions [2].

Treatment should be undertaken as soon as the tumor becomes symptomatic, or is suggested for cosmetic reasons [2, 7, 8]. In case of a rapid increase in size, surgical treatment is mandatory because of the possibility of a malignant degeneration. The surgical treatment should be nearly as total as possible without creating functional defects; recurrence is rare even when excision is incomplete though it has been reported as late as 14 years [2]. The radioresistance of the tumor leaves no place for radiotherapy [7]. Most patients had complete disappearance of their deficits or had only minor residual deficits after surgical decompression [1–4, 6–11]. Even patients with significant and long-standing deficits (up to 9 years) have improved [2].

In our case, symptoms and neurological signs progressed quickly enough so that there was minimal delay in presentation. During surgery, the main difficulty is due to the bony hard consistency of the tumor, which often calls for use of massive hardware in the immediate vicinity of delicate neural structures. Any maneuver which might increase the spinal cord compression, such as rocking movement of the exostosis and the insertion of rongeurs beneath the lamina must be avoided. The signs and symptoms resolved promptly after surgery with no recurrence at last follow-up.

Conclusion

Compressive myelopathy due to an osteochondroma arising from C7 vertebra in a case of nonhereditary multiple exostoses is being reported for the first time. Both CT and MRI demonstrated the origin, size, extent and relationship of the tumor to the vertebral and neural elements. Satisfactory improvement and recovery of functions after surgical decompression was achieved in this case. An osteochondroma of spine must always be

considered in all patients with multiple exostoses who have spine pain or develop neural deficit.

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