CASE REPORT

ACUTE TRANSVERSE MYELITIS IN CHILDREN

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A case of Acute Transverse Myelitis (ATM) is presented. ATM is a rare disease in childhood. The diagnosis stems from the clinical presentation, cerebrospinal fluid findings, appearance of the spinal cord on imaging and ruling out differential diagnoses like Guillain-Barre syndrome (GBS) and Poliomyelitis. The proposed treatment is intravenous (IV) methyl prednisolone with variable chances of recovery. A controlled multicenter study is suggested to assess epidemiology, etiology and prognosis of ATM.

Key Words: Transverse Myelitis, Sensory level, Methyl Prednisolone.

INTRODUCTION

Acute Transverse Myelitis (ATM) is a rare disease in childhood and adolescence and the incidence has been estimated at 1.34 in 1,000,000 people in Israel[1] but there has been no specific evaluation of pediatric incidence. It is characterized by bilateral spinal cord dysfunction presenting as lower extremity weakness with or without sensory symptoms and bladder dysfunction. It typically manifests over a period of hours to one week.[2, 3] Spinal magnetic resonance imaging (MRI), electrophysiological tests and cerebrospinal fluid (CSF) analysis are performed to diagnose ATM and exclude other treatable conditions. ATM can be caused by a number of disorders including trauma, space occupying lesions, vascular malformations, occlusive vasculitic disorders causing infarction of the spinal cord [4, 5, 6] autoimmune diseases[7, 8] and infections either bacterial, viral or spirochetal.[9] ATM has been described after infections with *Epstein-Barr* virus, cytomegalovirus, cytomegalovirus in immune compromised patients, concurrent cytomegalo- and herpes simplex virus infections, rubella, chickenpox, infectious mononucleosis, and measles.[2]ATM can be the initial presentation of acute lymphoblastic leukemia.[10] In our country, Poliomyelitis and Guillain-Barre syndrome remain the two most important differential diagnoses. After diagnosis of ATM, high dose IV steroid pulse therapy is the most promising treatment. Prognosis is variable and residual symptoms are common.

CASE REPORT

A previously well 13-year-old boy, vaccinated against poliomyelitis as an infant, developed weakness of both lower limbs and inability to pass urine twelve hours before presentation to our emergency room. He had prodromal symptoms of fever, headache, pain in the neck and back for two days and vomiting for one day. There was no history of trauma immediately preceding this illness and no photophobia. On admission, he was conscious and fully oriented. He was not able to stand and bear weight on both lower limbs. Tone was decreased in both lower limbs. Power was grade 5/5 in both upper limbs but grade 2/5 in right and 3/5 in left lower limb. Reflexes in the upper limbs were normal, knee jerks were absent bilaterally. Both ankle jerks were present. Planter reflex was flexor on the right and extensor on the left. Abdominal reflex was absent and urinary bladder was palpable up to umbilicus so he was catheterized. The signs of meningeal irritation were positive. No sensory level was identified on day one. Position sense was intact. The differential diagnoses considered at this point were GBS, Poliomyelitis and ATM.

His basic lab work up was unremarkable (Table 1). His Cerebrospinal fluid analysis revealed raised proteins with lymphocytic pleocytosis and normal glucose. Electromyography and Nerve conduction was normal with normal F wave latency.

Examination on day 2 showed that he had a sensory level at the midthorax (T_4). However, he had some patchy sensations present in both legs. Magnetic Resonance Imaging (MRI) of thoracolumbar spine was done and an increased signal was seen (Fig 1&2) throughout the cord except upper cervical region. Focal areas of enhancement were also seen throughout the cord, more in the lumbosacral area. A final diagnosis of Transverse Myelitis was made and he was given intravenous Methyl Prednisolone (IVMP) in a dose of 1g/1.73 m^2/day for five days.

On the third day, he started improving; he was able to stand with support and bear weight on both feet and was able to feel bladder distention. Urinary catheter was removed. He was discharged on oral prednisolone 1mg/kg/dose once daily for two weeks. At follow up a week after discharge he still had some residual feeling of incomplete bladder emptying, had started walking but was still unable to run, he was able to get up from the floor without support and climb stairs. Power in both lower limbs was 4/5. No sensory loss was present. After his two weeks of Prednisolone, steroids were tapered over the next two weeks and stopped. The child was followed

again after two months. By this time he had no complaints and had a normal neurological examination.

Table-1:Laboratory investigations

Hematology:
Hemoglobin: 14g/dl
Total Leukocyte Count: 13 x 10E9/L
Neutrophils: 71%
Lymphocytes: 19%
Platelets: 314 x 10E9/L
Biochemistry:
Random Blood Sugar: 117 mg/dl
Blood urea Nitrogen: 9 mg/dl
Creatinine: 0.7 mg/dl
Sodium: 141 mg/dl
Potassium: 4.7 mg/dl
Bicarbonate: 27 mg/dl
Calcium: 10 mg/dl
Cerebrospinal fuid analysis:
Glucose: 65 mg/dl
Chloride: 126 mg/dl
Proteins: 125 mg/dl
Total Leucocyte Count: 18/mm^3
Neutrophils: 5%
Lymphocytes: 95%
Red Blood Cells: 120/mm^3
Opthalmology consult: Normal (was done to exclude Multiple Sclerosis and Devics disease*)
Antinuclear and Anti ds DNA antibodies: Negative
Electrophysiology:
Visual evoked potentials: Normal
Somatosensory evoked potentials: Normal
Electromyography and Nerve Conduction: Normal study with no electrophysiological evidence of neuropathy.

*Devics disease is also called neuromyelitis optica. It is a combination of symptoms of optic neuritis and ATM.

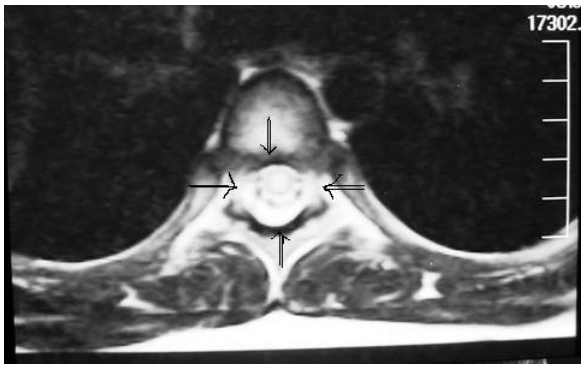

Fig 1: MRI of the spine showing increased signal in the cord

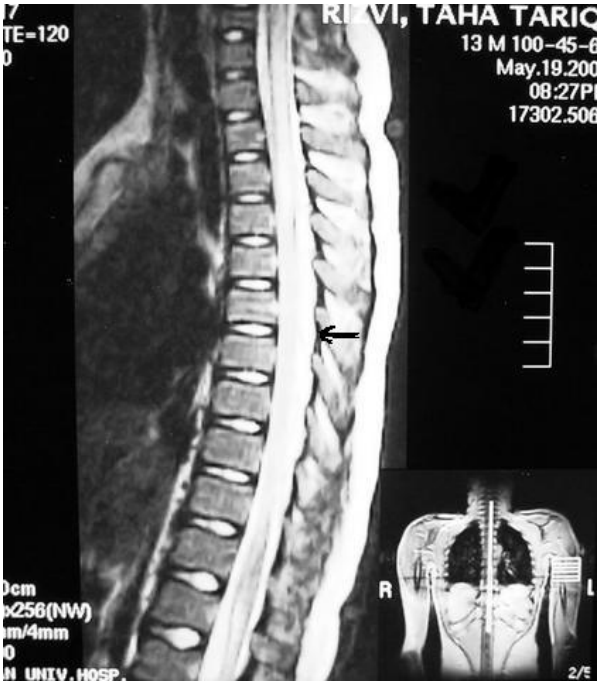

Fig 2: MRI showing increased signal throughout the cord except the cervical region.

DISCUSSION

The case that we have described experienced an acute loss of spinal cord function. All functions of the spinal cord including sensation, motor activity and sphincter control were disturbed to different degrees. Full-blown disease occurred within three days after onset of prodromal symptoms. All these findings are consistent with cases reported in literature. [11]

Our case had similar prodromal symptoms of fever, muscular ache and vomiting as described in literature.[11] Like other case reports our patient had paresis of both legs as the presenting symptom but did not have paresthesias.[11] The sensory examination was completely normal at presentation, which is also reported by Linssen and Gabreels.[2] Sphincter dysfunction with palpable urinary bladder was present at admission. The onset of urinary problems coincided with the onset of paraparesis [11, 12, 13] but developed before the onset of sensory disturbances, which is contrary to what has been reported.[11, 12, 13] The upper-segmental level of neurological dysfunction developed during the stay in the hospital, which is definite and fixed in about 56% of patients.[11] In rest of the cases this level has reported to be ascending. In our case, this level did not ascend and there was no incidence of asphyxia as the cervical cord was unaffected. Our patient had involvement of the thoraco-lumbar spinal cord. According to literature the lesion occurs at the thoracic spinal cord level in nearly 80% of cases, the lumbosacral and

cervical regions are affected in about 10% of cases.[11] The muscle tone, power and deep tendon reflexes were all decreased, abdominal reflex was absent and a unilateral left sided babinski's was present.[2, 11] Signs of meningeal irritation were positive in our child which is consistent with reports published.[2]

As reported in literature, the CSF analysis revealed lymphocytic pleocytosis and an elevated protein level. Glucose concentration and pressure were normal. [11]

Magnetic Resonance Imaging (MRI) of thoracolumbar spine showed increased signal throughout the spinal cord except upper cervical region. There were focal areas of enhancement also seen throughout the cord more on lower region. According to the literature, half of the ATM cases show an enlargement of the spinal cord on T_1 weighted images. On, T_2 weighted images, an increased signal over some segments and across the complete cross section are noticed. The lesions are unifocal in more than 80% of the cases. [11, 14] MRI also excluded other lesions that can cause paraparesis.

Somatosensory evoked potentials (SEP) showed no abnormalities, which is comparable to literature.[2] They may show prolonged SEP latencies or missing SEP responses in conjunction with normal sensory nerve action potentials.[11]

Intravenous steroids are often instituted for patients with ATM. Though there is no randomized, placebo-controlled study that supports this approach, evidence from related disorders and clinical experience support this treatment. Additionally, several small studies support the administration of steroids in patients with ATM.[14] One study quoted from Belgium[1] states IVMPS as one of the most promising treatments for ATM. According to this study, IVMPS had a significant effect on the proportion of patients walking independently at one month and on the proportion with full recovery at one year. We treated our patient with intravenous methylprednisolone (IVMPS) in similar doses and time to full recovery in our case was two months.

According to literature, 44% have good outcome and either have no residual symptoms or are left with mildly disturbed micturition, minimal sensory loss or pyramidal tract signs.[11] Our patient had full recovery with no sequelae after two months.

The indicators for poor prognosis cited in literature are backache as the first symptom, rapid onset of paralysis in six hours, extensive hyperintensity on spinal cord T_2 weighted MRI, anterior horn cell involvement, signs of spinal shock and sensory level upto the level of cervical dermatomes.[11, 15] Our patient had backache as one of the symptom complexes but it was not precisely the first symptom. None of the other factors were present in our case, which could account for his rapid and complete recovery.

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