A rare case of a giant cervical osteochondroma.

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Abstract

Background: Osteochondroma or osteocartilaginous exostosis is the most common benign tumor of bone. They account for 35% to 50% of benign bone neoplasms and 10% to 15% of all primary bone tumors. Usually these tumors are found in the appendicular skeleton, especially in the metaphyseal region of long bones. Spinal osteochondromas, however, are uncommon especially at cervical level.

Clinical presentation: We report the case of a 16 year old boy presenting a hard, gradually progressing, large swelling mass, over the posterior part of the neck causing pain. Radiological images revealed a giant solitary osteochondroma arising from C3 to C6 laminae. The patient underwent complete surgical excision of the tumor. Postoperative outcome was good and recovery from the pain achieved.

Conclusion: Solitary osteochondroma of the cervical spine is a rare manifestation of a common bony tumor. Patients affected may present with a multitude of symptoms and complications depending on the size and location of the lesion. Advanced imaging modalities are required for characterization, diagnosis, surgical planning and management of these rare cervical lesions.

Keywords: Osteochondroma, Exostosis, Cervical spine, Bone tumor, Spine surgery.

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Introduction

Osteochondroma, also known as osteocartilaginous exostosis or just exostosis, was initially described in 1843 by Reid [1]; it is a common benign tumor of the bone accounting for 10% to 15% of all bone tumors and 20% to 50% of all benign bone tumors [2]. These lesions are the most common benign tumors of the appendicular skeleton, located in the metaphyseal region of long bones, particularly the distal femur and proximal tibia while occur less frequently in the axial skeleton [3-5]. They occur in a solitary or multiple form; also called osteochondromatosis or hereditary multiple exostosis (HME) [6-8]. Spinal involvement accounts for approximately 3% of cases [5-9]. Any portion of the vertebral body may be affected. Spinal osteochondromas represent 1.3% to 4.1% of all osteochondromas and for less than 4% of spinal neoplasms [3,10-15]. The role of the genes or of the immune system in the pathogenesis of osteochondroma seems to be less understood as compared to other diseases so as far for neoangiogenesis [16-20].

Approximately 7% of hereditary osteochondromas occur along the vertebral column, which is the double of the rate of sporadic spinal involvement [21]. Anyway vertebral osteochondromas rarely cause spinal cord compression [2,5]. Cervical osteochondromas usually involve C1 and C2 vertebrae while subaxial cervical spine localization (C3 and below) is quite rare [4,22].

The age of onset for spinal osteochondromas is the second and third decade of life and there is a male predominance (M: F ratio of 2.5:1) [4,9,14].

Spinal osteochondromas may produce a wide variety of symptoms and complications depending on their location and relationship to surrounding structures. These may include compressive myelopathy, nerve root compression, pathological fractures and malignant degeneration [8,14].

We present a rare pediatric case of a giant sporadic osteochondroma arising from C3-C6 spinous processes and laminae producing neck pain.

Case Report

A previously healthy 16 year old boy was referred to our Department because he noticed a gradually progressing hard posterior neck mass causing pain and discomfort. Pain developed when the neck was turned or when local pressure was applied. No family history of osteochondroma or other
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Abnormalities of the long-bones and appendicular skeleton was referred. Physical examination revealed an osseous protuberance, firmly attached to the cervical spine at the base of the neck slightly larger on the left. The mass seemed to be continuous with the cervical vertebrae and terminal ranges of neck’s movements were restricted. He denied numbness, tingling, or other neurologic symptoms: no motor deficit, sensory disturbance, or hyper reflexia was noted.

Computerized tomography scan of the cervical spine was done: it revealed a well-defined, pedunculated mushroom-shaped mass with a calcified cartilage cap, measuring about 35*26*37 mm, arising from posterior arch of the C3-C6 vertebrae (spinous processes and corresponding laminae) with corticomedullary continuity and bone remodelling (Figures 1a and 1b). There was no extension within the canal. Pre-and post-contrast magnetic resonance imaging confirmed the presence of the large multilobulated, cystic, and irregularly enhancing mass with calcified components (Figures 1c and 1d). The lesion was suspicious for a solitary osteochondroma.

The differential diagnosis for this spinal bone lesion included benign osteoma, osteoblastoma, osteosarcoma, meningioma, hypertrophy with calcification of the ligamentum flavum, unusual synovial cyst manifestation and chondrosarcoma.

Surgical treatment was chosen and performed. Under general anaesthesia and through a posterior-midline cervical approach the lesion was clearly visualized: it was lobulated, well-circumscribed with the top surface smooth (Figures 1b and 2a). The cartilaginous lesion was first resected using osteotomes and the residue lesion was then thinned down to the level of the laminae. A wide C3-C6 laminectomy without facet sacrifice was performed with en bloc resection of the tumor; there were no adhesions with the dura mater. Considering the patient’s normal cervical lordosis and facet preservation, fusion was not thought to be necessary. The resected piece was sent to the Pathology Department for the histopathological examination. As hemostatic agent the Vivostat® system (Vivostat A/S, Alleroed, Denmark) was used, forming an extremely thin white coat without compression of the neural structures [23-26]. A drain was applied for 24 h.

The results of a pathological examination were consistent with a benign osteochondroma with no evidence of any malignant transformation. The bony edges of the laminectomy margins were in healthy tissue.

The postoperative course was uneventful and the patient recovered soon after surgery; the pain immediately began to decrease and the neck movements to improve. The patient was discharged after 3 days. A new examination was performed one month later; he showed complete clinical recovery and the postoperative CT scan demonstrated a gross-total resection with stable sagittal alignment (Figures 3a and 3b). The patient was completely relieved of his symptoms until the latest follow up, conducted 1 year postoperatively.

Discussion

Spinal osteochondromas can be of two types: multiple osteochondromas in patients with hereditary multiple exostoses, autosomal dominant with full penetrance syndrome, and solitary osteochondromas occurring in the spine [4].

The pathophysiology of both remains unclear. Ionizing radiation could have a role and an association is found in 12 to 15% of cases [27].

Solitary osteochondromas are not considered true neoplasms: they originate from within the peristeme when a cartilaginous fragment grows progressively by enchondral bone formation [8]; this often happen at sites of rapid bone growth in the immature skeleton [7]. Therefore the lesion is composed of
ostochondromas and in less than 1% in the solitary forms. It occurs more commonly in axial degeneration, usually into a chondrosarcoma, is the most fearsome complication. It occurs due to the relatively increased mobility in this portion compared with the others and its predisposition of increased stress and microtrauma to the epiphysis and promoting exostotic growth [13,28]. Osteochondromas arise most frequently from the posterior elements of the spine such as the spinous, transverse, and articular processes but can also arise from the vertebral body, pedicle, and rarely the facet joints [12,13]. This tendency thought to be related to the abundance of secondary ossification centers within the neural arch [28].

As in our case, most solitary cervical spine osteochondromas do not produce neurological symptoms [10]; rarely do they cause radiculopathy or myelopathy. Symptoms and complications depend on location and relationship to associated structures and are the result of progressive encroachment of the slowly expanding osteochondroma [9,32]. The most frequent presentation is myelopathy, more common than radiculopathy and occurs more than twice as frequently in the familial form than the solitary osteochondroma group [10].

Often it evolves slowly and do not manifest until 20-30 years of age and for this reason represents a diagnostic challenge [2,33]. A palpable mass, local pain and cosmetic deformity may occur with neural arch lesions because these lesions tend to be large [8]. However, in some cases, the onset of symptoms is acute after sudden hyperextension of the spine or after a fall. In the literature some rare clinical presentations are described. Pharyngeal mass, as a calcified goiter, or with hoarseness or dysphagia due to compression of the oesophagus, larynx and trachea by anterior spinal osteochondroma in the neck have been described [8,14,21]. Cervical osteochondromas may produce Horner syndrome due to compression of the cervical sympathetic chain by lesion developing anteriorly into the lateral spinal nerve groove. Arnold’s (occipital) neuralgia has been seen in case of C2 involvement and a case of sudden death has also been reported in odontoid osteochondroma [34]. But also symptoms as headaches, cranial nerve palsy, vertigo or obstructive sleep apnea are described [15]. Finally, complications of cervical osteochondromas include, compressions of vertebral artery, subclavian artery and the common carotid artery, pathologic fracture and bursal and pseudoaneurysm formation [8,34,35]. In our case, there was no neurological deficit. However, there was a neck pain and movement limitation caused by the effect which arose from the large mass of the osteochondroma.

Apart from the risks of local compression, malignant degeneration, usually into a chondrosarcoma, is the most fearsome complication. It occurs more commonly in axial osteochondromas and in less than 1% in the solitary forms [6,8,22]. On MRI, a feature suggestive of malignancy is when the cartilage cap thickness is >2 cm in adults and >3 cm in children [2,6].

Diagnosis is difficult on plain radiographs because of the complex image formed by the spine in the area of the lesion; therefore multiple imaging modalities are often needed to detect spinal osteochondromas. CT is the imaging modality of choice to demonstrate cortical and medullary continuity in spinal osteochondromas and all others typical signs of spinal osteochondromas [8,12,13,34]. Magnetic resonance imaging is more useful in defining the intracanal extradural component of the tumor and the dural compression and to visualize the hyaline cartilaginous cap [8,12]. These lesions generally do not show contrast enhancement, although cases of unusual gadolinium uptake have been described. Furthermore, bone scintigraphy may be helpful in the assessment of osteochondromas and can evaluate for the presence of additional osteochondromas, which are found in approximately 50% of patients with a cervical spine osteochondroma [35].

Treatment of symptomatic cervical osteochondromas consists in surgical excision [13]. The surgical treatment should be total, including the cartilaginous cap and underlying intraspinal component. Usually a laminectomy with decompression of the neural elements has no major complications and gives good functional results [29].

Incomplete resection, for example in case of intralesional excision, is associated with a recurrence rate between 2% to 5% of cases [8]. If surgery causes instability due to sacrificing a significant portion of facets, consideration should be given to posterior stabilization by instrumentation [12].

Conclusion

Although unusual, cervical osteochondromas should be considered in the differential diagnosis of spinal tumors. These lesions have a wide spectrum of clinical presentations depending on the extent and on the relationship with surrounding structures. This case report discusses about a rare location in a pediatric patient of an osteochondroma arising from the C3-C6 spinous processes and causing neck pain. Treatment consists in total surgical excision.

References


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