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CASE REPORT

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Headache due to an osteochondroma of the axis

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Abstract We reported a case of a 42-year-old man with a 3-year history of headache due to a spinal osteochondroma. Repeated neurological evaluation, including EEG studies and CT of the cerebrum, revealed no pathology. More recently the patient presented with persistent headache and a slight limitation of neck motion. MRI studies of the cerebrum including the cervical spine showed a high cervical extradural tumor. Additional CT angiography showed a bony tumor suspected of being a spinal osteochondroma. An en bloc resection of the tumor was performed; histological evaluation confirmed the diagnosis. Immediately after intervention, all symptoms disappeared. In most patients with a spinal osteochondroma, the lesion causes no symptoms, or symptoms are aspecific. Therefore, there is often a significant delay between initial complaints and the diagnosis, as in the current case.

Keywords Osteochondroma · Vertebra · Cervical spine · Exostosis

Osteochondroma, or osteocartilaginous exostosis, is the most common skeletal neoplasm. It accounts for 36% of benign bone tumors and 8.5% of all bone tumors. Osteochondroma may be solitary or multiple. When multiple, there is a strong familial incidence, in which case, it is known as hereditary multiple exostoses (HME), an autosomal dominant disorder with variable penetrance [1, 3].

In this report, we present the clinical course of a patient seeking medical treatment over years for a severe, unexplained headache, caused by a solitary, cervical spinal osteochondroma.

Case report

A 42-year-old man presented with a 3-year history of severe headache. Over time, repeated analysis by means of neurological examination, EEG studies and a CT scan of the cerebrum did not show any abnormalities. The headache, located frontally, reacted partially to frequent pain relieving medication. More recently the patient presented with a limitation of neck movement, particularly rotation. Physical examination revealed slight limitation of neck rotation and increased reflexes at both arms and legs. Plain radiographs of the spine showed a synchondrosis/articulation between the neural arches of C1 and C2 at the right side (Fig. 1). Bone scintigraphy showed increased uptake high in the cervical spine. An MRI scan showed an extradural tumor at the level of the neural arches C1–C2 at the right side, giving clear deformation of the dural pouch and deviation of the myelum to the left. CT angiography and 3D reconstructions demonstrated an extradural bony mass that seemed to rise from the lateral mass on the pedicle of C2 (Fig. 2a, b and c). In addition, the slowly growing tumor produced a neoarthrosis between the tumor and the lamina C1 by compression. The mass filled approximately 50% of the spinal canal. There was no vascular abnormality. All imaging studies suggested either a hypertrophic osteoarthrosis of the right facet joint or an osteochondroma (Enneking stage 2). Surgery included a C1–C2 laminectomy followed by en bloc resection of the tumor (Fig. 3). Because the facet joint itself was left intact, posterior fixation was not necessary. Histological evaluation was consistent with an osteochondroma (Fig. 4). The surgical margins were wide. The postoperative course was uneventful. At follow-up 12 months postoperatively, the patient is in good health and does not complain of headache or limited neck motion. There is no evidence of local recurrence (postoperative CT studies). However, the lack of appropriate follow-up does not allow making definitive conclusions about the prognosis.

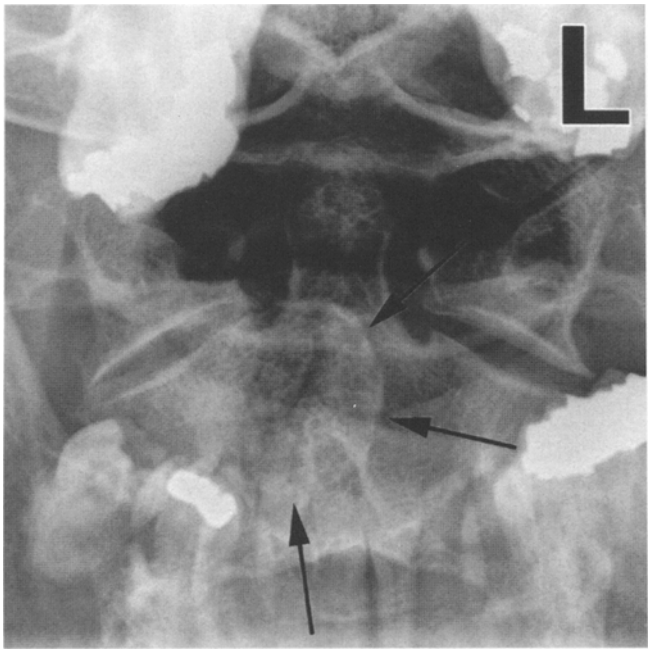

Fig. 1 Postero-anterior radiograph of the dens demonstrating an oval mass (*black arrows*) between C1 and C2 at the right side

Discussion

Only 1.3–4.1% of solitary osteochondromas originate in the spine. They make up 0.4% of intraspinal tumors or 3.9% of solitary spinal tumors. The prevalence of spinal involvement is higher in HME than in solitary exostosis (9%) [1]. The most common location of this tumor is in the posterior elements of the spinal column [3, 6]. The predilection for the neural arch could stem from the fact that it has numerous secondary ossification centers (e.g., spinous process, transverse processes and articular processes) that appear between the ages of 11 and 18 years [3]. The bulk of the tumor grows outside the spinal canal in the surrounding soft tissues of the neck [6]. In total 38 cases of cervical osteochondromas have been described. The C2 vertebra is the single commonest site of origin (28% of solitary lesions and 44% of multiple lesions) and approximately 70% of cases occurs in male patients [6]. Osteochondromas arise only in bone formed by enchondral ossification and consist of a bony outgrowth covered with a cartilage cap of variable thickness [2] (Fig. 3 and Fig. 4).

Patients in whom the osteochondroma has stopped growing at skeletal maturity with no symptoms are often not identified or not reported. Therefore, true frequency of spinal localization is probably underestimated [4]. However, some patients with spinal osteochondromas may present with a palpable mass, local pain or symptoms due to neurological or vascular compression [6]. Neurological complications, including spinal cord compression, myelopathy or nerve root irritation are rare [1, 7] and are even

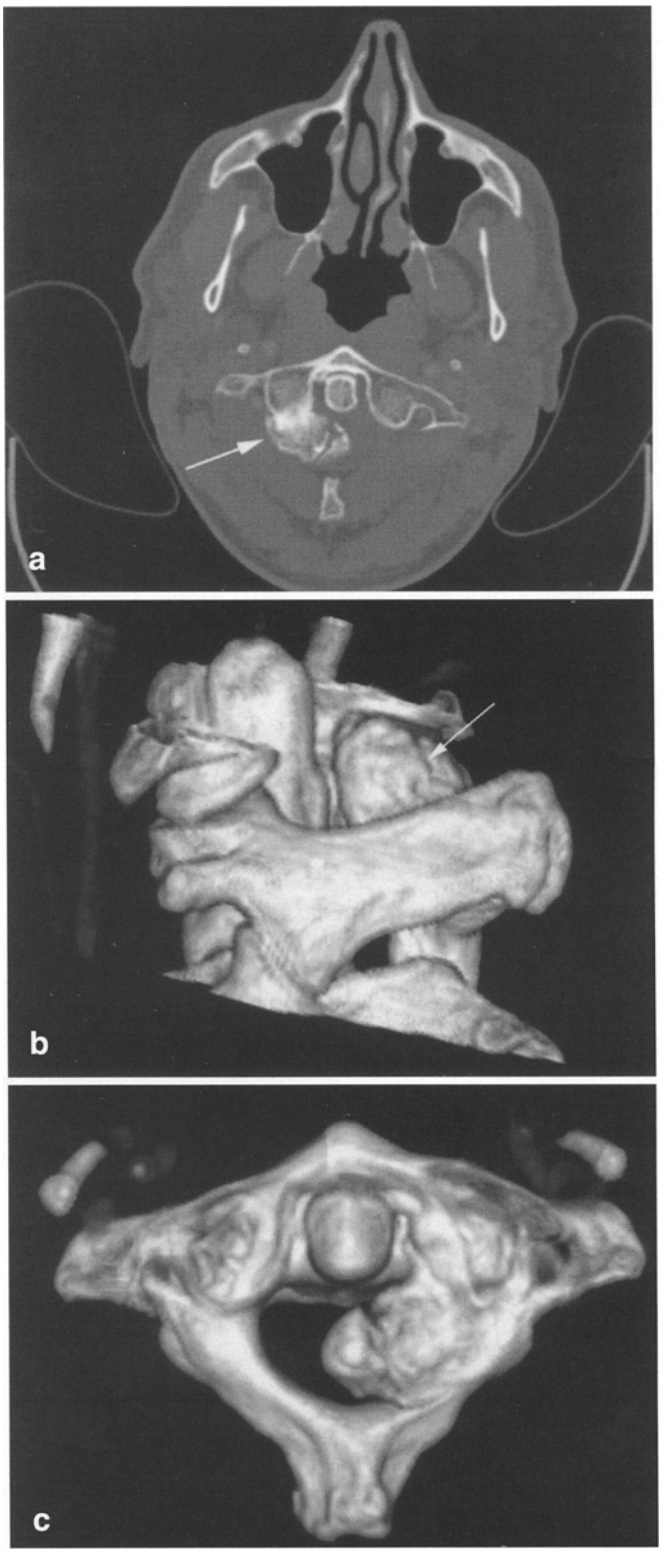

Fig. 2 **a** Axial computed tomography showing an intraspinal bone-like mass (*white arrow*) arising from the enlarged right lamina of C2; **b** lateral 3D reconstruction of CT scan showing the tumor (*white arrow*); **c** axial 3D reconstruction of CT scan showing the tumor

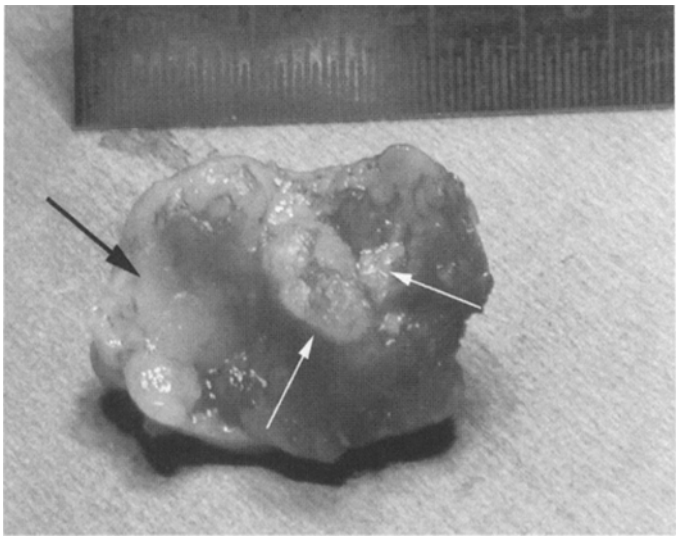

Fig. 3 Photograph of the en bloc resected tumor. Cartilage cap (*black arrow*), bony stalk (*white arrows*)

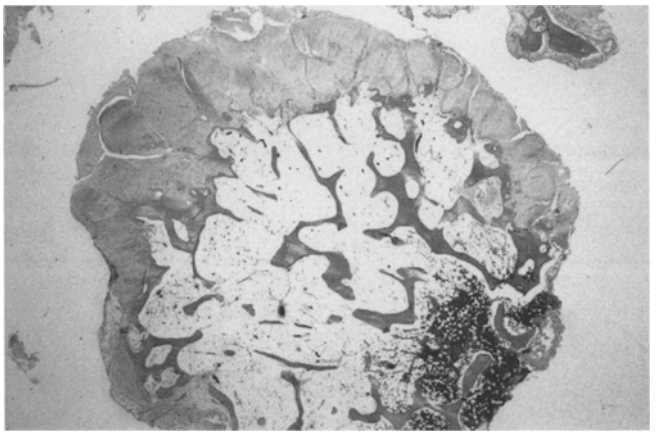

Fig. 4 Micrograph of histologic section of the tumor demonstrating a cartilage cap (*pink*) and enchondral ossification in the central part (*purple*). The base of the tumor contains bone marrow (*red*). This picture is suggestive of an osteochondroma

less common in patients with solitary spinal osteochondromas [6]. However, the slow growth of the osteochondroma may result in gradual development of these neurological symptoms, and sometimes acute symptoms may develop, e.g., after sudden hyperextension of the spine or after a fall [1]. In addition, vascular-related symptoms (compression of vertebral, carotid or subclavian arteries) could occur [1]. In all reports there is a long delay between the onset of symptoms and the final diagnosis, as in the current case.

Spinal osteochondromas are difficult to detect on X-rays because of the complex image formed by the spine. Computed tomography is the most useful technique in evaluating osteochondromas. It determines the cartilaginous and osseous components and delineates the tumor's extent and its relationship to the vertebral and neural elements of the spine. The following CT-scan findings may be considered as typical of spinal osteochondromas:

1. Roundish, sharply outlined mass
2. Bone-like density with scattered calcifications
3. Paraspinal, dumbbell or eccentric intraspinal location
4. Osteosclerotic changes in neighboring bone
5. Lack of contrast enhancement [1] (see Fig. 2a)

The relative insensitivity of MRI to small areas of ossification, compared to CT, forms a potential pitfall in the diagnosis of these lesions [6]. However, the thickness of the cartilage cap and the relationship of the lesion to other structures (e.g., an extradural intracanal component and its relation to the dural sac, spinal cord or nerve roots) are best defined with an MRI scan [1]. Bone scintigraphy can be used to stage the lesion and to exclude metastases.

Malignant transformation into a chondrosarcoma occurs in 1–2 % of patients with solitary osteochondromas [8]; therefore, treatment of asymptomatic osteochondromas is not necessary. When the tumor causes pain or neurological complications due to compression, it should be excised at its base. However, the bony consistency of spinal osteochondromas often makes it necessary to use chisels near delicate neural structures, which makes it difficult to resect the lesion en bloc. If possible, the entire cartilage cap has to be resected, as incomplete removal may lead to tumor recurrence [1]. The overall recurrence rate of these lesions is less than 2% [5].

In conclusion, aspecific symptoms caused by a spinal osteochondroma may lead to a long delay between the onset of symptoms and the final diagnosis. Especially in complex radiographic areas like the spine, lesions may be concealed from view and therefore not detected. Therefore, in the case of persistent, aspecific symptoms, further investigation by means of computed tomography is indicated to exclude spinal pathology.

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