Acute Transverse Myelopathy in Children

Coleen Adams and Derek Armstrong

ABSTRACT: Twenty-three children with acute transverse myelopathy (ATM) are reviewed. Antecedent minor trauma or exercise was reported in 10 patients. Despite a positive history in 7 patients no preceding infection was documented. Two patients had a history of less severe ATM followed by recovery prior to a second more severe episode. The most common initial symptom was back pain and the most prominent clinical signs were weakness, sensory level and sphincter disturbances. Myelography and CT myelography at presentation was performed to exclude a compressive lesion. Spinal cord enlargement was demonstrated in 6 of 21 cases. Magnetic resonance imaging (MRI) of the spinal cord, performed in one patient, showed enlargement of the cord. Poor prognostic features were severity of weakness at the time of maximum deficit and a delayed onset of recovery. Maximum motor recovery occurred at a mean of 6 1/2 months but did not occur in one patient until 1 1/2 years. Normal or good outcome was obtained in 64%.

RÉSUMÉ: Myélopathie transverse aiguë chez les enfants Nous revoyons les cas de 23 enfants atteints de myélopathie transverse aiguë (MTA). Un traumatisme mineur ou un exercice préalable avait été rapporté chez 10 patients. Malgré une histoire positive chez 7 patients, aucune infection antérieure n'a été documentée. Deux patients avaient une histoire de MTA moins sévère, suivie d'une guérison, antérieure à un deuxième épisode plus sévère. Le symptôme initial le plus fréquent était une douleur dorsale et les signes cliniques les plus évidents étaient la faiblesse et les perturbations sensitives et sphinctérielles. Une myélographie et une myélographie par CT ont été pratiquées à la consultation initiale afin d'éliminer une lésion pas compression. Une augmentation de volume de la moelle épinière a été mise en évidence chez 6 des 21 cas. L'imagerie par résonance magnétique de la moelle épinière, pratiquée chez un patient, a montré une augmentation du volume de la moelle. La sévérité de la faiblesse au moment où le déficit était à son maximum et un processus de récupération à début tardif étaient associés à un pronostic sombre. La récupération motrice était à son maximum à 6 mois et demi en moyenne, alors que dans 1 cas elle ne s'est produite qu'à 1 an et demi. Chez 64% des cas, il y a eu un retour à la normale ou une bonne récupération.

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Acute transverse myelopathy (ATM) has been reported in 2 large pediatric series,[1,2] in combined adult and pediatric reviews[3-5] and in adult series.[6] Onset has been noted to follow febrile illness, suggesting a post viral etiology. Minor antecedent trauma with delayed onset of signs of ATM suggests an ischemic etiology with involvement of the anterior spinal artery.[7,8] Other associations include multiple sclerosis (MS),[3-6] systemic lupus erythematosus[9,10] and mycoplasma.[11,12] Prognosis in children is better than in adults.[1,2] We describe the preceding events, clinical course, investigations and long-term outcome in 23 children with ATM.

Methods

The charts of children admitted to The Hospital for Sick Children Toronto between 1960-1988 with the diagnosis of ATM were reviewed. Criteria for inclusion were age up to 15 years, no known antecedent neurological disease, acute onset of a spinal cord syndrome and myelography to exclude a compressive spinal cord lesion except where recovery was rapid. Patients with a known cause of acute myelopathy, such as trauma or irradiation, were excluded.[2] Patients with encephalitis as the main feature were excluded. Clinical histories, noting antecedent illness and trauma, were reviewed. The initial symptom, duration of symptoms to maximum deficit, and signs at maximum deficit along with investigations were reviewed. Myelography was reviewed by one neuroradiologist (DA). An oil-based contrast was used as the myelography agent in 10 patients and a water soluble contrast in 11 patients. CT myelography was used in 8 patients. Follow-up was by contacting the

Table 1: Preceding Symptoms and Onset

	Number of Patients	
Illness in previous 3 weeks	7	(30%)
Preceding minor trauma	4	(17%)
Preceding exercise	6	(26%)
First Symptom:		
thoracic pain	10	(43%)
limb pain	9	(39%)
limb paresthesias	3	(13%)
weakness	4	(17%)
Temp >38.5°	2	(9%)
Nuchal rigidity	8 of 20	(40%)

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patients or their family doctor or from chart documentation. Time from maximum deficit to recovery was noted. Degree of recovery was graded as: normal; good (slight limp, minor sensory loss but no significant functional impairment); fair (walking with marked limp or with aid of apparatus, permanent hypesthesia, some but not constant sphincter control); poor (completely or largely immobilized, permanent anesthesia, only partial sphincter control of "automatic type").[1]

Results

Twenty-three children satisfied the criteria for inclusion. Age of onset was from 19 months to 14 years (mean 9.4 years). The preceding symptoms and clinical onset are outlined in Table 1. Preceding minor trauma occurred just prior to symptoms in one child and at 1, 2 and 12 hours in 3. Four children were exercising at the onset of symptoms and another 2 had been skiing in the previous 24 hours. In 2 patients a quickly resolving first episode of ATM was followed in 12 days and 6 weeks respectively by a second more severe episode. Of the total group the mean time from onset to maximum deficit was 50 hours (range 5 mins to 10 days). Where minor trauma or exercise was a possible contributing factor the mean time from onset to maximum deficit was 23 hours (range 1 to 24 hours).

Symptoms and signs at the peak of the illness are outlined in Table 2. Reflexes were reduced or absent in the legs in 15

Table 2: Symptoms and Signs at Peak Involvement

Patient	Grade		Strength		Respiratory Involvement	Abdominal Reflexes		Sensory Level		Retained Vibration + Proprioception	Urinary Symptoms	Bowel Symptoms
	Arms		Legs									
	Rt	Lt	Rt	Lt		U	L	Rt	Lt			
1.	0	2	0	0	diaph	0	0	C_3	C_6	–	I	No
2.	5	5	0	0	No			T_3	T_3	Yes	I	Yes
3.	0	0	0	0	vent	0	0	–	–	–*	C	Yes
4.	4	4	0	0	No	0	0	C_7	C_7	–	C	Yes
5.	5	5	4	4	No	0	0	T_6	T_6	–	C	Yes
6.	5	5	0	0	No	√	0	T_{12}	T_{12}	Yes*	C	Yes
7.	5	5	0-3	4	No	0	0	L_1	L_1	No	I	Yes
8.	5	5	0	2	No	√	0	T_{11}	T_{11}	No	C	Yes
9.	4	3	5	4	No	√	√	T_4	No	Yes*	I	No
10.	5	5	0-2	0-2	No	√	√	S_3	S_3	–*	C	Yes
11.	3	4	0	0	No	0	0	T_1	T_1	–*	C	No
12.	5	5	1-2	1-2	No	0	0	T_8	T_8	No	C	Yes
13.	2-3	2-3	2-3	3-4	diaph	–	–	T_4	T_4	Yes*	C	Yes
14.	5	3	0	0	diaph	0	0	T_3	T_6	Yes	C	Yes
15.	4	4	3-4	3-4	No	√	√	C_6	C_6	–	C	No
16.	decreased throughout				No	0	0	C_4	C_4	–	C	Yes
17.	5	5	0	0	No	√	0	T_{10}	T_{10}	–*	C	Yes
18.	3-4	3-4	2	2	No	0	0	C_5	C_5	–	I	No
19.	5	5	3	3	No	0	0	T_8	T_8	Yes*	C	Yes
20.	0-2	3-4	0-2	3-4	No	0	0	C_7	C_7	Yes*	C	Yes
21.	1	2	0	2	No	RO L√	RO L√	T_4-T_{10}	T_4	Yes*	No	No
22.	0	0	0	0	vent	0	0	C_4	C_4	No	C	Yes
23.	3	2	4	3	vent	0	0	C_3	C_3	Yes	C	No

Grade strength from Medical Research Council Memorandum No. 45 London.
diaph — diaphragmatic respiration
vent — ventilated

Abdominal reflexes 0: absent; √: present; U: upper; L: lower; Rt: right; Lt: left

Sensory level C: cervical; T: thoracic; L: Lumbar; S: Sacral

Urinary symptoms I: incontinence; C: bladder catheter

* history of preceding minor trauma or exercise.

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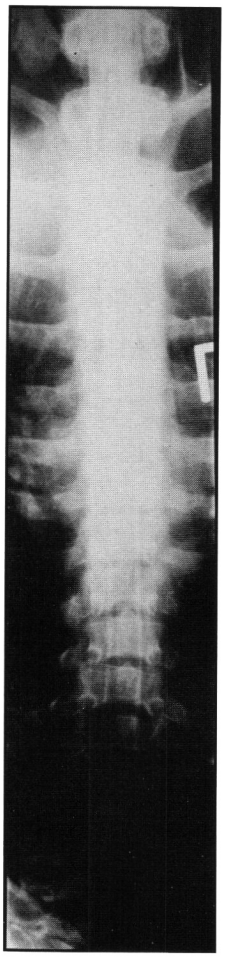

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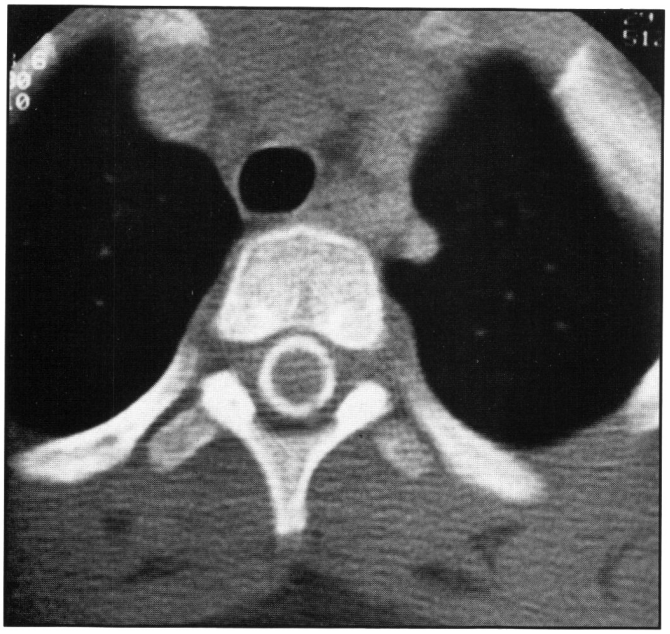


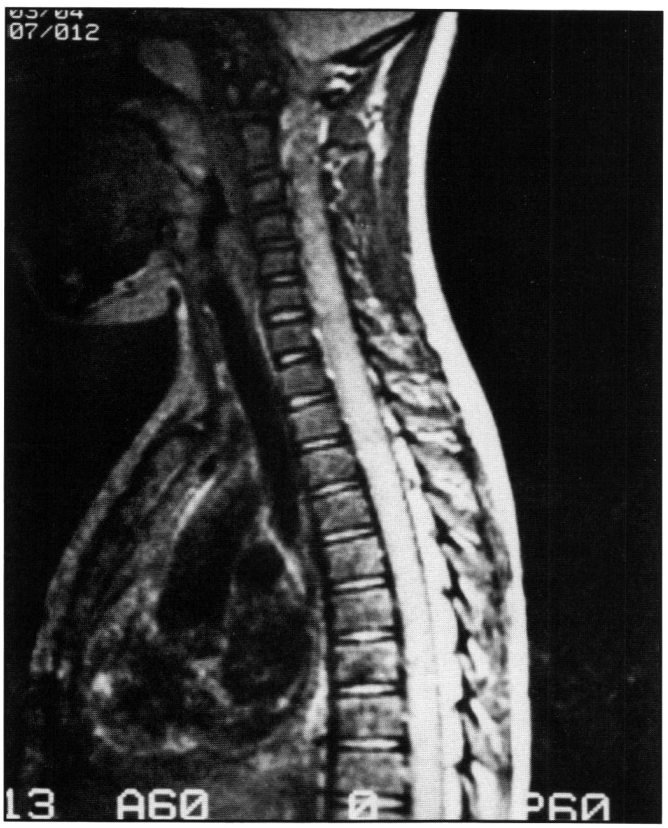
B

Figure 1(a) — Myelography of patient 22 showing diffuse enlargement of spinal cord from cervical cord (C_4) to conus.

Figure 1(b) — CT upper thoracic spine (post myelography) showing swollen spinal cord.

Figure 1(c) — MRI (TR 1364, TE 35) cervical and thoracic spine showing diffuse enlargement of spinal cord from the level of C_3.

Figure 1(d) — MRI (TR 500, TE 20) thoracolumbar spine showing diffuse enlargement of spinal cord to the level of the conus.



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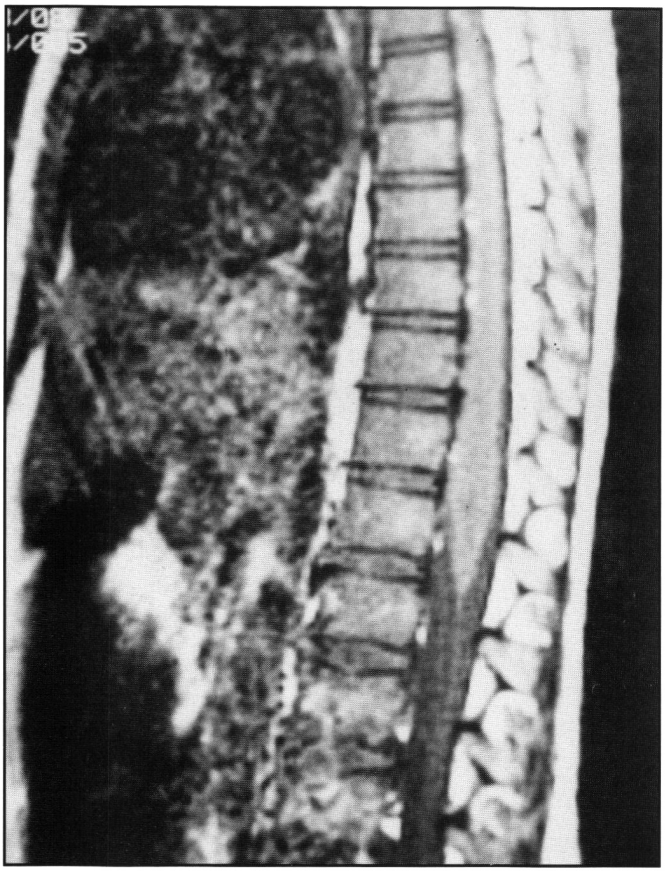

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patients. In 8 the reflexes in the legs were increased when first tested. A sensory level was not noted in one child (#3) but the other clinical features of initial bladder symptoms and long-term outcome of a spastic paraparesis could not have been due to another illness such as Guillain-Barré syndrome. Sensory loss was asymmetric in 4 and motor findings were asymmetric in 10. Urinary retention was never the first symptom but always occurred within the first 2 days.

Twenty-one patients had myelography. One who was improving by the time he was admitted and another who had a lumbar puncture to exclude a manometric block did not have myelography. Myelography was performed between 1 and 15 days (mean 3.5 days) after onset of symptoms. Myelography was normal in 15 of 21. Five myelograms showed localized enlargement of the cervical or upper thoracic cord over 2 to 10 spinal cord segments (mean 6.8 segments). One showed diffuse cord enlargement from the third cervical segment to the conus (#22). This patient also had magnetic resonance imaging (MRI) which showed the same diffuse enlargement as seen on CT myelography (Figure 1). Cerebrospinal fluid (CSF) was drawn in all either before or at the time of myelography. Eleven had normal CSF protein. Of the 12 elevated protein levels the range was 0.43 to 1.84 g/L (mean 0.85 g/L). CSF leukocyte counts were normal in 14. Eight had moderately raised CSF leukocyte counts. Both lymphocyte and polymorph counts were elevated. CSF bacterial culture was negative in all. CSF viral culture was negative in 7, however only 2 patients with a preceding illness had CSF viral cultures. CSF oligoclonal banding in 2 patients showed more than 2 bands in one. Blood viral titers were not elevated in 11 patients tested. Of these 11 patients 3 had a preceding illness. Blood mycoplasma titers in 4 patients and cold agglutinins in another 4 patients were negative. Seventeen patients were given steroids. Dexamethazone and prednisolone at approximate doses of 0.5 mg/kg/d and 1 - 2 mg/kg/d respectively were used. Other steroids included hydrocortisone and adrenocorticotropic hormone in one case each. Duration of medication was for a mean of 13.5 days (range 3 days to 2.5 months). Effect of steroids could not be assessed.

Follow-up was obtained in 22 patients. Length of follow-up was 1 month to 17 years (mean 5.8 years). The start of recovery after maximum deficit in 22 varied from 1 day to 1 month with a mean of 5.9 days. Seventeen children walked unsupported. Of these the time of walking after the maximum weakness in 14 ranged from 3 days to 8 weeks with a mean of 16 days. Time at best motor function was documented in 13. The range of duration of improvement was 5 days to 1.5 years with a mean of 6.5 months. Of the 22 patients followed the bladder had been affected in 21. Of 14 subsequently reported normal functioning bladders 11 had documentation of time of recovery. Of these 7 had recovery under 11 days and 4 had improvement after 7 weeks with one patient reporting improvement up to one year. Urological investigations were performed in few. Satisfactory details of return of sensation was not obtained. Final outcome was normal in 7 (32%), good in 7 (32%), fair in 3 (13%) and poor in 5 (23%).

Patient number 1 died 11 years after ATM. At autopsy sections of the spinal cord at the low cervical region showed nearly total myelin and neuronal loss with a marked fibrillary gliosis. Above this level there was myelin loss and gliosis in the sensory tracts and corticospinal tracts and below this level there was loss of myelin and gliosis in the corticospinal tracts.

Discussion

Previous reports of ATM have reviewed children alone,[1,2] adults and children together[3-5] or adults alone.[6] In pediatric reports the incidence of preceding illness is 38% and 60%[1,2] and 35% in an adult report.[6] The illness may have settled prior to ATM suggesting a postinfectious autoimmune response. Reported associated infections include measles, varicella,[1] mumps,[13] rubella,[14] herpes simplex and zoster,[2,5,15] echo virus,[16] CMV and hepatitis A[17] and mycoplasma.[11,12] Other viral causes are listed.[17] Seven or 30% of the patients in the present series had a preceding illness. In 4 the illness had settled before the onset of the ATM. No seasonal variation was noted.

Minor antecedent trauma has been reported. In one pediatric review this occurred in 8%.[1] In combined pediatric and adult reports minor trauma occurred in 8% and 4.8%.[3,5] In the present report 4 patients (17%) had minor trauma. The age range in these 4 patients was 19 months to 5 years with a mean of 4.1 years. Six patients (26%) had been exercising prior to symptoms. Of these 6 exercising patients the age range was 10 to 14 years with a mean of 13 years. This age difference probably reflects the incidence of minor trauma in younger children and exercise in older children. Vibration and proprioception were preserved in the 6 patients with antecedent exercise. The 4 patients with minor antecedent trauma did not have vibration and proprioception documented. Sudden onset of myelopathy following minor trauma may be due to the anterior spinal artery syndrome with preservation of vibration and proprioception. This has been previously reported in children.[7,8] Also non-traumatic anterior spinal artery syndrome has been reported.[18,19] The present review did not find antecedent trauma or exercise to be a prognostic factor in outcome.

The history of an initial episode of ATM which recovered and then recurred with a more severe episode has not been reported previously. One patient was a 13-year-old girl (#2) who had no illness prior to her initial episode of ATM. Her initial episode involved thoracic pain and paraplegia which resolved over 3 weeks. Three weeks after her recovery she had a recurrence with flaccid paraplegia developing over 5 minutes. Her CSF protein and cells were normal. Oligoclonal banding was not performed. Myelography was normal. Her outcome was poor. MS was not diagnosed in her 5-year follow-up. The other patient (#3) was a 14-year-old boy who awoke with his initial episode. He had thoracic pain, paraplegia and a thoracic sensory level with preservation of vibration and proprioception. CSF had normal protein and cells. Recovery was almost complete over 3 weeks when he had a severe recurrence while riding his bicycle. CSF and myelography were again normal. Outcome was fair at 3-year follow-up. He had no progression to suggest MS. In these 2 patients the intermittent course may have been due to ischemia, initially intermittent, subsequently becoming complete. This explanation is favoured by the finding of preservation of vibration and proprioception in both, which is in keeping with the anterior spinal artery syndrome. Neither MS nor vasculitis appear to be involved.

Previous reports have noted varied durations of symptoms to the time of maximum deficit.[1-6] Apart from 2 reviews[3,4] each of

which reported patients with delayed evolution, all others reported maximum deficit within 2 weeks. One review reported that acute onset, with development of maximum deficit with 12 hours, had a worse prognosis.[4] In the present review the 5 patients with a poor outcome had a mean time to maximum deficit of 24 hours compared to the overall mean time to maximum deficit of 50 hours. In the present review rapid development did not correlate with antecedent trauma nor exercise. In previous reports the main symptom at the peak of the illness was weakness of legs in all cases with arms involved in one-third to one-half of cases where reported.[1-6] In the present review leg weakness was also the most prominent symptom. Legs were involved in all and arms in 14 (61%). In those 7 patients with flaccid legs outcome was poor in 3, fair in 2 and good in 2. In the 2 patients who had flaccid arms and legs the outcome was poor in both. Therefore those patients who were flaccid had a worse outcome.

In previous reports CSF was either normal or had increased protein or white cell count. In all reports the CSF findings did not correlate with outcome.[1-6] In the present report the CSF cell count did not correlate with severity of symptoms or outcome. Also, the CSF protein did not generally correlate with severity except for the 2 patients who were completely flaccid and required ventilation (#3, 22). Both of these patients had CSF protein elevated to >1.8 g/L. This degree of elevation did not occur in the other patients. It is possible that these patients may have had a myelo-radiculopathy. CSF protein did not correlate with outcome. The two patients with CSF protein >1.8 g/L had poor outcome, however 2 patients who had normal CSF protein also had poor outcome.

In the large reports of ATM with myelography a total of 139 myelograms were performed. 5% of myelograms were abnormal with spinal cord widening being the most common finding.[2-6] Myelographic findings did not correlate with outcome. CT myelography was not used in these patients. More recent reports have shown the usefulness of CT myelography and MRI in ATM.[14,20,21] The present review had 6 of 21 myelograms showing cord widening over a varying number of interspaces and involving the total cord in patient 22. This patient also had an MRI and both myelogram and MRI demonstrated total spinal cord widening. This patient had a poor outcome. Otherwise myelographic findings did not correlate with outcome. In 5 patients with abnormal myelography the outcome was normal or good. The increased incidence of widening found in the later years of this review demonstrates the value of CT myelography. MRI which is non-invasive will replace CT myelography for these types of spinal cord studies.

In the present review the start of recovery occurred sooner in those patients with a better outcome. The start of recovery from the time of maximum deficit in those with a normal or good outcome varied from 1 day to 1 week with a mean of 2.8 days. In those with a poor or fair outcome the start of recovery was at 4 days to 1 month with a mean of 11.7 days. In previous reviews maximum recovery is usually by 3 to 6 months.[1-6] However, recovery has been reported to occur for 16 months to 4 years.[1,4,6] In the present review the time of maximum motor improvement in 13 patients ranged from 5 days to 1 1/2 years with a mean of 6 1/2 months.

It has been reported that ATM may be the first sign of MS. In previous reviews incidence of MS is from 0% to 14.6%.[2-6] In the present review with a mean follow-up of 5.8 years no patients developed MS. However, in the initial chart review one patient, who presented as a possible case of transverse myelitis developing over a period of 3 weeks, relapsed after discontinuation of steroids and also developed a visual acuity deficit. She had a subsequent diagnosis of MS and was excluded from the study.

Table 3. Degree of Recovery in Previous Reports

Ref	Number	Normal	Good	Fair	Poor
1.	25 (c)	32%	28%	24%	12%
2.	21 (c)	38%	19%	43% fair or poor	
3.	67 (a+c)	1/3		1/3	1/3
4.	52 (a+c)	33%		42%	25%
5.	62 (a+c)	39%		36%	25%
6.	34 (a)	31%		31%	38%, 3 died, 3 no FU

c: children
a: adults
FU: follow-up

As can be seen from Table 3 a normal or good outcome occurred in 60% and 57% in the pediatric reviews. The present result of 64% with normal or good outcome is in agreement.

Conclusion

This is the third reported series of ATM in children. A preceding infectious agent was not documented despite a positive history in 30%. In 43% antecedent minor trauma or exercise, with the clinical sign of preservation of vibration and proprioception in 60%, may have contributed to the anterior spinal artery syndrome. Two patients had an intermittent course of possible ischemic etiology. No children apparently developed MS on follow-up. ATM probably has several etiologies including infectious or post infectious, vascular and demyelinating.

We recommend CT myelography, or preferably MRI of the spinal cord where available, at presentation of a spinal cold syndrome to exclude a compressive lesion and to document the extent of the spinal cord involvement. Spinal cord enlargement, however, was not prognostically valuable.

Poor prognostic features were severity of weakness at the time of maximum deficit and delay in onset of recovery. Normal or good outcome was obtained in 64%.

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References

1. Paine RS, Byers RK. Transverse myelopathy in childhood. Am J Dis Child 1953; 85: 151-163.
2. Dunne K, Hopkins IJ, Shield LK. Acute transverse myelopathy in childhood. Dev Med Child Neurol 1986; 28: 198-204.
3. Altrocchi PH. Acute transverse myelopathy. Arch Neurol 1963; 9: 111-119.
4. Ropper AH, Poskanzer DC. The prognosis of acute and subacute transverse myelopathy based on early signs and symptoms. Ann Neurol 1978; 4: 51-59.

5. Berman M, Feldman S, Alter M, et al. Acute transverse myelitis: Incidence and etiologic considerations. Neurology 1981; 31: 966-971.
6. Lipton HL, Teasdall RD. Acute transverse myelopathy in adults. Arch Neurol 1973; 28: 252-257.
7. Blennow G, Stark L. Anterior Spinal Artery Syndrome. Report of seven cases in childhood. Pediatr Neurosci 1987; 13: 32-37.
8. Ahmann PA, Smith SA, Schwartz JF, et al. Spinal cord infarction due to minor trauma in children. Neurology 1975; 25: 301-307.
9. Warren RW, Kredich DW. Transverse myelitis and acute central nervous system manifestations of systemic lupus erythematosus. Arthritis Rheum 1984; 27: 1058-1060.
10. Pedersen C, Bonen H, Boesen F. Transverse myelitis in mixed connective tissue disease. Clin Rheumatol 1987; 6: 290-292.
11. MacFarlane PI, Miller V. Transverse myelitis associated with mycoplasma pneumoniae infection. Arch Dis Child 1984; 59: 80-82.
12. Cotter FE, Bainbridge D, Newland AC. Neurological deficit associated with mycoplasma pneumoniae reversed by plasma exchange. Br Med J 1983; 286: 92.
13. Silverman AC. Mumps complicated by a preceding myelitis. Report of a fatal case. N Engl J Med 1949; 241: 262-266.
14. Bitzan M. Rubella myelitis and encephalitis in childhood. A report of two cases with magnetic resonance imaging. Neuropediatrics 1987; 18: 84-87.
15. Wiley CA, VanPatten PD, Carpenter DM, et al. Acute ascending necrotizing myelopathy caused by herpes simplex virus type 2. Neurology 1987; 37: 1791-1794.
16. Barak Y, Schwartz JF. Acute transverse myelitis associated with ECHO Type 5 infection. Am J Dis Child 1988; 142: 8.
17. Tyler KL, Gross RA, Cascino GD. Unusual viral causes of transverse myelitis: Hepatitis A virus and cytomegalovirus. Neurology 1986; 36: 855-858.
18. Foo D, Rossier B. Anterior spinal artery syndrome and its natural history. Paraplegia 1983; 21: 1-10.
19. Puntis JWL, Green SH. Ischemic spinal cord injury after cardiac surgery. Arch Dis Child 1985; 517-520.
20. Awerbuch G, Feinberg WM, Ferry P, et al. Demonstration of acute post-viral myelitis with magnetic resonance imaging. Pediatr Neurol 1987; 3: 367-369.
21. Merine D, Wang H, Kumar AJ, et al. CT myelography and magnetic resonance imaging of acute transverse myelitis. J Comput Assist Tomogr 1987; 11: 606-608.

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