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R. Ergün
A. İ. Ökten
E. Beşkonakli
G. Akdemir
Y. Taşkin

Cervical laminar exostosis in multiple hereditary osteochondromatosis: anterior stabilization and fusion technique for preventing instability

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R. Ergün · A. İ. Ökten · E. Beşkonakli
G. Akdemir · Y. Taşkin
Department of Neurosurgery,
Ankara Numune Hospital, Ankara, Turkey

R. Ergün (✉)
Güçlüler Sokak no. 1/7, Cebeci,
06340 Ankara, Turkey
Tel. +90-312-362 4963

Abstract Multiple hereditary osteochondromatosis is a genetically transmitted disorder consisting of multiple projections of bone capped by cartilage, which are called exostoses. Spinal cord compression due to expansion of a laminar osteochondroma is rare but well recognized. Surgical decompression usually improves the patient's neurological status but, in cervical exostosis, postlaminectomy kyphosis and instability problems, especially in the high-risk adolescent group, form the most significant potential difficulties in the postoperative period. We report a case of cervical laminar exostosis that was treated by anterior stabilization and fusion and discuss the benefits of this technique.

Key words Cervical exostosis · Osteochondroma · Postlaminectomy kyphosis · Stabilization

Introduction

Osteochondroma is a cartilage-capped bone projection that originates in any part of endochondral bone formation. It originates from within the periosteum and grows progressively by endochondral ossification of its cartilaginous cap in a fashion that mimics physeal growth, which is an aberrant cartilage of the growth plate. Many authors therefore consider them to be developmental anomalies rather than true tumors [15].

Osteochondromas constitute 8.5% of all primary bone tumors and 36% of all benign bone tumors. They occur in two different patterns: as a solitary lesion with no genetic component, or as multiple lesions in the form of the genetic disorder known as multiple hereditary osteochondromatosis (MHO). MHO was first described by Stanley in 1849, and its hereditary nature was documented by Stocks and Barrington in 1925, who reported a family history in 65% of cases [10]. MHO has many synonyms coined in recent years, such as hereditary multiple exostoses, multiple congenital osteochondromata, diaphyseal aclasis, dyschondroplasia, cartilaginous exostoses, exostotic dysplasia, and osteogenic disease [14]. The disease is transmitted as a single autosomal dominant gene, and occurs more often in males, demonstrating a male/female ratio of about 1.5/1 and appearing most commonly before the age of 20 years [10]. The prevalence of this condition worldwide is unknown and difficult to estimate from available data. Patients with multiple osteochondromas account for approximately 12% of all cases [4]. Exostoses most often involve the metaphysis of long tubular bones, but can be found in any bone formed by enchondral ossification. Tubular bones such as long bones of extremities (95%), iliac crest (62%), and ribs (42%) are the most frequently affected bones, with involvement of vertebrae (9%) and carpal and tarsal bones (7%) being more rare [12].

We report the case of an MHO patient with a huge cervical laminar exostosis that extended through from C3 to C6. The clinical symptoms and signs, radiological investigations, and surgical treatment including the technique of anterior stabilization and fusion are reviewed.

Case report

A 16-year-old boy had noticed multiple swellings on his four extremities, right shoulder, and neck, since the age of 7 years, which were totally asymptomatic. He later experienced progressive motor weakness that involved all four limbs, which had developed over a period of 1 year. Similar multiple lesions were also revealed in his family members (his father and one of his brothers).

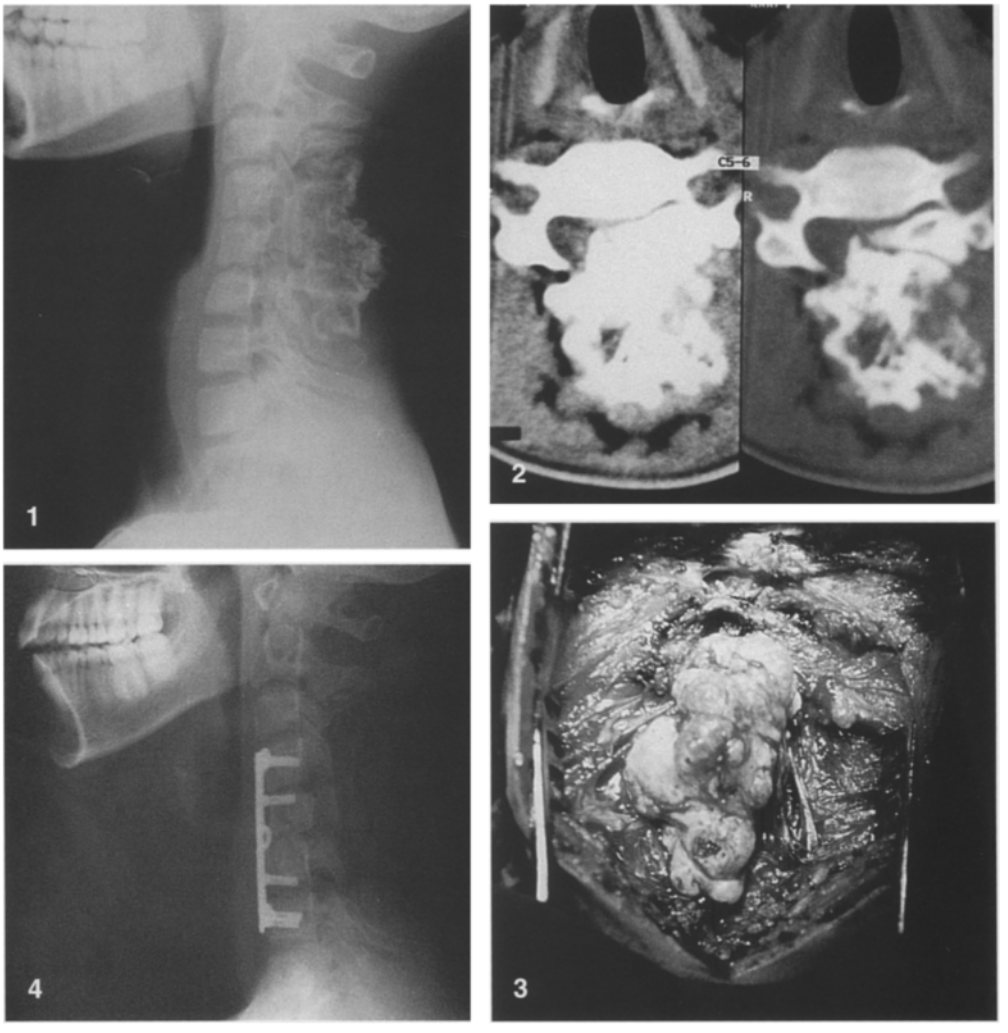

Fig. 1 Lateral plain radiograph of the cervical spine shows a well-delineated lesion between the levels of C3 and C6

Fig. 2 Axial CT scan demonstrates a heavily calcified irregular mass that stenosed the vertebral canal at the level of C5-C6

Fig. 3 Intraoperative view of cervical osteochondroma with cauliflower-like appearance

Fig. 4 Postoperative lateral plain radiograph of the cervical spine showing that the mass was removed and anterior stabilization and fusion were performed

Detailed neurological examination revealed quadriparesis – the power of the left upper extremity was grade 3 of 5, the right upper extremity grade 4 of 5, the left lower extremity was grade 3 of 5 and the right lower extremity was grade 4 of 5 – associated with moderate spasticity. There was significant hypoesthesia over the trunk and four extremities and loss of position and vibration senses on sensory examination. Deep tendon reflexes were hyperactive in all extremities. There were clonus, Hoffman, and Babinski signs bilaterally.

Plain cervical spine radiographs demonstrated a well-delineated lesion with scattered calcifications at the levels of C3, C4, C5, and C6 involving spinous processes. The height of the vertebral bodies and the cervical lordosis were well preserved (Fig. 1). CT revealed a heavily calcified mass between the posterior elements of C3 and C6, which extended into the vertebral canal, compressing the cord laterally. The tumoral mass mostly involved the left pedicles and intervertebral joint at the level of C5-C6. The vertebral canal was nearly 80% stenosed (Fig. 2).

The patient underwent surgery in sitting position by a standard posterior midline exploration. The cartilage-capped mass had the convoluted cauliflower-like appearance characteristic of an osteochondroma (Fig. 3). He underwent posterior decompressive laminectomies of C3 through C6 and the tumor was totally removed. The cord was found to be free and pulsatile, indicating the adequacy of the decompression procedure. The operation was then continued in supine position using the standard anterior approach to cervical spine. After C5-C6 anterior discectomy and fusion (Cloward operation) by cortico-cancellous bone grafting, which was taken from the anterior iliac crest, anterior stabilization of C4 through C7 vertebral bodies was performed using the Cervical Spine Locking Plate system (Synthes Spine, Paoli, Pa.), including a titanium plate implant with six titanium Morscher screws (Fig. 4).

Histopathological examination of the tumor showed features of an osteochondroma composed of hyaline cartilage cap of variable thickness and subchondral bony trabeculae separated by fatty marrow.

The postoperative course of the patient was uneventful. On follow-up examinations 18 months later, his right hemiparesis had improved to normal, on the left side the power of the extremities was grade 4 of 5.

Discussion

Involvement of the spinal cord from a vertebral exostosis is indeed rare. It can affect any part of the vertebral column; it originates from the neural arch and is most fre-

quently located close to secondary ossification sites, commonly near the tip of spinous or transverse processes [8]. Nevertheless, osteochondromas can take their origin from any part of the vertebra such as intervertebral joints, pedicles, spinous processes, costovertebral articulations, and vertebral body. Osteochondromas of the spine are located mainly in lumbar (34%), followed by cervical (23%), sacral (7%), and both lumbar and sacral (1.5%) regions [2].

Spinal osteochondromas usually cause a variety of signs and symptoms including those of spinal cord or root compression. These are the result of progressive encroachment of the slowly expanding mass on neural structures. The tumoral lesion can be well demonstrated by radiological investigations using plain radiographs, CT, and MRI. Plain roentgenograms are often but not always, diagnostic. Prior to skeletal maturity, the lesion has a hyaline cartilage cap that is undetectable on plain radiography if less than 2.5 cm thick [9]. MRI is more useful in defining an extradural intracranial component of the tumor, the importance of compression of the neural structures, and the true levels of the lesion, but it will not show precisely the extent of the tumor [5]. CT not only demonstrates the cartilaginous and osseous components of the tumor, but also defines clearly its extension and relationship to the vertebral and neural elements of the spine [1, 6, 11].

The osteochondroma grows at its tip as the cartilage undergoes ossification, and if the cartilaginous cap is incompletely resected, it can recur. The rate of recurrence has been reported to be nearly 50% in incompletely resected cases. This has been described in the literature to occur as early as 6 months [13] and as late as 14 years postoperatively [17]. Recurrence or a sudden acceleration in the growth of an osteochondroma may herald malignant transformation, usually chondrosarcoma. This occurs in about 1–2% of solitary osteochondromas, but in MHO cases the reported risk is higher, at 5–25% [7, 16]. The radioresistance of this tumor leaves no place for radiotherapy [2]. As malignant degeneration and recurrence risks are the most significant potential problems in MHO patients, careful postoperative follow-ups are essential.

The other important complication in the postoperative period, especially in cases of multiple-level posterior laminectomies, as in our case, is development of a progressive postlaminectomy kyphosis and subsequent instability in the high-risk adolescent patient group. We do not recommend posterior stabilization, which was reported recently in literature [3], because of the potential risks of recurrence or malignant transformation associated with the tumor. In conclusion, we strongly recommend the technique of anterior stabilization and fusion, especially in similar cases affecting adolescents, who have a high risk of instability, particularly in view of the possibility of recurrent operations.

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