

Original Article

Laminectomy with posterior spinal fusion for multilevel cervical spondylotic myelopathy in an adult patient with osteopetrosis: a case report and literature review

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Abstract: Background: Osteopetrosis is a sclerosing bone disorder caused by defective bone resorption due to dysfunctional osteoclasts. A variety of orthopedic presentations have been described in autosomal dominant osteopetrosis patients. However, cases of multilevel cervical spondylotic myelopathy in osteopetrosis patients have been seldom discussed in the literature. The aim of this study was to present a case of cervical spondylotic myelopathy in association with osteopetrosis and to discuss its management. Case presentation: A 43-year-old man presented to our hospital with a 7-month history of progressive stiffness and numbness in the upper and lower extremities. Imaging findings, intraoperative pitfalls, and 4-year follow-up data are reported. The patient successfully underwent a posterior laminectomy and spinal fusion with instrumentation from C3 to C6. He had an uneventful postoperative recovery. At the 4-year follow-up, the Japanese Orthopaedic Association (JOA) score had increased from 10 to 14, and the patient had stable fixation with no evidence of implant migration or disruption. Conclusions: The occurrence of multilevel cervical spondylotic myelopathy in patients with osteopetrosis is rare. When surgery is required, laminectomy with posterior spinal fusion is indicated and reasonable. Intraoperative difficulties due to sclerosing and brittle bone should be anticipated.

Keywords: Osteopetrosis, cervical spondylotic myelopathy, laminectomy

Introduction

Osteopetrosis is characterized by osteoclast failure that results in intense sclerosis of the bone [1]. It is a rare genetic disorder that was first described by Albers-Schönberg. Recently, several genetic defects, including TCIRG1, CLCN7, OSTM1, RANKL, and TNFSF11, have been identified in the disease [2, 3]. Based upon its inheritance and clinical manifestation, three clinical types of osteopetrosis can be categorized: an autosomal recessive type that is severe and fatal within the early years of life; an intermediate autosomal recessive form that appears during childhood with a mild pathological condition; and an autosomal dominant type, which is usually considered benign, but has a full life expectancy with a variety of orthopedic problems.

To establish the diagnosis, autosomal dominant osteopetrosis (ADO) are marked by bone

within bone and a classical “sandwich vertebra” (endplate thickening of the vertebra) appearance on the radiographs of the spine. Orthopedic presentations, including fractures, arthritis and osteomyelitis, have been widely described in ADO patients [4, 5]. Additionally, spinal involvement through vertebral endplate thickening [6], spondylolysis [7], cervical spine fractures [8], and lumbar pedicle fractures [9] has been reported in the literature. However, to our knowledge, there has been no prior report of spinal arthrodesis performed in an adult ADO patient who had cervical myelopathy. It is important to note that the extremely hard and brittle nature of osteopetrotic bone imposes technical difficulties on the decompression and instrumentation of the spine [10]. Here, a case of multilevel cervical spondylotic myelopathy (CSM) in association with osteopetrosis is presented along with a discussion of its management.

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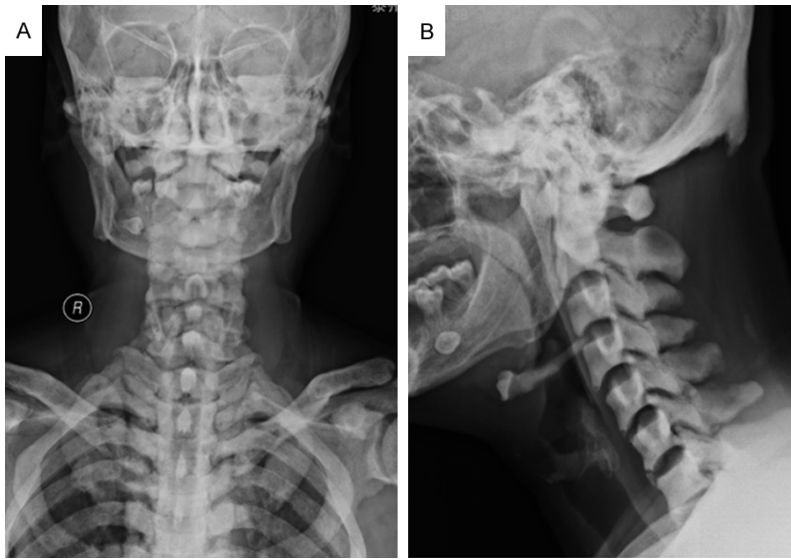


Figure 1. Preoperative anteriorposterior and lateral views showing general sclerosis of vertebrae and the base of the skull consistent with the diagnosis of osteopetrosis.

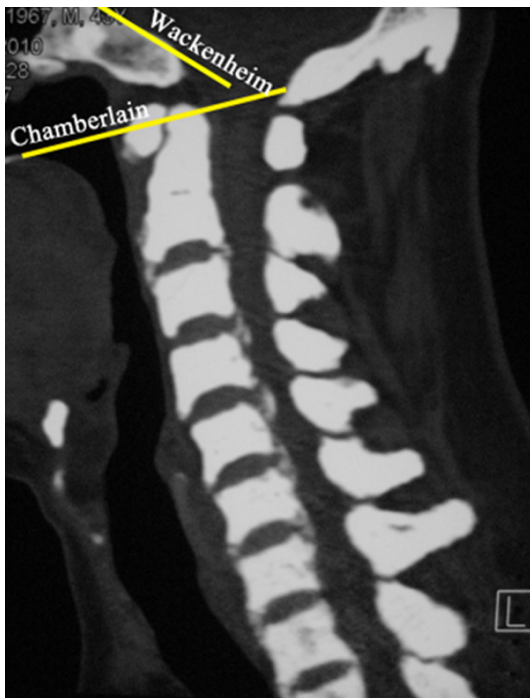


Figure 2. Sagittal CT scan image showing cervical canal stenosis with development of ossification of the posterior longitudinal ligament. Note the generalized increased density of the vertebrae.

Case presentation

Ethical approval for the study was obtained from the Institutional Review Board (NO: 2016-

146). Consent for publication was obtained from the patient. A 43-year-old man presented to our hospital with a 7-month history of progressive stiffness and numbness in the upper and lower extremities. He did not have hearing loss or vision problems, but he reported increased difficulty walking. Physical examination showed diminished sensation of the forearms and hands, objective motor weakness (grade 4/5 power) in the muscle groups of the upper and lower extremities, exaggerated deep tendon reflexes in the left lower extremities and the presence of pathological reflexes (Hoffmann's

sign). The serum level of alkaline phosphatase was 345 U/L (normal range, 45 to 125), the phosphorus level was 0.43 mmol/L (normal range, 0.87 to 1.45), and the calcium level was within normal range. Plain radiograph showed thickening and sclerosis of the vertebrae and at the base of the skull (**Figure 1**). Computed tomography (CT) scans revealed cervical canal stenosis with generalized increased density of the bones (**Figure 2**). Axial CT imaging demonstrated flattening of the spinal cord due to ossification of the posterior longitudinal ligament (OPLL) and thickening of the lamina (**Figure 3**). The occipitocervical junction was evaluated by radiographic lines and showed no abnormal finds. Sagittal T2-weighted MRI showed obliteration of cerebrospinal fluid and cord compression from C3 to C6 (**Figure 4**). The patient also had a positive maternal family history of established osteopetrosis.

After discussion with the patient and his family, surgical treatment was deemed appropriate. Because of the brittle nature of the cervical lamina and the lack of a medullary canal, the practice of laminoplasty was abandoned. Operative management consisted of a laminectomy from C3 to C6 and spinal fusion with instrumentation (VERTEX System, Medtronic Sofamor Danek, Memphis, TN, USA). Since bones in osteopetrotic patients are so sclerotic and hard, a power drill (System 6, Stryker



Figure 3. Axial CT image showing spinal stenosis due to ossification of the posterior longitudinal ligament and thickening of the lamina.

Corporation, USA) was used to create lateral mass pilot holes. Although the drill (Diameter 2 mm) was cooled with water frequently, it broke at the left mass of C4. Because removal of the broken drill bit would increase surgical time and the complexity of the surgery, attempts to remove the broken bit were abandoned and another hole was successfully created for the screw. After thinning the lamina using a high-speed burr, a 1-mm Kerrison laminectomy rongeur was used to remove the lamina. The facet joints at the involved levels were fused with the bone grafts taken from dissected lamina and cancellous allograft bone. It took 3 hours to complete the operation, and estimated blood loss for the procedure was 300 cc.

Postoperatively, the patient was asked to wear a Philadelphia collar for 3 months. He had an uneventful recovery. At the final follow-up, which was 4 years after surgery, the Japanese Orthopaedic Association (JOA) score had increased from 10 to 14. Radiographic images revealed no evidence of implant migration or disruption (**Figure 5**).

Discussion and conclusions

Osteopetrosis is a sclerosing bone disorder caused by defective bone resorption due to dysfunctional osteoclasts. The prevalence of osteopetrosis has been reported at 5.5/



Figure 4. Sagittal T2-weighted MRI showing obliteration of cerebrospinal fluid and cord compression from C3 to C6.

100,000 people [11]. The lack of bone resorption leads to diffuse osteosclerosis and bone fragility. Some distinct clinical involvement of the spine, such as scoliosis [12], spondylolysis [7], subacute odontoid fracture [8] and Chiari I malformation [13], has been described in the literature. This report presents the rare occurrence of CSM in a patient with osteopetrosis. Although the coexistence of CSM and osteopetrosis might be purely coincidental, it should be considered that there may be a causal relationship between CSM and osteopetrosis. CSM is the clinical manifestation of compression of the spinal cord caused by degenerative changes of the spine, including cervical disc herniation, developmental spinal canal stenosis, and OPLL. Based on the clinical features of the disease, two contributing mechanisms for cervical myelopathy in osteopetrosis have been proposed by McCleary [14]. One mechanism is the lack of remodeling from comprised osteoclast activity that result in thickening of the neural arch structures, which leads to spinal cord encroachment. This mechanism is further enhanced by entrapment of the cranial nerve

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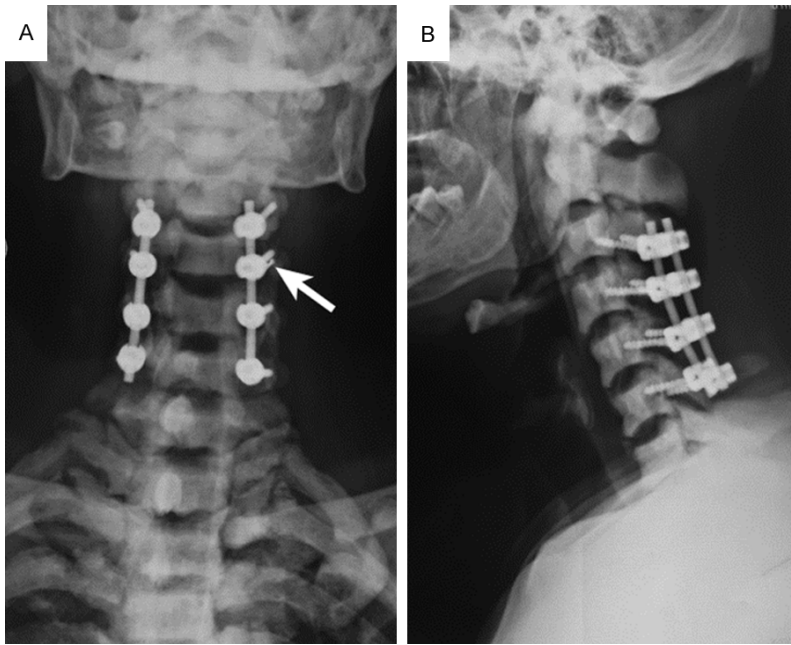


Figure 5. At 4 years final follow-up, anterior-posterior and lateral views showing a stable fixation with no evidence of implant migration or disruption. Note the broken drill bit at the C4 mass (arrow).

[15] and development of Chiari I malformation [13] in osteopetrosis patients. The second mechanism is that spinal canal stenosis may be secondary to the shunting procedure, which decreases the expansile force delivered by cerebrospinal fluid pulsations. In our patient, the laminae of the cervical spine were thickened to double their normal size, but the patient did not present with ventriculomegaly or have a shunting procedure. The latter mechanism may not be involved in the development of spinal stenosis in our presented case. However, OPLL were found in the posterior aspect of the intervertebral disc (**Figure 3**). Therefore, in our case, a combination of thickening laminae and OPLL contributed to the formation of spinal stenosis and finally led to the development of CSM.

The unique nature of osteopetrotic bone imposes technical challenges during surgery [10, 16]. A few reports in the literature have described surgical treatment methods in this population. Martin [7] reported that one of 5 patients with spondylolysis at the cervical spine underwent posterior arthrodesis, and at forty-five months follow-up, a pseudarthrosis was found at the fixation level. In contrast, Auerbach described a successful spinal arthrodesis in a patient with osteopetrosis and subacute odontoid fracture

using transarticular screws and interspinous wiring. In their patient, a solid posterior cervical arthrodesis was achieved at 2 years post-operation [8]. Additionally, Westerlund [12] reported on the successful operative management of a patient with scoliosis using Luque rods, sublaminar wires and allograft bone augmentation. McCleary [14] presented the congenital osteopetrosis of a 15-year-old boy with upper cervical myelopathy. The boy also presented with ventriculomegaly and had three shunting procedures. Finally, two levels of laminectomy without fixation were performed to successfully relieve the spinal stenosis. However, the boy did not have full neuro-

logical recovery after surgery. In our patient, while planning for surgery, laminoplasty was not chosen. The osteopetrotic bone was found to be extremely brittle and easily fractured. In addition, it was shown that the laminae of the cervical spine were hyperostotic and without medullary canal. These unique natures of the bone made it impossible to perform laminoplasty for the patient. Therefore, a laminectomy with posterior instrumented fusion was performed. While drilling holes for screw insertion at the left mass of C4, the drill bit broke, which increased the surgical complexity. After failed attempts to remove the broken bit, the screw were inserted at the lateral mass. Technical difficulties have been described during surgery in patients with osteopetrosis. Bhargava [10] present the common surgical problems and general management principles for the treatment of osteopetrosis patients. Based on a literature review and our experience, it is important to note that preoperative planning is essential to manage the possible complications.

Delay, or non-union, is another common complication in osteopetrosis because of the diminished blood supply [7]. Allogenic cancellous bone combined with prolonged postoperative

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immobilization has proven effective for achieving a solid fusion in osteopetrotic patients [8, 12]. The present case was also found to have stable fixation with no implant migration or disruption at the 4-year follow-up.

In conclusion, the occurrence of multilevel CSM in patients with osteopetrosis is rare. When surgery is required, laminectomy with posterior spinal fusion is indicated and reasonable. Intraoperative difficulties due to sclerosing and brittle bone should be anticipated.

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Disclosure of conflict of interest

None.

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