CASE REPORT

Cervical myelopathy caused by an exostosis of the posterior arch of C1

Y. S. Chooi, Y. S. Siow, C. S. Chong

From the University Malaya Medical Centre, Kuala Lumpur, Malaysia

We report a case of vertebral osteochondroma of C1 causing cord compression and myelopathy in a patient with hereditary multiple exostosis. We highlight the importance of early diagnosis and the appropriate surgery in order to obtain a satisfactory outcome.

Solitary osteochondromas are commonly found in the appendicular skeleton. They occur less frequently in the axial skeleton, and compression of neural elements is rare.

Hereditary multiple exostosis (HME), also called diaphyseal aclasis, has a higher incidence of spinal involvement and neurological complication, compared with the solitary variety. Most vertebral exostoses grow from the external parts of the lamina but none have been reported arising from the posterior arch of the atlas (C1) of a patient with HME, causing cord compression.

Case report

A 23-year-old woman with HME, presented with a six-month history of progressive numbness and weakness of her right arm and both legs. Writing became difficult, worsened by a weakened grip. She walked unsteadily and with difficulty. Neck pain, dull in nature, radiated to her right shoulder. She described an electrical shock-like sensation radiating down her right arm when she turned her head to the right. She also had urinary and bowel incontinence, with a sensation of incomplete emptying. There was a family history of HME, her father and four of her six siblings being affected.

She was disproportionately short, with an ataxic, broad-based gait. There was neither tenderness nor palpable swelling in her neck but there were multiple asymptomatic exostoses in the extremities. Weakness and spasticity were noted (power 4 on MRC grading), with diminished sensation to light touch and pinprick in the C3-S1 dermatomes bilaterally. The sensation to vibration and proprioception was absent in both legs. There was generalised hyper-reflexia. Clonus was elicited at both ankles and knees with bilateral extensor planter responses. Hoffman's test, Lhermitte's sign, the scapulohumeral reflex, the inverted radial flex and the finger escape sign were also positive. She also failed the 10-second finger grip and release test. Evaluation of severity of myelopathy, using the Japanese Orthopaedic Association (JOA) scale, gave a score of 2 of 16.

Radiographs showed a faint area of increased sclerosis in the posterior arch of the atlas (Fig. 1). CT scan revealed a pedunculated bony outgrowth from the right anterior aspect of the posterior arch of C1, projecting into the spinal canal (Fig. 2). MRI confirmed the presence of an exostosis arising from the right side of the posterior arch of C1 causing compression of the cord (Fig. 3).

A posterior decompression with left hemilaminectomy of C1 was undertaken. The posterior arch of C1 was removed slowly using a...
high-speed burr. The exostosis, tethered to the underlying dura was removed piecemeal, allowing the indented dura to expand. Histological examination confirmed the benign nature of the lesion (Fig. 4). She was allowed to return home three weeks after surgery, with some neurological improvement. Her gait became steadier and she no longer required a walking aid. Fine and gross motor function of her hands had improved.

There was significant neurological recovery at seven months with normal tone and power of the extremities. Sensation to light touch and proprioception were normal except for in her right arm and left leg. The hyper-reflexia and ankle clonus persisted to some extent but her gait and handwriting improved. Her JOA scores improved to 9 of 16.

**Discussion**

HME, diaphyseal aclasis, multiple osteochondromatosis, Ehrenfried disease, hereditary deforming chondrodysplasia, and osteogenic disease are synonyms of the same condition which was first described by Boyer in 1814.2–6 Its pathophysiology remains unknown. It has an estimated prevalence of between 0.9 and 2 per 100 000 in Caucasian populations.7 Realistically, the true prevalence remains unknown, since asymptomatic lesions evade diagnosis. There was a delay in the diagnosis of our case and delays of up to ten years have previously been reported.8 It is an autosomal dominant disorder, with full penetrance and has an equal gender dominance.9

These benign tumours frequently arise from the metaphyseal regions of long bones, usually of the legs and occasionally from the ribs. The actively growing cartilaginous cap increases in size during normal bone growth, in both childhood and adolescence.10
Spinal involvement of solitary osteochondromas is between 1% and 4%, but in HME, it is between 7% and 9%. In the vertebral column, exostoses usually arise from the posterior elements (the secondary centres of ossification), most commonly near the tips of the spinous processes. Neurological damage is usually caused by the progressive encroachment by the slowly expanding lesion. Cord compression is uncommon; myelopathy is more common with HME than with solitary osteochondromas.

On plain radiographs, an osteochondroma in a long bone typically appears as a pedunculated or sessile bone-like projection. Spinal osteochondromas are more difficult to detect on plain radiographs because of the complex image formed by the spine. CT scan is the diagnostic imaging modality of choice. It shows the extent of the cartilaginous and osseous components, and its relationship to the vertebral and neural elements of the spine. MRI is more useful than CT in defining the extradural component and cord compression. A sudden increase in size or the onset of pain suggest malignant transformation. The incidence of malignant transformation into chondrosarcoma is reported to be between 5% and 11%.

The management of this case was aimed at relieving neurological compromise. Improvement and satisfactory recovery of function after decompression and resection of the lesion are to be expected in most cases. Our patient’s favourable prognostic factors included her youth, the duration of her symptoms for less than 12 months and the adequate space within the spinal canal at the level of the lesion. This was reflected in the significantly improved JOA functional scores seven months after surgery.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References