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GRAND ROUNDS

Surgical challenges in the management of cervical kyphotic deformity in patients with severe osteoporosis: an illustrative case of a patient with Hajdu–Cheney syndrome

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Abstract

Purpose No standard strategy exists for the management of cervical kyphotic deformity in patients with severe osteoporosis. In fact, in such subpopulation, standard algorithms commonly used in patients with normal bone mineral density may not be applicable. In this Grand Rounds, the authors present a challenging case of a patient with Hajdu–Cheney syndrome, a rare disorder of bone metabolism induced by a Notch-2 mutation, who presented with cervical kyphotic deformity and severe osteoporosis.

Methods A 65-year-old female patient with a previous diagnosis of Hajdu–Cheney syndrome presented with cervical myelopathy and cervical kyphotic deformity. The initial MRi demonstrated multilevel cervical canal stenosis. The CT-scan also revealed marked spondylolisthesis of C6 over C7 as well as numerous laminar and pedicle fractures, resulting in a cervical kyphosis of approximately 50 degrees.

Results The patient was submitted to 360-degree decompression and fusion of the cervical spine consisting of a staged C6 anterior corpectomy and multilevel microdiscectomies with wide opening of the posterior longitudinal ligament in order to provide a satisfactory release of anterior spinal structures, followed by 24 h of cervical halo-traction, a second anterior approach for bone graft implantation in the site of the corpectomy as well as insertion of allografts and completion of the ACDF C2–T1 and plating, and, finally, a posterior C2–T3 pedicle screw instrumentation using intra-operative CT-scan (O-arm) navigation guidance.

Conclusions This case illustrates some intra-operative nuances as well as specific surgical recommendations for cervical deformity surgery in patients with severe osteoporosis, such as avoidance of Caspar pins for interbody distraction, use of intra-operative fluoroscopy for achievement of bicortical purchase of anterior cervical screws and placement of pedicle screws during posterior instrumentation. Moreover, such illustrative case demonstrates that, in the subpopulation of patients with severe osteoporosis, it may be possible to successfully apply cervical distraction after an isolated anterior approach with a satisfactory improvement in the cervical alignment, possibly avoiding more laborious 540-degree approaches such as the previously described back–front–back or front–back–front surgical algorithms.

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Case presentation and diagnostic imaging

A 65-year-old female patient with a previous diagnosis of Hajdu–Cheney syndrome (Fig. 1) presented with complaints of tingling and numbness in the upper and lower extremities that started 3 days before following a sudden movement of her neck. The patient also reported worsening gait in the last months with progressive weakness in her lower limbs. The neurological exam revealed clear signs of cervical spondylotic myelopathy with presence of hyperreflexia in upper and lower limbs with sustained clonus, Babinski and Hoffman's signs bilaterally. The strength was preserved in her upper limbs, but was slightly reduced in the lower limbs (Grade 4 +/5).

The initial MRi of the cervical spine demonstrated multilevel cervical canal stenosis with severe spinal cord compression at the level of C6, with the antero-posterior diameter of the spinal canal measuring approximately 4 mm at this level (Fig. 2). The CT-scan revealed a marked spondylolisthesis of C6 over C7 as well as numerous laminar and pedicle fractures, resulting in a cervical kyphotic deformity of approximately 50 degrees (Fig. 2).

Fig. 1 Photographs of the superior and inferior extremities of the patient with Hajdu–Cheney syndrome presented in this article demonstrating the extensive bone and connective tissue deformation induced by the Notch-2 gene mutation

Historical review: the Hajdu–Cheney syndrome

Hajdu and Kauntze first described a very rare connective tissue disorder, which they called 'cranioskeletal dysplasia', in 1948 [1], with Cheney providing a further analysis of the clinical course of such disease in 1965 [2]. This syndrome, nowadays better known as Hajdu–Cheney syndrome (but which has also been referred as 'arthro-dento-osteodysplasia' or 'cranio-skeletal dysplasia with acro-osteolysis') is a rare bone and connective tissue metabolism disorder which exhibits autosomal dominant inheritance. The clinical presentation of this disease includes skull malformations, hyperextensive joints, short stature, periodontal disease, coarse hair, ptosis, delayed closure of cranial sutures, absent frontal sinuses and severe osteoporosis [3, 4]. Notable spinal complications associated with Hajdu–Cheney syndrome include atraumatic fractures, multilevel vertebral fractures, syringomyelia, basilar invagination, cervical kyphotic deformity and cervical instability.

Recent studies have identified mutations in the exon 34 of the C-terminus portion of the Notch-2 gene as the underlying genetic defect of the Hajdu–Cheney syndrome [5, 6]. However, the exact mechanisms through which such mutation leads to the overexpression of the Notch-2 protein and subsequent osteopenia are not clearly defined. It has been suggested that overexpression of Notch-2 may lead to both increased osteoclast-mediated bone resorption [7] as well as abnormal osteoblast or osteoid function [8]. An alternative theory suggests that elevated levels of the Notch-2 protein may lead to the accumulation of mast cells and osteolytic cytokines, ultimately causing bone degradation [9].

Rationale for treatment and evidence-based literature

Kyphotic deformities account for the majority of subaxial cervical spine imbalances [10]. Often, cervical kyphosis develops secondarily or in association with other clinical conditions including multilevel degenerative disc disease, trauma, ankylosing spondylitis, or neoplastic disease [10]. Additionally, an iatrogenic etiology, related to the disruption of the posterior spinal elements after non-instrumented laminectomies, may also account for a significant percentage of cases of cervical kyphotic deformities [11].

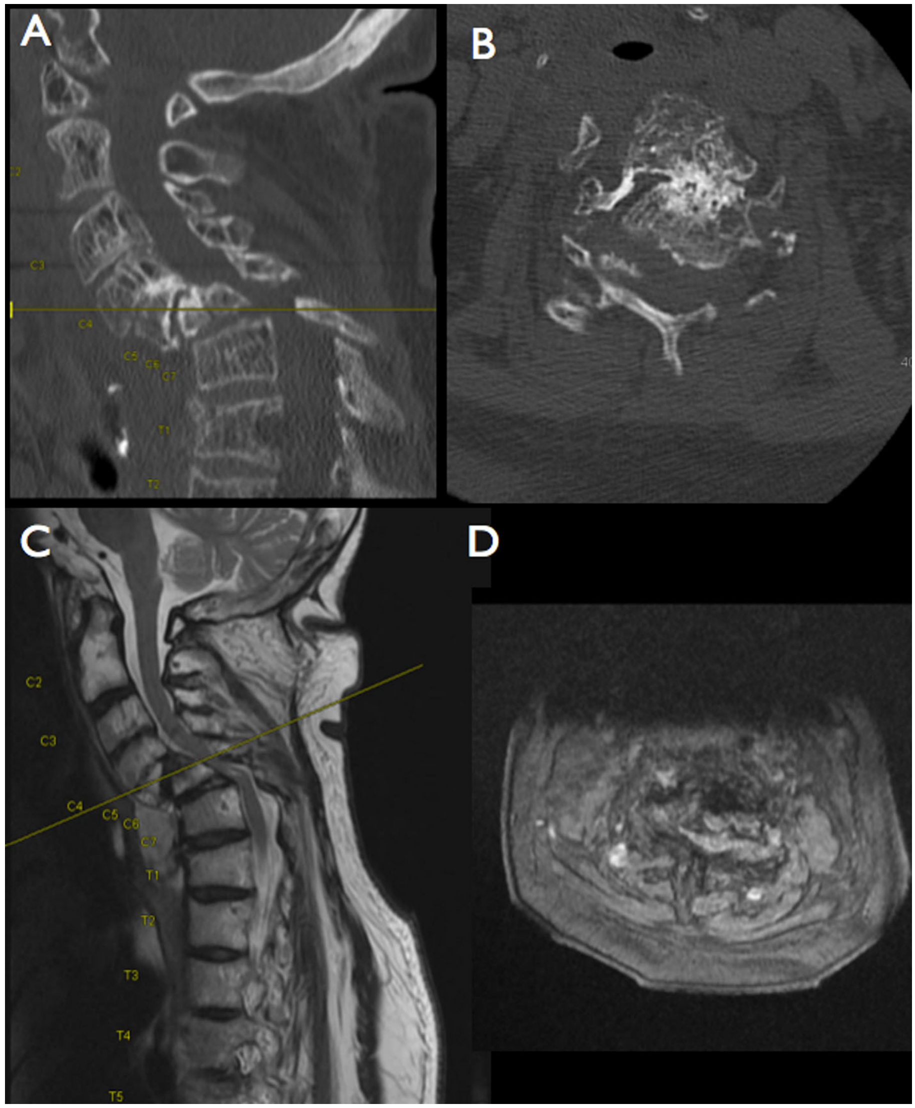

Fig. 2 Midline sagittal view (*left*) and axial slice at the C5–C6 level (*right*) of CT-scan (*top*) and MRi (*bottom*) demonstrating multilevel cervical canal stenosis (more prominent at the C5–C6 level) as well as severe cervical kyphosis

In the initial course of the disease, patients exhibit loss of cervical lordosis or flat neck, but may otherwise be asymptomatic or present only minimal pain and mild impairment in their quality of life [10]. However, with the progression of the kyphotic deformity, pain, neurological deficits (including radiculopathies and myelopathy), as well as more prominent limitations to daily activities due to the sagittal imbalance become apparent [12, 13]. In the worst scenario, patients may progress to the debilitating condition of chin-on-chest deformity.

The necessity of surgical intervention in patients with cervical kyphosis is often determined with basis on the degree of deformity (as it has already been demonstrated that the severity of kyphosis has a direct influence upon the ability to maintain a normal horizontal gaze [14]), the existence of associated neurological deficits, and the presence of residual flexibility of the cervical spine (in other words, if the kyphotic deformity is fixed or flexible). According to a recent literature review [15], horizontal gaze (as quantified by the kyphosis angle) was the most important factor for indicating surgery in previous surgical series. In relation to the presence of myelopathy, experimental studies have already demonstrated that progressive kyphosis may lead to neuronal loss, white tract demyelination, and atrophy of the spinal cord, likely related to the chronic compression of the spinal cord vasculature [16].

Unfortunately, there is no unique standard strategy for the surgical management of cervical kyphotic deformities and, ultimately, the type and number of approaches should be tailored according to each patient's anatomy, bone quality, clinical and neurological status, as well as specific imaging characteristics. In such scenario, the etiology of the cervical kyphosis also constitutes an important factor which should be taken into account when planning the most appropriate surgical strategy.

In the setting of severe cervical kyphotic deformity and marked sagittal imbalance [17, 18], some authors have defended the necessity of osteotomies for proper correction of the spinal alignment. In such scenario, C7 has been considered the ideal level for such osteotomies for the following reasons [17]: the vertebral artery usually enters the foramen transversarium at C6; the region between C7 and T1 contains a larger spinal canal compared to the mid-cervical spine; any potential damage to the nerve roots still allows the patient to retain a reasonable degree of function of the upper extremities. Nevertheless, cervical osteotomies at the C6 level for the treatment of severe cervical kyphosis have also been reported [18].

Other strategies for the management of cervical kyphotic deformity involving an anterior, posterior, combined anterior–posterior or 540-degree surgical approaches have also been described [12]. Anterior approaches have the advantage of not only relieving anterior compression to the spinal cord (which is very common in cases of cervical kyphotic deformity), but also of providing the best strategy for restoration of the cervical lordosis by the insertion of an interbody graft strut or expandable cage [12]. Moreover, complication rates of anterior approaches are usually lower than those of posterior approaches. The main indication for posterior approaches is a flexible kyphosis. Isolated posterior approaches are rarely indicated, as in the setting of cervical kyphotic deformity, long posterior cervical instrumentations (specially those relying on lateral mass screws) in the absence of a supplementary anterior instrumentation seem to lead to unacceptable rates of pseudarthrosis and construct failure. In the majority of the cases, the debate is between choosing an isolated anterior approach or a combined strategy. Especially if posterior ankylosis of the vertebral joints exists (as in patients with ankylosing spondylitis), a single anterior approach is limited in its capacity of correcting the cervical kyphotic deformity.

With regard to combined strategies, it is traditionally believed that, at least theoretically, anterior grafts may loosen and migrate if further posterior corrections are attempted [13, 19]. Therefore, anterior corrections are usually the last surgical stage and, if any further posterior approach is performed, it is recommended that it should be restricted to instrumentation without further deformity correction. In patients suffering from significant comorbidities, particularly those which may lead to impairment of the bone quality, a combined approach has been found to maximize long-term clinical and radiographic outcomes [12, 20]. In such scenario, 540-degree approaches have been proposed as the option which provides the highest rates of a satisfactory and sustained correction of the cervical kyphosis. It must be remembered, however, that, although 540-degree approaches allow for a greater correction of the cervical kyphosis due to the possibility of manipulating both the anterior and posterior vertebral columns, they have also been demonstrated to be associated with longer operative times and higher complication rates [12].

The proposed front–back–front paradigm consists of initial anterior discectomies (and if necessary cervical osteotomies and unilateral pediculectomies) followed by a posterior fixation with cervical screws and rods with correction of the deformity through repositioning of the Mayfield clamp, followed by a second anterior approach for placement of structural interbody grafts (i.e., allografts, long bone struts or expandable cages, depending on the anterior column defect) and plating [12]. The disadvantage of such algorithm is that the performance of two sequential anterior approaches some days apart may increase the risk of complications related to pre-vertebral soft tissue swelling and upper airway edema. Additionally, in patients with severe cervical kyphotic deformity, flipping a patient to prone position after an isolated anterior decompression without fixation may lead to higher risks of spinal cord injury due to the significant spinal instability caused by the initial surgical procedure.

he back–front–back sequence is usually considered when the initial kyphosis is too severe to perform the anterior approach as the first stage. Additionally, in patients with severe multilevel spinal canal stenosis, it provides the easiest and fastest method for decompressing the neural elements. In such cases, the first posterior approach consists of multilevel posterior facetectomies and osteotomies to enable further correction of the deformity from the front. Some surgeons may opt to perform the lateral mass or pedicle screw instrumentation during this first initial approach. In such scenario, the rods are not inserted to enable further correction of the deformity during the anterior approach. If the second stage is planned to be carried out on a different day (due to anesthetic limitations or any other factor related to the patient's clinical condition), most surgeons would choose to place the patient in a halo-vest to provide some sort of temporary stability to the spine. The second stage consists of an anterior approach with multilevel discectomies (and/or corpectomies), with correction of the deformity, anterior column reconstruction (with either allografts, a long anterior strut graft or an expandable cage) and plating. During this step, the application of cervical traction (with a Gardner-Wells tongs, for example) is extremely useful to restore the cervical lordosis. Finally, the third stage consists of a second posterior approach for instrumentation of the spine (if it has not yet been performed during the initial posterior approach), placement of the rods and final locking of the construct, usually without any further attempt to correct the deformity in order to avoid loosening of the anterior plating and displacement of the anterior grafts.

Despite the availability of several surgical algorithms for correction of cervical kyphosis, most of them are derived from patients with ankylosing spondylitis [13, 14]. In such disease, there is an associated inflammatory process that leads to severe arthritic changes of the intervertebral joints, ultimately resulting in intense bone remodeling and a fixed kyphotic deformity [14]. In fact, although combined 540-degree strategies may be required for adequate correction of the cervical kyphosis in patients with fixed deformities and normal bone quality, they may not be necessary in patients with significant osteoporosis. In such patients, the anterior release through multilevel discectomies (and, if necessary, corpectomies) followed by a short period of halo-traction may be enough to offer a significant reduction of the kyphotic deformity, avoiding the necessity of flipping the patient without instrumentation (which may carry a non-negligible risk of spinal cord injury), as required in 540-degree algorithms. The staging between the first anterior approach for disc-ligamentous release and the second anterior approach for reconstruction of the anterior column is justified by the benefits in terms of correction of the cervical kyphotic deformity that the sustained halo-traction provides. The application of halo-traction without the anterior surgical release is not only unlikely to offer a satisfactory reduction of the cervical kyphosis, but may also lead to worsening of pre-existing neurological deficits, especially in those patients with severe spinal stenosis, as in the presented case. Similarly, intra-operative application of cervical traction for only a short period of time during the first surgical approach is also unlikely to lead to a satisfactory reduction of the kyphotic deformity. Conversely, sustained application of traction with progressive weights after the initial anterior release (for one or more days, as in the presented case) benefits from the increased viscoelasticity of the soft tissues and bony structures of patients with osteoporosis, ultimately enabling a significant reduction of the kyphotic deformity.

Finally, the surgical management of cervical kyphotic deformity in the subpopulation of patients with low bone quality present several other important differences when compared to traditional algorithms used in patients with normal bone mineral density. For example, although the presence of osteoporosis may induce most spine surgeons to opt for keeping the halo-vest postoperatively for some weeks or months (as an attempt to avoid pseudarthrosis and a possible catastrophic instrumentation failure in such challenging cases), there may be a significant morbidity associated with the prolonged use of a halo-vest in such patients, as demonstrated by the presented illustrative case. Therefore, based on our experience, we recommend that, especially in those cases of diffuse osteoporosis induced by genetic syndromes, the use of halo be limited to short periods between the staged surgical approaches.

Operative procedure

After an extensive discussion about the best surgical strategy for decompressing, reducing and stabilizing the cervical spine in the case under discussion, the authors supposed, with basis on the patient's very low bone quality as demonstrated by a previous bone mineral density scan, that it would be possible to avoid a 540-degree back–front–back approach by performing an anterior one-level (C6) corpectomy as well as multilevel microdiscectomies followed by application of 24 h halo-traction. If a satisfactory reduction of the deformity could be achieved, the surgical plan would include a second anterior approach for insertion of a bone strut at the level of the corpectomy, completion of the multilevel ACDFs with implantation of allografts and instrumentation with plate and screws followed by a final long posterior cervico-thoracic instrumentation for supplementing the anterior construct.

The patient was submitted to an anterior cervical approach for a C6 corpectomy (Fig. 3a) and C2–C3, C3–C4, C4–C5 and C7–T1 microdiscectomies with decompression of the spinal cord and wide opening of the posterior longitudinal ligament in order to provide proper release of the anterior spinal structures. Several anatomical particularities were observed during the anterior cervical approach, such as a substantial carotid dolichoectasia and significant bleeding from the porous and soft osseous structures. Special intra-operative recommendations for cervical approaches to the spine in patients with severe osteoporosis include avoiding Caspar pins for interbody distraction (as they may easily lead to vertebral body fractures) and use of intra-operative fluoroscopy for achieving bicortical purchase of the anterior cervical screws during plating.

After the initial anterior procedure, the patient was placed in halo-traction with progressive weights up to 30 pounds until the kyphotic deformity was satisfactorily reduced (Fig. 3b). At this point (24 h after the initial surgery), the patient was taken back to the OR and the ACDF at the C2–C3, C3–C4, C4–C5 and C7–T1 levels was completed with insertion of allograft bone in the disc spaces followed by reconstruction of the anterior column at the site of the corpectomy with a tricortical bone strut (Fig. 4a). Additionally, an anterior plate and screws from C2 to T1 were inserted. Finally, at the same operative setting, the patient was flipped and a C2–T3 instrumentation was performed with O-arm navigation-guided insertion of pedicle screws at the cervical spine (Fig. 4b).

Due to the low bone quality, the authors decided to keep the patient in a halo-vest. Four weeks later, after verifying the loosening of the cranial screws used for halo fixation, the authors tightened the screws. Although no problem was

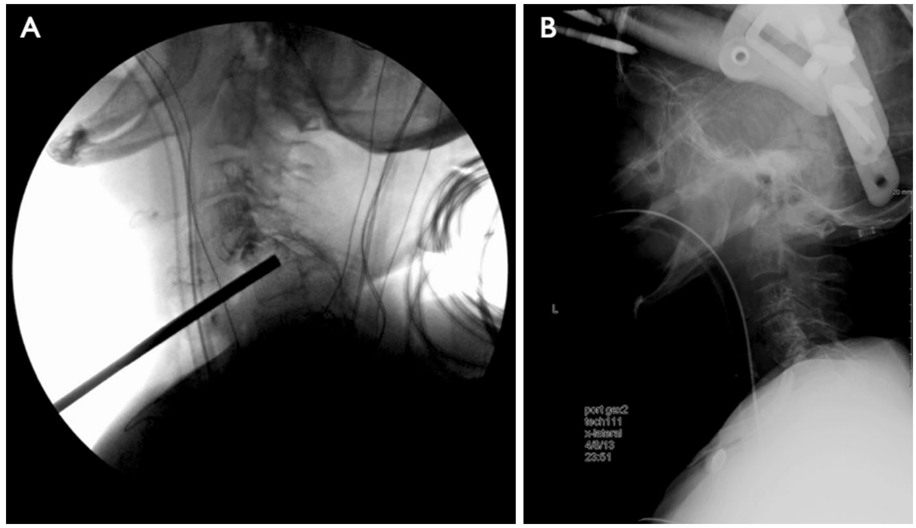

Fig. 3 **a** *Left* Intra-operative fluoroscopy after the C6 corpectomy and C2–C3, C3–C4, C4–C5 and C7–T1 microdiscectomies with wide opening of the posterior longitudinal ligament. **b** *Right* Postoperative X-ray demonstrating the remarkable reduction of the kyphotic deformity after 24 h of halo-traction

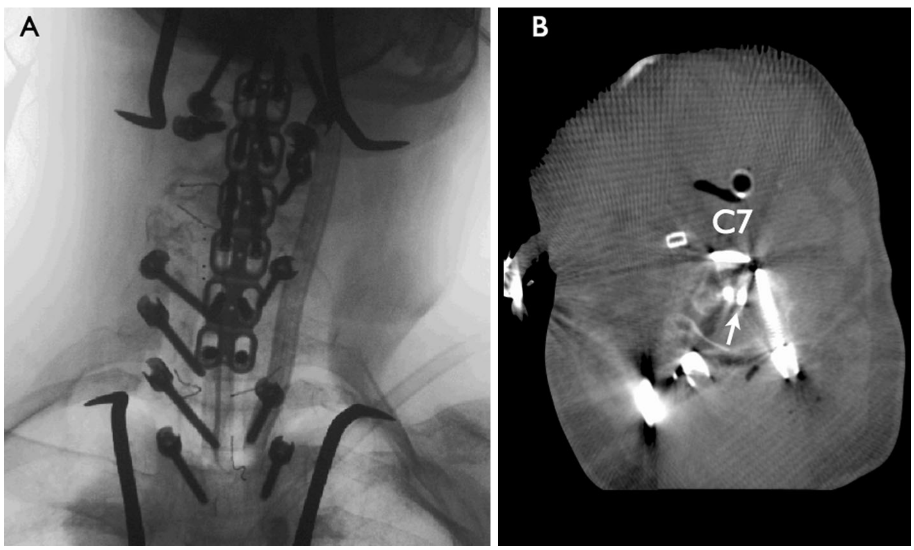

Fig. 4 **a** Intra-operative fluoroscopy after the completion of the C2–T1 ACDF with insertion of interbody grafts at the site of the C2–C3, C3–C4, C4–C5 and C7–T1 microdiscectomies and a bone strut at the site of the C6 corpectomy, followed by the supplementary posterior C2–T3 instrumentation (*left*). **b** Intra-operative CT-scan (O-arm) demonstrating the C2–T1 ACDF and plating (note the bicortical anterior screws—*arrow*) and posterior C2–T3 instrumentation with pedicle screws (note that due to the extensive bony destruction as well as severe osteoporosis it was not possible to insert pedicle screws at some levels)

observed at this time, a follow-up CT-scan demonstrated that the left parietal screw perforated the inner cortex of the skull (Fig. 5a). At this point, the head-fixating screws were removed and the halo-vest was kept in place supported by a soft cranial pressure system.

Follow-up

At the 1-year follow-up, the patient was already out the halo. Neurologically she presented a significant improvement in hand coordination and strength, and was already able to feed herself and write. At this point, she reported no neck pain. The CT-scan showed good position of her spinal instrumentation, no significant evidence of hardware loosening, and fusion in progression (Fig. 5b).

Conclusions

Standard algorithms for correction of cervical kyphotic deformity in patients with normal bone mineral density may not be applicable to patients with severe osteoporosis. In fact, in such population, due to the increased viscoelasticity of the spinal structures, 540-degree approaches may not be required. In this technical note, we present a

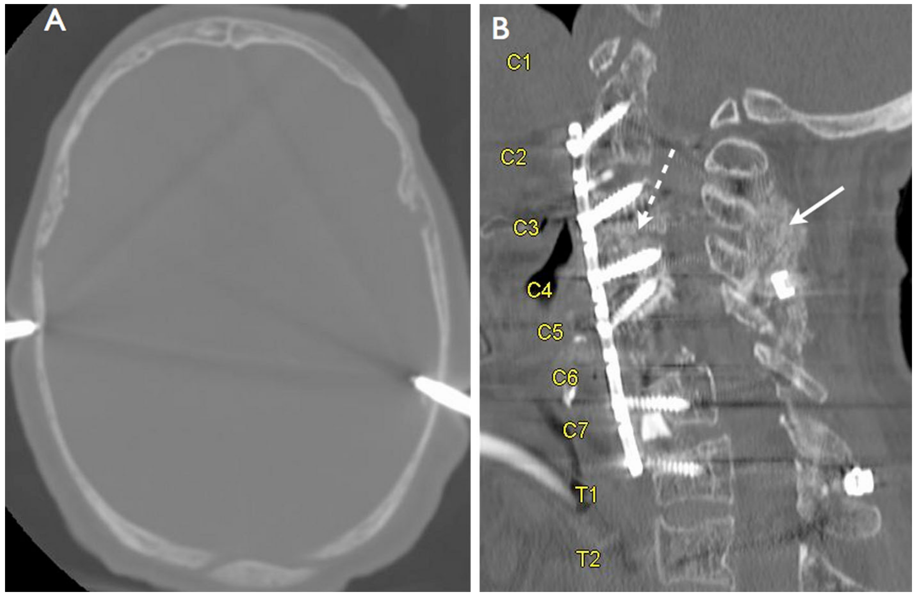

Fig. 5 **a** 2-weeks postoperative CT-scan after attempted tightening of the halo-fixating cranial screws, demonstrating perforation of the inner cortex of the skull by the left parietal screw. **b** 6-months postoperative CT-scan demonstrating fusion in progression, especially between the posterior elements (*arrow*) and anteriorly at some of the intervertebral spaces (interrupted *arrow*)

new algorithm for treatment of cervical kyphotic deformity in such patients, which consists of an anterior approach for anterior spinal cord decompression and release of the anterior bony and disc-ligamentous structures followed by a short period of sustained halo-traction with progressive weights. When a satisfactory reduction of the kyphosis is obtained, a new anterior approach for anterior column reconstruction (with either interbody grafts, bone struts or expandable cages, depending on the defect) and instrumentation with plate and screws is then performed. Finally, a long posterior cervico-thoracic (or if necessary cranio-cervico-thoracic) instrumentation to supplement the anterior construct is performed. The use of such algorithm was illustrated with the presentation of a patient with Hajdu–Cheney syndrome with severe osteoporosis and a cervical kyphotic deformity in whom such combined approach was successfully employed. Finally, specific intra-operative nuances as well as specific surgical recommendations for cervical deformity surgery in patients with severe osteoporosis (such as the avoidance of Caspar pins for interbody distraction, use of intra-operative fluoroscopy for achievement of bicortical purchase of anterior cervical screws and placement of pedicle screws during posterior approaches) were highlighted.

Conflict of interest The authors declare that there is no conflict of interest and that no funding was received for this work.

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