HIRAYAMA DISEASE: ABOUT TWO CASES

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INTRODUCTION

Hirayama's disease or distal juvenile muscular atrophy is a rare myelopathy associated with cervical flexion. It mainly affects adolescent boys.

It manifests with progressive muscle weakness and distal atrophy of the upper limbs, generally unilateral.

The diagnosis is confirmed with functional cervical MRI, where anterior displacement of the dural sac is observed during neck flexion, causing compression of the cervical spinal cord, producing atrophic changes and ischemic disease of the anterior horn.

The indicated treatments consist of reducing cervical mobility: cervical orthoses, spinal fusion or duraplasty associated with fusion

OBJECTIVE

Report two clinical cases of Hirayama Disease, a rare pediatric myelopathy associated with a cervical flexion.

MATERIAL AND METHOD

Retrospective and descriptive study of the clinical history of two patients with Hirayama Disease

RESULTS

Patient 1

17 years old boy, presents with progressive weakness of upper right limb with a 14-month progression. Physical examination showed weakness at flexo-extension of forearm and hand muscles associated with hypothrophy.

Patient 2

16 years old boy, presents with involuntary movements of both hands on neck flexion with associated paresthesia. Physical examination showed hypothrophy of left biceps and wasting of bilateral hypothenar eminence

In both cases, diagnosis was confirmed with functional MRI, which showed forward displacement of the spinal cord and increased posterior epidural space on neck flexion (Figure 1 and 2).

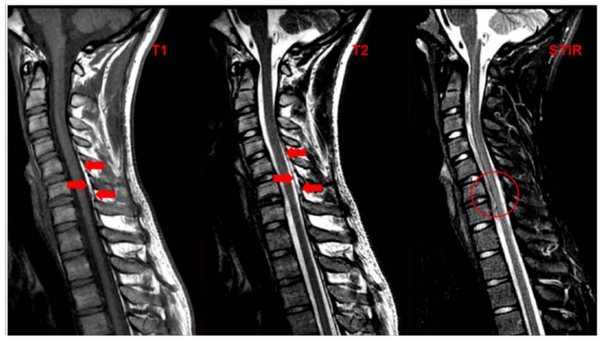

Figure 1- Sagittal section in T1-T2-STIR sequences in neutral position. A hypointense line indicated by the arrow is observed forward, which is the dura mater that became detached from the vertebral bodies, and backwards the posterior limit of the epidural space. The hyperintensity between the arrows is the epidural space with the venous plexuses and the epidural fat that are pathological.
In STIR, a decrease in the anteroposterior diameter of the spinal cord is observed.

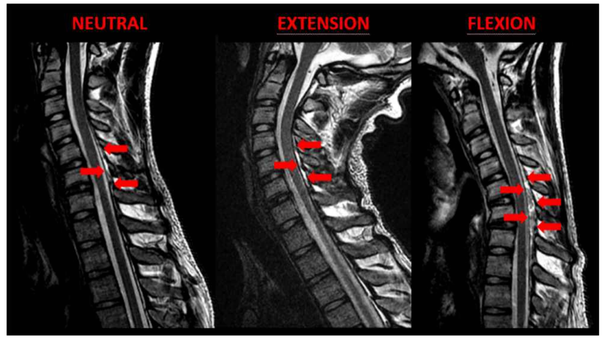

Figure 2- The 3 positions of the functional MRI in a T2 sagittal cut. It is observed that in the neutral position and in extension the alteration that is revealed is slight, while in flexion the anterior migration of the dura mater and the increase in the posterior epidural space can be clearly observed.

CONCLUSION

A thorough understanding and recognition of this pathology is essential in order to request a cervical MRI with flexo-extension, because if it is not functional, no alteration can be observed.
Reaching an accurate diagnosis is necessary to indicate treatment and to avoid clinical progression.

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