MR Imaging of the Cervical Cord in Juvenile Amyotrophy of Distal Upper Extremity

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We report the MR studies of the cervical cord in seven patients presenting with juvenile muscular atrophy of distal upper extremity. This illness, also known as monomelic amyotrophy or benign focal amyotrophy, is distinct from the other motor neuron diseases. Seen in young males, it is characterized by muscular atrophy of the hand, and usually of the forearm, most often unilateral. The underlying process, of unknown origin, affects the anterior horn cells in the lower cervical cord. The gradual onset of purely motor disturbances may mimic early amyotrophic lateral sclerosis. This latter diagnosis may be excluded because of clinical stabilization and lack of pyramidal tract involvement. In our series, five MR studies were positive. In three cases we were able to demonstrate focal and unilateral atrophy in the lower cervical cord limited to the anterior horn region. Morphologic MR findings correlated with clinical and electromyographic features. In two other cases the MR-clinical correlation was more complex. No pathologic MR signal was detected on either T1- or T2-weighted images.

Although the diagnosis of monomelic muscular atrophy is based on neurologic and neurophysiologic data, MR provides confirmatory evidence as well as useful information contributing to an understanding of this disease.

The appearance of progressive unilateral atrophy of the intrinsic hand muscles suggests muscular or nervous lesions of varying origin. When the clinical examination, electromyogram (EMG) findings, and nerve conduction velocity show evidence of a chronic anterior horn lesion, the most likely diagnosis is a motor neuron disease. Motor neuron diseases, involving the upper and/or the lower motor neurons, are a heterogeneous group of syndromes with various clinical presentations and a predominantly poor prognosis. It has been reported, however, that some young patients develop an anterior horn disease involving only one extremity that has a benign course [1–6]. This illness, corresponding to juvenile amyotrophy of distal upper extremity, was first described in 1959 by Hirayama et al. [1] and constitutes a separate entity among motor neuron diseases. It is also named monomelic amyotrophy [6] or benign focal amyotrophy [7]. The disease selectively affects the anterior horn cells in the lower cervical cord. The clinical characteristics are juvenile male occurrence, insidious onset, and unilateral muscular atrophy in the hand and forearm. The motor deficit and muscular atrophy progress for 1 to 3 years before stabilizing over a sufficient period to underline its benignity. This clinical picture permits differentiation from more severe forms of progressive muscular atrophy. The differential diagnosis includes juvenile spinal muscular atrophy, chronic poliomyelitis, syringomyelia, and amyotrophic lateral sclerosis (ALS). This latter is the most serious disease. A juvenile form of ALS may start with muscular atrophic changes of a hand; progression is usually rapid, with pyramidal tract involvement, and life expectancy is usually not more than 3 years.

Attention to this kind of pathology has been infrequent in the radiologic literature. Improved contrast resolution of the CT scan and the advent of MR have renewed interest in these conditions previously thought to be without radiologically identifiable spinal cord abnormalities.

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MR findings in juvenile amyotrophy of distal upper extremity have not been published previously, and a review of the literature found only four reported cases [7, 8] studied by CT with intrathecal metrizamide. We present the MR findings in our series of seven patients and discuss the value and significance of MR in this disease.

Materials and Methods

The MR studies of seven patients presenting with juvenile muscular atrophy of distal upper extremity were reviewed.

All patients were male, and they ranged in age from 15 to 31 years (average, 20.4 years). Onset of symptoms was between ages 12 and 26 years (average, 18 years). None of the patients had a family history of neuromuscular disease. Six of seven patients were extremely active in sports; four of them were at a competitive level. Sports included volleyball (case 1), soccer and aikido (case 2), karate (case 3), gymnastics and weight lifting (case 4), rugby (case 6), and water skiing and tennis (case 7). Sports activity preceded the onset of symptoms by several years. There was no relation between the involved side and the patient's dominant side.

The patients presented only with severe amyotrophy of one hand and of the distal forearm, causing a motor deficit (Fig. 1). The appearance was that of an Aran-Duchenne hand. Neurologic examination revealed hyperactive, normal, or hypoactive reflexes in the distal involved upper extremity. There were no sensory deficits. Frequently seen were cramps (all patients), fasciculations (cases 1 and 5), vasomotor disturbances (cases 2 and 7), and a fine resting tremor of the fingers (cases 2 and 6). Cold considerably aggravated the findings; in two patients (cases 2 and 3) the deficit initially appeared only in cold weather. All the patients stabilized after a period of evolution lasting 1 to 3½ years. This stabilization was permanent as far as the follow-up of 1 to 8 years (average, 3 years) allowed us to determine.

MR studies were obtained on a superconductive unit* operating at 0.5 T. Multiecho sequences and a multislice program were available. Surface coils were used in all examinations. T1-weighted images, 400/14,26, (TR/sagittal images TE, axial images TE), were obtained in sagittal and axial sections; T2-weighted images, 2000/60, 120 (TR/first echo TE, second echo TE), were obtained in axial sections and, in one patient, also in the sagittal plane. Axial sections were obtained parallel to the disk plane. Two echoes were obtained in the multiecho sequences. Images were reconstructed by 2D Fourier transform on a 256 × 256 matrix. Slice thickness was 5 mm in the sagittal plane and 8 mm in the axial plane.

Cord size was evaluated on both sagittal and axial T1-weighted images. Localized atrophy was defined as a decrease in cord size in comparison to the normal cord above and below the affected level. Sagittal images suggesting atrophy were verified on the appropriate contiguous axial sections, since it is possible to make an erroneous interpretation of atrophy on sagittal cuts when the spinal cord is not truly in midline. On axial images, comparison of the affected level with lower slices is more reliable than with upper slices. The cervical cord normally tapers inferior to the physiological enlargement; thus, increased cord size inferiorly signifies atrophy superiorly (see Fig. 4).

Myelography (in four patients) and nonenhanced cervical CT scans (three patients) were normal. The EMG study invariably demonstrated severe anterior horn involvement of the clinically symptomatic side. However, the asymptomatic side also demonstrated pathologic EMG changes that were mild. Motor and sensory conduction velocities were normal. Antiviral antibody (IgG) tests were negative. CSF was normal.

* CGR Magniscan.

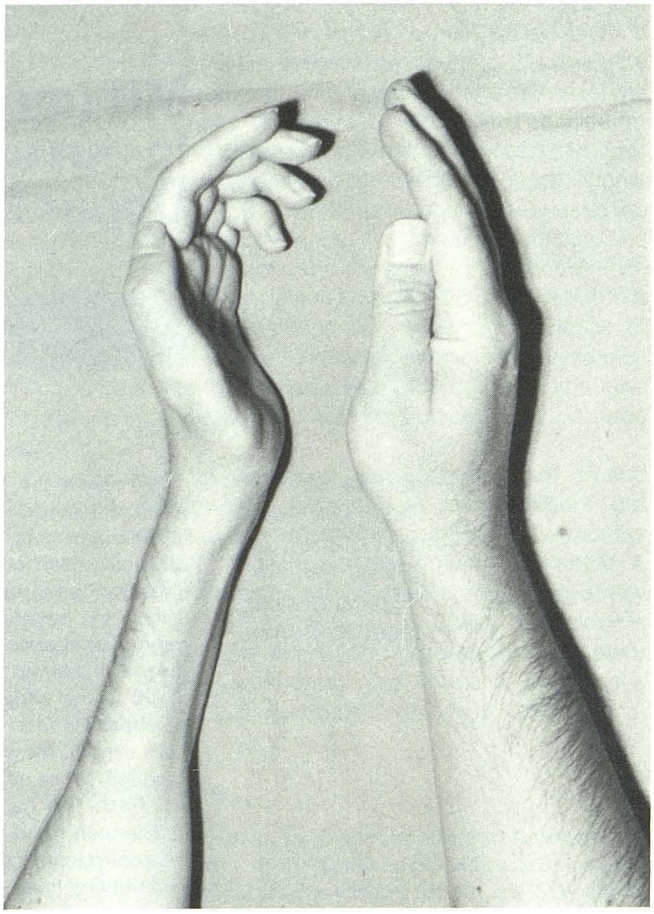


Fig. 1.—Juvenile amyotrophy of distal upper extremity. Typical severe muscular atrophy of left hand and distal forearm.

Results

In five of seven patients MR showed pathologic findings. Morphologic spinal cord changes were well demonstrated on T1-weighted images. In five patients, sagittal T1-weighted images showed a diminished anteroposterior medullary diameter at the lower cervical vertebral level: C6 (case 1), C6–C7 (cases 2 and 6), C7 (case 3), C7–T1 (case 7). The pre- and retromedullary subarachnoid spaces were clearly shown in all cases and no compressive pathology was detected. On axial section at the same levels MR confirmed the atrophic changes of the spinal cord. Three patients (cases 1, 2, and 6) presented with a flattened appearance of the anterior part of the spinal cord, which was limited to one side (Figs. 2 and 3). This finding correlated with clinical distribution. In one patient (case 3) MR defined bilateral anterior atrophy with flattening of the ventral surface. In another patient (case 7) global atrophy was seen at the C7–T1 level and the spinal cord appeared round and small (Fig. 4). In all patients the spinal cord was normal above and below the atrophied region. In particular there was no evidence of syrinx, posttraumatic cyst, or tumor. On T2-weighted images, no pathologic signal intensity was observed. In no patient was there widening of the anterior median fissure.

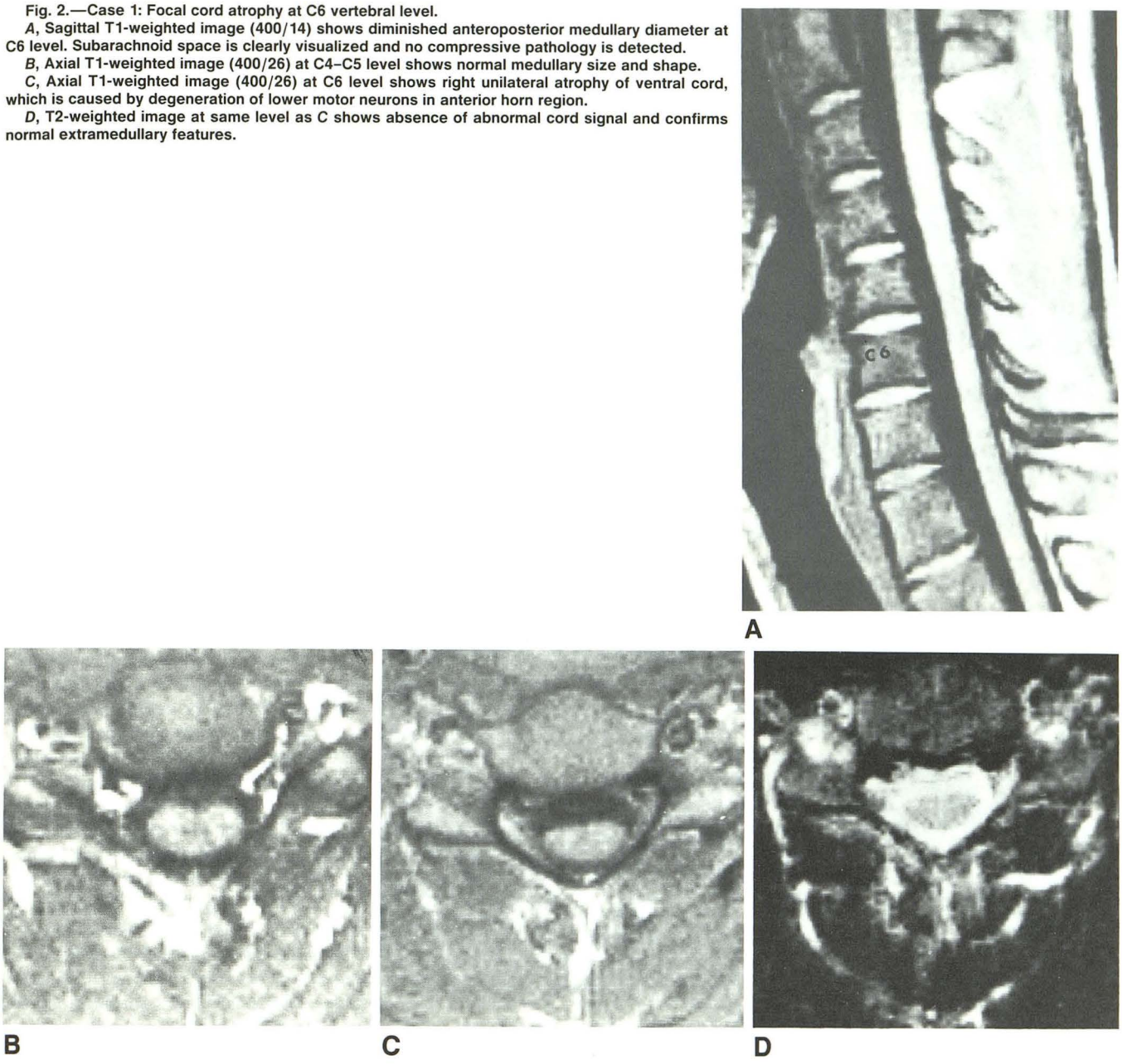
Fig. 2.—Case 1: Focal cord atrophy at C6 vertebral level.

A, Sagittal T1-weighted image (400/14) shows diminished anteroposterior medullary diameter at C6 level. Subarachnoid space is clearly visualized and no compressive pathology is detected.

B, Axial T1-weighted image (400/26) at C4–C5 level shows normal medullary size and shape.

C, Axial T1-weighted image (400/26) at C6 level shows right unilateral atrophy of ventral cord, which is caused by degeneration of lower motor neurons in anterior horn region.

D, T2-weighted image at same level as *C* shows absence of abnormal cord signal and confirms normal extramedullary features.



The two patients (cases 4 and 5) with normal MR examinations had the shortest clinical history. The progression and stabilization periods were comparable to those of the other patients with positive MR studies (Table 1).

Discussion

The value of MR in the evaluation of the spine and spinal cord has been reported [9–12]. MR with short TR and TE permits direct visualization of the cord with sharp anatomic delineation and provides good definition of its size and shape. MR with long TR and TE allows evaluation of signal changes caused by pathologic processes. Gray and white matter usually are not readily distinguishable.

Different types of spinal lesions (such as degenerative, vascular, traumatic, inflammatory) produce distinct patterns of atrophy. In addition, nonspecific atrophy of the cord can be seen with tumors, syringes, AVMs, etc. Except for some cases of cord atrophic changes that occur after trauma, surgery, multiple sclerosis, and ALS [11–14], MR findings in medullary atrophy have not yet been reported. Radiologic features of spinal cord atrophy have been described on CT studies performed after intrathecal metrizamide [8, 15]. The most common patterns are: *diffuse atrophy* (transverse myelopathy), in which the spinal cord is round and small and the anterior median fissure (AMF) is usually not visualized. *Hemiatrophy* (Brown Sequard syndrome), in which one-half of the cord appears normal while the other half demonstrates

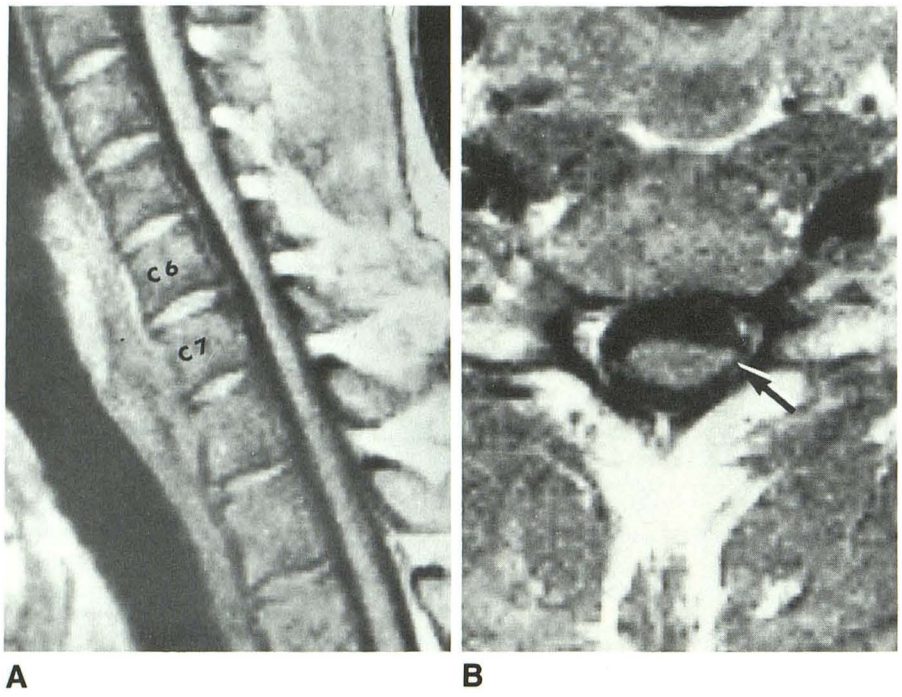


Fig. 3.—Case 2: Focal cord atrophy at C6–C7 vertebral level.

A, Sagittal T1-weighted image (400/14) shows atrophy of spinal cord at C6–C7 level.

B, Axial T1-weighted image (400/26) at C6–C7 level confirms cord atrophy; this latter is unilateral and limited to left anterior horn region. Beaking of left lateral tract is observed (*arrow*).

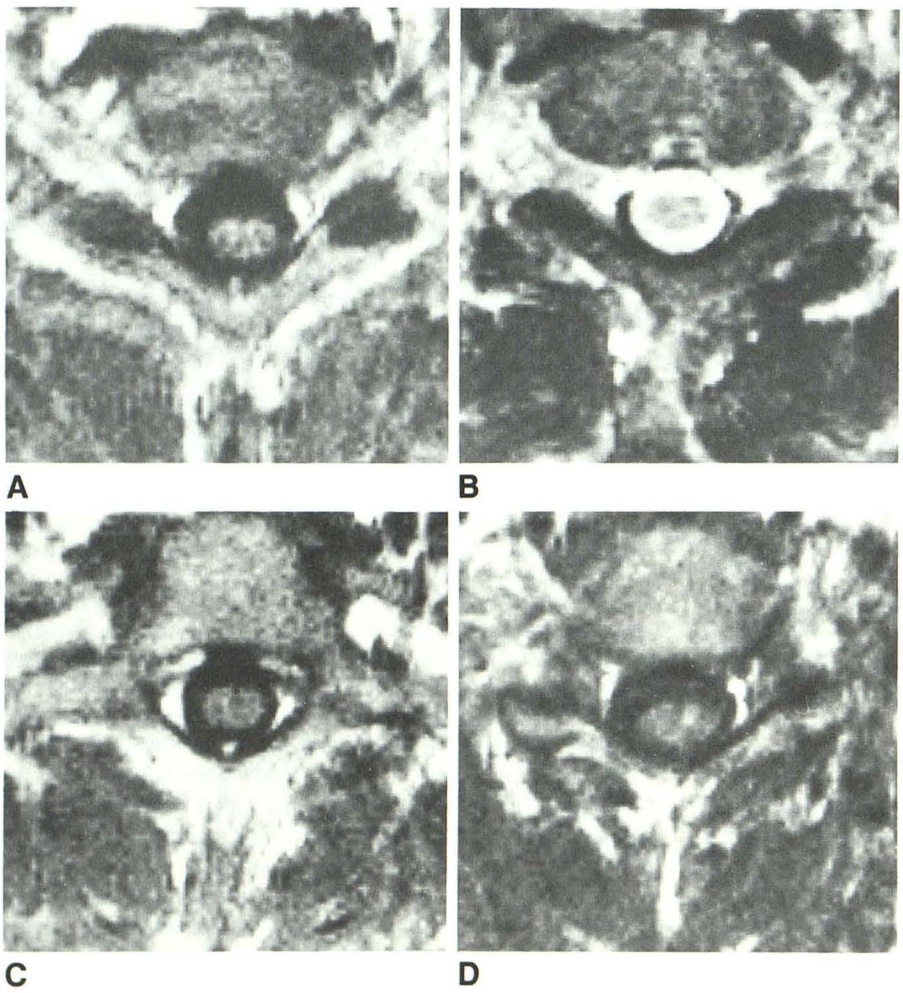


Fig. 4.—Case 7: Medullary atrophy at C7–T1 vertebral level.

A, Axial T1-weighted image (400/26). In this patient with left-sided clinical involvement, global atrophy is seen at C7–T1 level.

B–D, T2-weighted image (*B*) (2000/120) at same level shows no pathologic MR signal intensity. Atrophy at C7–T1 level is clearly seen in comparison with the contiguous lower (*C*) and upper (*D*) T1-weighted images of normal-sized cord.

TABLE 1: Seven Patients with Juvenile Amyotrophy of Distal Upper Extremity

Case No.	Age	Gender	Side Involved	Age at Onset	Duration of Disease (years)	Period of Evolution (years)	Period of Stabilization (years)	MR Atrophy
1	22	M	Right	18	4	3	1	C6, anterior–lateral right
2	25	M	Left	16	9	1	8	C6–C7, anterior–lateral left
3	22	M	Right	18	4	3	1	C7, anterior-bilateral
4	15	M	Right	12	3	1	2	Negative
5	22	M	Left	19	3	2	1	Negative
6	26	M	Right	20	6	1	5	C6–C7, anterior–lateral right
7	31	M	Left	26	5	3, 5	1, 5	C7–T1, global

atrophic changes; the AMF clearly delineates the midline. *Anterior atrophy* (anterior spinal artery syndrome, cervical spondylosis), in which the cord has a decreased anteroposterior diameter and flattening of the ventral surface, giving a bean-like shape; widening of the AMF is also seen. Central flattening and infolding of the cord are typical of spondylosis and may sometimes be produced by direct osteophytic compression; beaking of the cord caused by the acquired relative prominence of the lateral tracts is often observed. *Posterior atrophy*, due to involvement of the dorsal columns, presents as posterior flattening. In these forms the length of cord involvement is variable.

Chronic multiple sclerosis may show diffuse atrophy or focal degeneration due to old demyelinated plaques; accentuation of surface sulci and widening of the AMF have been reported [8]. In *ALS* the anterior and lateral portions of the cord are flattened, reflecting atrophy of both anterior horns and corticospinal tracts; the AMF is normal. Four cases of *monomelic amyotrophy* have been reported on metrizamide CT scans [7, 8] showing unilateral atrophy of the anterior portion of the spinal cord.

In all our patients with positive MR studies, the medullary atrophy was located in the lower cervical cord corresponding to the metameric nervous distribution of the distal upper extremity. In three patients MR in the axial plane demonstrated hemiatrophy limited to the anterior lateral portion of the spinal cord corresponding to the anterior horn (Figs. 2 and 3). This likely reflected the underlying degeneration of lower motor neurons. The lesion correlated with the involved clinical side. In one patient MR showed anterior atrophy on both sides of the spinal cord and the radiologic-clinical correlation was lacking. Our series contains only the unilateral form of the disease, which represents the most common type, but a bilateral form (usually asymmetric, more rarely symmetric) has been described [2, 5, 16]. EMG changes were always bilateral, but predominated on the affected side. In this patient neurologic signs were localized to one hand while EMG and MR findings suggested lesions on both sides. If EMG is a sensitive diagnostic tool in detecting slight involvement of the contralateral anterior horn without neurologic findings, it is more difficult to explain morphologic MR abnormalities lacking clinical expression. In another patient MR demonstrated global atrophic modifications at C7–T1 (Fig. 4). In this case cord atrophy did not correspond to any neurologic feature except for the amyotrophy of one hand. This can be explained by the fact that lower motor neurons of the anterior horns are the most easily injured structures in the cord [16, 17]; but here again the unilateral deficit is not well explained.

Only a single autopsied case of a patient with bilateral asymmetric clinical manifestations has been reported [16]. The gross appearance of the cord was similar to the MR images of our patients. The cord showed a decreased anteroposterior diameter, chiefly at the C7 and C8 root levels. This aspect was due to selective atrophy of the two anterior horns, which was more severe on the side with greater clinical involvement. Histologic findings demonstrated lower motor neuron degeneration. The white matter was well preserved. The anterior roots showed thinning caused by the loss of myelinated fibers, but the myelin sheath was intact.

The two patients whose clinical courses were the shortest, although stabilized, had normal MR examinations. The length of the clinical history appeared to be the determining factor, insofar as MR findings are concerned. The rate of progression and stabilization periods did not seem to correlate well with MR changes and were comparable to those of the patients with positive MR (Table 1). These results suggest that the atrophy, not initially seen on MR, can continue to evolve even after clinical stabilization, and can eventually be discerned.

The origin of juvenile spinal amyotrophy remains unknown. Different theories have been proposed [2, 3, 16]: degenerative, ischemic (anterior spinal artery), viral, and other infectious or aseptic inflammatory causes. The absence of abnormal MR signal in the spinal cord on both T1- and T2-weighted images permits a priori the exclusion of an ischemic anterior spinal artery or inflammatory type lesion. The strenuous physical activities of the patients are striking to us, as they were to Hashimoto et al. [3]. One can speculate on the possible role of repeated subclinical cervical trauma as a cause of chronic microcirculatory disturbances. It is known that the anterior horn cells are most sensitive to arterial or venous ischemia and perhaps the lower cervical cord is most susceptible to vascular compromise (torsion effects with rotatory motion, small spinal canal?).

MR images are not pathognomonic for this disease. Norman [12] reported similar localized atrophy in a patient who had sustained injuries in a motor vehicle accident; unilateral anterior cord flattening was found at the C5–C6 level, while in our cases involvement was more typical in the lower cervicothoracic region. In ALS, MR findings show, in addition to atrophic changes in the anterior horn region, atrophy of the corticospinal tracts, reflecting both the upper and lower motor neuron nature of the process. Some authors [14] have re-

ported a hyperintense signal in the cord. We think this hyperintensity could be due to demyelinating changes associated with the axonal disease. In ALS, degeneration of the myelin sheath of corticospinal fibers and also demyelination in the lateral columns extending beyond the region of the pyramidal tract have been described [18]. The discrepancy with our results lacking MR signal hyperintensity may be explained by the absence of myelin in the anterior horn cells, resulting in a purely degenerative atrophic process. The myelin sheath of the axons of these cells begins in the intramedullary portion of the anterior root [19]. Although in the anterior roots there is a reduction of axonal fibers, the myelin seems intact [16, 20]. If there should be slight demyelination, this may not be revealed on MR images. Differential diagnosis between an early ALS and juvenile amyotrophy of distal upper extremity is based above all on clinical findings. In the latter, involvement remains localized at the lower cervical level after a period of progression lasting 1 to 3 years. ALS (including the rare juvenile form) may start with unilateral hand atrophy, which progresses to pyramidal tract involvement. The course of this disease is relentless.

In conclusion, we have presented the first series of patients with juvenile amyotrophy of distal upper extremity studied by MR. The diagnosis of this disease is based on neurologic findings and EMG studies. MR provides confirmatory evidence. It reveals structural alterations in the lower cervical cord, reflecting the underlying atrophy of the anterior horn and correlating with clinical and neurophysiological data. Evaluation of MR findings and their temporal changes may contribute to a better understanding of the disease in its pathophysiological and historical aspects.

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