

Surgical Algorithm for Management of Cervical Deformity in Acro-osteolysis (Hadju-Cheney Syndrome)

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Introduction

Acro-osteolysis (Hadju-Cheney Syndrome) is a rare disorder of bone metabolism characterized by progressive destruction of the distal phalangeal bones (Figure 1), aplasia of the facial sinuses, persistence of the cranial sutures and severe spinal deformities.

Methods

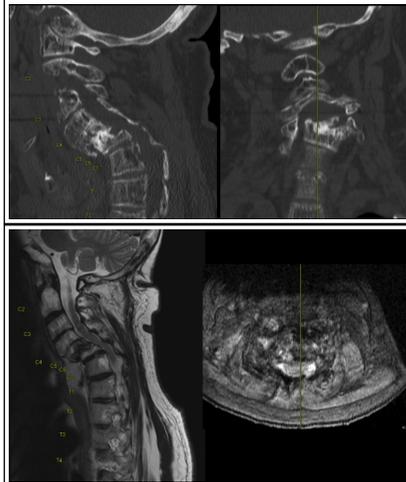
There is no standard strategy for management of cervical deformity and instability in such patients and, as such syndrome involves severe degrees of osteoporosis, standard algorithms for deformity correction (such as the standard back-front-back approach) in healthy patients may not be applicable.



Figure 1

Photos of the patient superior and inferior extremities demonstrating the soft tissue deformity caused by the NOTCH2 mutation.

Figure 2

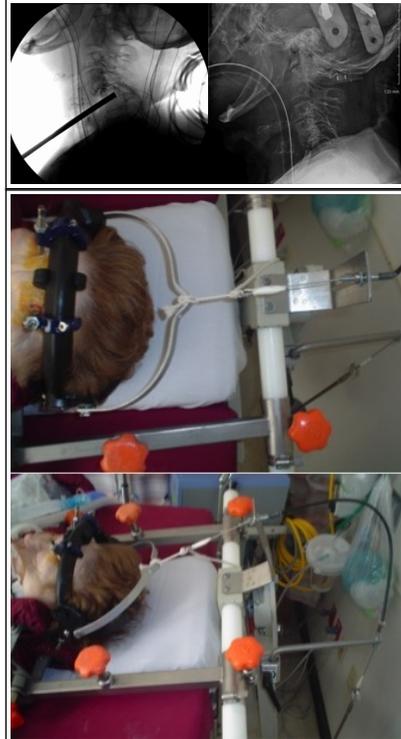


Sagittal (left) and axial (right) slices of CT-scan (top) and MRI (bottom) demonstrating the multi-level cervical canal stenosis (more prominent at the C5-C6 level - axial slice of MRI) as well as the severe severe cervical kyphoscoliosis.

Results

In this report we describe the case of a 65-year old female patient with Hadju-Cheney syndrome who presented with cervical myelopathy and severe cervical kyphoscoliosis (Figure 2). The patient was submitted to 360° cervical decompression and fusion in a staged anterior consisting in: corpectomy (Figure 3/top), followed by 24h of halo traction (Figure 3/bottom), followed by another anterior approach for bone graft implantation and ACDF C2-T2, and finally complemented by a posterior C2-T3 fusion with navigation guidance with intra-operative CT-scan (O-arm)-Figure 4.

Figure 3



Left: Intra-operative fluoroscopy after the C6 corpectomy. Right: Post-operative x-ray demonstrating the remarkable reduction of the kyphotic deformity after 24h of halo traction (bottom)

Figure 4



Intra-operative fluoroscopy after C2-T2 ACDF (left) and posterior C2-T3 fusion

Conclusions

In this report the authors highlight the particularities of the surgical management of cervical deformity in patients with Hadju-Cheney Syndrome. According to our experience several nuances should be expected in such patients, such as: substantial carotid dolichoectasia and increased bleeding from osseous structures. In our experience we recommend the following surgical strategies: no use of Casper pins for interbody distraction, use of intra-operative fluoroscopy for achievement of bicortical purchase of anterior cervical screws, placement of pedicle screws (instead of lateral mass screws) during posterior approaches and maintenance of halo-vest in the post-operative period for at least 3 months. It was also verified that it is possible to successfully apply cervical distraction after an isolated anterior approach with corpectomy with a satisfactory improvement in cervical lordosis without a posterior approach for bilateral facetectomies. The proposed therapeutic algorithm and surgical nuances may have broader applications for management of cervical deformity in other non-syndromic patients presenting with severe reduction in bone density.

Learning Objectives

- To be able to describe the main clinical and genetic features of Hadju - Cheney Syndrome (Acro-osteolysis)
- To be able to describe the possible challenges for management of cervical deformity in patients with severe osteoporosis
- To understand the possible salvage strategies as well as the intra-operative nuances during surgical treatment of cervical deformities in patients with reduced bone density such as in the presented case

References

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