Juvenile Amyotrophy of the Distal Upper Extremity (Hirayama Disease), a Disease of Transition to Adulthood: Case Series and Surgical Management

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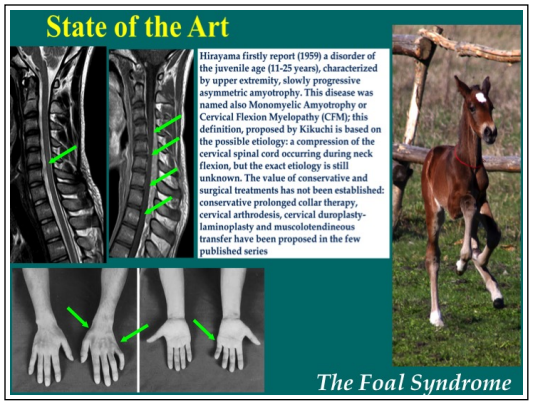


Introduction

Hirayama firstly reported (1959) a disorder of the juvenile age (11-25 years), characterized by unilateral or asymmetric slowly progressive amyotrophy of the distal upper extremity. The initial progressive course is followed by a spontaneous arrest. Named Hirayama Disease (HD) or Monomyelic Amyotrophy or Cervical Flexion Myelopathy (CFM), proposed by Kikuchi basing to the possible etiology: a compression of the cervical spinal cord during neck flexion. Etiology is still debated. The value of conservative or surgical treatments has not been established: collar therapy, cervical arthrodesis, cervical duroplasty – laminoplasty and muscolotendineous transfer have been proposed.

Methods

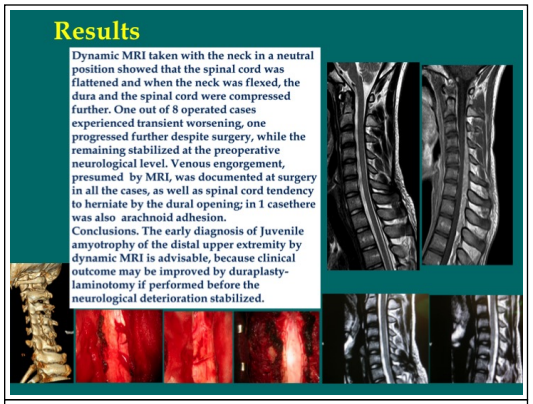
The clinical and radiographic characteristics of 16 patients affected by CFM were examined. Age at onset ranged between 14 and 25 years, with a pick at 16. We observed a male prevalence (13/15), as already reported. Mean age at onset was 16 years. Dynamic MRI and Flexion Neurophysiology were performed before and after surgery. 9 patients deteriorated were operated with laminotomy, expansive duraplasty and suspension; 7 remained stable and were just treated conservatively (collar) and followed

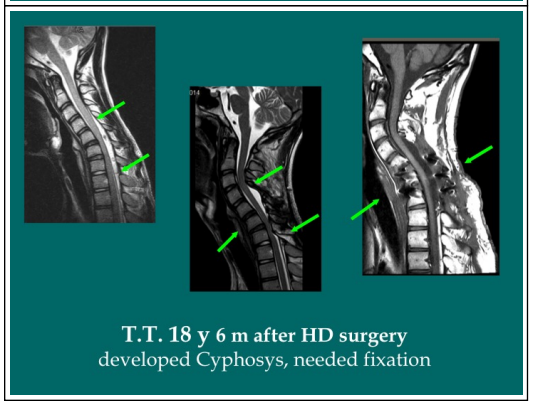
Neutral and fully flexed neck MRI showed posterior dural loss of attachment and cervical spinal cord flattening

Venous engorgement, presumed by MRI, was documented at surgery in all the cases, as well as spinal cord tendency to herniate by the dural opening; in 1 case there was also arachnoid adhesion.

Series

	Age at Diagnosis	Preoperative Deterioration	Type of Surgery	Neurological Outcome
CD	20	Biphasic, Evolutive	Sitting, LT, duroplasty & suspension	Stabilized
CC	18	Amiotrophy, Evolutive	Sitting, LT, duroplasty & suspension	Transient Worse, than Stabilized
DAM	19	4 y, amiotrophy	Prone, LT, duroplasty & suspension	Stabilized
GS	19	Evolutive	Sitting, LT, duroplasty & suspension	Stabilized
RG	20	Evolutive	Sitting, LT, duroplasty & suspension	Stabilized
PN	22	Tremor, Amiotrophy	Sitting, LT, duroplasty & suspension	Stabilized
VN	22	Evolutive	Prone, LT, Duroplasty & suspension	Stabilized
MS	20	6y, progressive amiotrophy	Prone, LT, Duroplasty + Suspension; Reoperation	Worsened
TT	16	Amiotrophy, Evolutive	Sitting, LT, duroplasty & suspension	Stabilized





Results

In our series of HD patients clinical diagnosis was confirmed by dynamic MRI, confirming the importance of neck flexion for proper diagnosis.

All the operaed patients were stabilized by surgery; two experience transient worsening: one was immediate, due to surgical trauma, and improved with fisiotherary; another one experienced delayed worsening, due to progressive instability and improved only after laminoplasty repositioning and anterior stabilization.

Conclusions

The early diagnosis is a milestone because clinical outcome may be strikingly improved by early surgery. If rapidly progressive disease occurs a surgical treatment may be considered before the spontaneous arrest, whilst mild HD may have a conservative approach.

Insyability should be carefully searched and treated by fixation; the role of differential pressure levels between cranium and spinal canal need to be furtherly investigated.