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ABSTRACT

Acute Transverse Myelitis (ATM) is a rare central nervous system (CNS) disorder in children and has variable prognosis.

We reported a case of ATM in a 10 year old Malay girl. She initially presented with sudden onset left sided hemiparesis and unable to walk. Otherwise she had no history of trauma, fever or headache.

Upon arrival at ED, patient was alert. The muscle's power of left upper and lower limbs was 2/5, hypotonia, hyporeflexia. Sensory was intact. CNS examination on the right side was normal. Other systems were unremarkable. Routine blood investigations and CT brain came back as normal. She was then admitted to ward for acute myelopathy and possibility of acquired demyelinating syndrome.

On second day of admission, patient developed right sided hemiparesis with reduced sensation at T5 level and below. Intravenous methylprednisolone was commenced as patient was treated as ATM. MRI of the spine revealed ill-defined T2-hyperintense lesion at the cervical cord extending from C3/C4 to C6 levels.

After completion of IV methylprednisolone for five days, patient showed significant improvement and able to ambulate with help upon discharged.

Because ATM is a diagnosis of exclusion, deliberate consideration should be given to the differential diagnosis including tumour, compressive myelopathy, infection and others. Therefore a careful history taking and thorough examination are essential in formulating the diagnosis. Spinal cord symptomatology in childhood ATM usually evolves over 2–4 days to peak at 5–6 days as was typically presented in this patient.

It has been suggested that the use of high-dose corticosteroids can reduce the length of disability and improve outcomes. In this case, early treatment with methylprednisolone has contributed to rapid recovery and reduced the morbidity.

INTRODUCTION

Acute transverse myelitis (ATM) is a serious medical condition with variable prognosis (1). It is a rare demyelinating and immune-mediated disorder of central nervous system (CNS) which involves the spinal cord (2). ATM contributes about 20% of children experiencing a first acquired demyelinating syndrome (1). Surveillance studies in the Canada and UK have estimated that the incidence of ATM is 2 per million children every year for the age group below 16 years. In the United States, roughly 300 paediatric patients affected annually (3). The female-to-male ratio is approximately 1:1 for < 10 years old, with a female predominance observed after 10 years old.

CASE REPORT

A 10 year old Malay girl with no previous medical illness presented with sudden onset left sided hemiparesis. It started after she woke up from an afternoon nap. She was well and able to attend her classes earlier in the morning. She also complained of numbness over bilateral upper and lower limbs. Otherwise she had no history of fever, fitting episode, headache, and also no previous history of trauma.

CASE REPORT (CONT.)

Upon arrival at Emergency Department (ED), patient was alert, conscious with full GCS. Vital signs recorded: blood pressure 115/80mmHg, heart rate 105 beats per minute, temperature 38 degree celcius, SPO2 99% under room air, glucometer 5.9mmol/L. CNS examination revealed the muscle's power of left upper and lower limb was 2/5 with hypotonia and hyporeflexia. Plantar reflex was equivocal and the sensory was intact.

Meanwhile, the examination of right upper limb and lower limb were normal. Her pupils were 2mm bilaterally reactive, no facial asymmetry, gag reflex was present. Other systems were unremarkable. Routine blood investigations and CT brain came back as normal. She was then admitted to ward for acute myelopathy and possibility of acquired demyelinating syndrome.

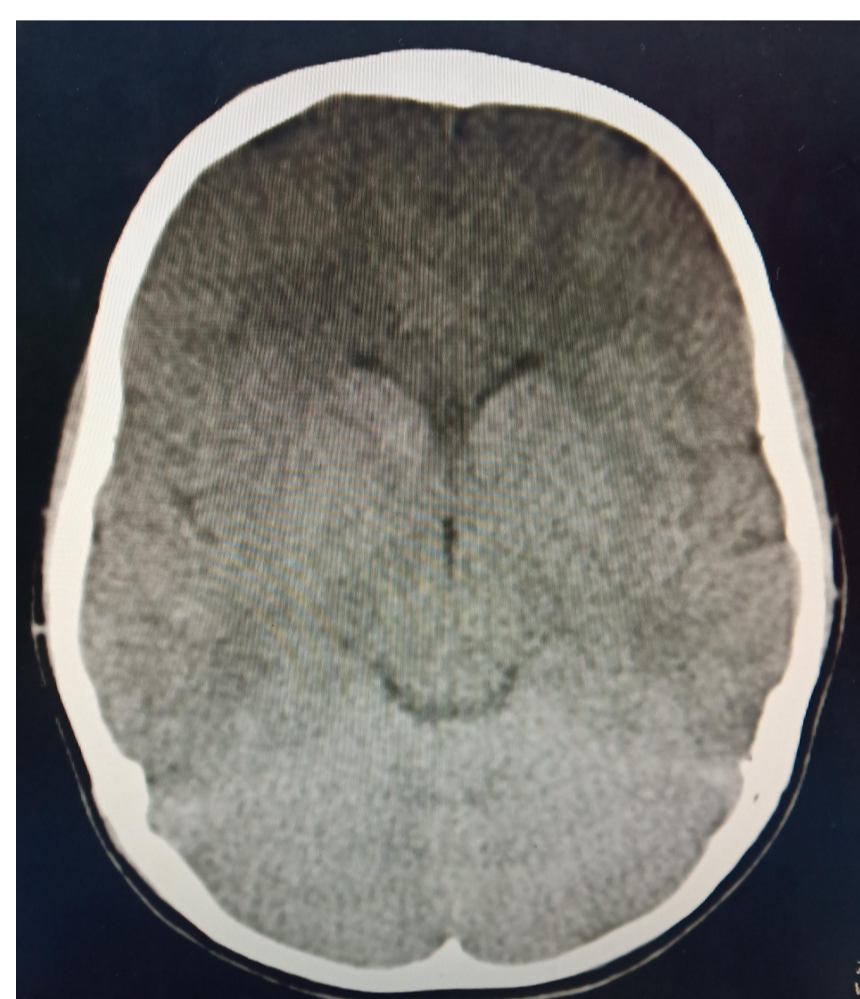


Figure 1. CT brain shows no abnormality on admission.

Subsequently, on second day of admission, patient complained of urinary incontinence and started to develop right sided body weakness and swallowing difficulty with persistent left hemiparesis. She also had reduced sensation at the T5 level and below. Nasogastric tube was inserted as her gag reflex was absent. Patient was then transferred to paediatric ICU (PICU) and high dose of intravenous corticosteroid, IV methylprednisolone was commenced as patient was treated as ATM. In PICU, patient developed mild respiratory distress thus she was started on high flow nasal cannula.

Urgent MRI of the spine was done and it revealed a long segment ill-defined T2-hyperintense lesion seen at the cervical cord extending from C3/C4 to C6 levels associated with cord oedema. This was in consistence with the diagnosis of ATM.



Figure 2. MRI spine shows ill defined T2-hyperintense lesion at the cervical cord extending from C3/C4 to C6 levels.

After few days, patient showed improvement in terms of muscle's power and tone. The oxygen supplement and nasogastric tube were off. IV methylprednisolone was completed for 5 days and oral prednisolone was then started. Patient showed significant improvement afterwards and subsequently she was able to ambulate with help upon discharged after almost a month in the hospital.

DISCUSSION

Challenges in making diagnosis

Because ATM is a diagnosis of exclusion, deliberate consideration should be given to the differential diagnosis including cerebrovascular accident, tumour, compressive myelopathy, ischemic myelopathy and infection (3). Therefore a careful history taking and thorough examination are essential in formulating the diagnosis.

Spinal cord symptomatology in childhood ATM usually evolves over 2–4 days to peak at 5–6 days (1). They can first presented with the symptom of back pain then develop motor and sensory deficit or bowel/urinary dysfunction. Most children will have urinary retention and require catheterisation as can be seen in this patient.

In 2002, the Transverse Myelitis Consortium Working Group has proposed the criteria for idiopathic ATM (4). This criteria also could be use a a guide in making the diagnosis of ATM.

Criteria for idiopathic acute transverse myelitis	
Inclusion criteria	
1.	Development of clinical dysfunction attributable to the spinal cord
2.	Defined sensory level and bilateral signs or symptoms
3.	Exclusion of extra-axial compressive etiology
4.	Inflammation within the spinal cord demonstrated by CSF pleocytosis or positive IgG index or gadolinium enhancement
5.	Symptoms from onset to reach maximal deficit between 4 hours and 21 days
Exclusion criteria	
1.	Previous radiation to the spine
2.	Clear clinical deficit attributable to anterior spinal artery occlusion
3.	Presence of arteriovenous malformations
4.	Serologic or clinical evidence of connective tissue disease
5.	Central nervous system manifestations of bacterial or viral diseases
6.	Brain MRI abnormalities suggestive of multiple sclerosis
7.	History of clinically apparent optic neuritis

Table 1. Criteria for idiopathic acute transversemyelitis (4).

Therapeutic measures

No medical therapy has been approved for patients with ATM in view of lack of controlled clinical trials (1). In treating ATM, medications are used based on experience and data from retrospective analysis primarily from studies involving adults. It has been suggested that the use of high-dose corticosteroids can reduce the morbidity and improve outcomes (3). It can be seen in this case that early treatment with methylprednisolone has contributed to rapid recovery and reduce the morbidity.

CONCLUSION

ATM is a serious and potentially life threatening condition to both adults and children. Early recognition and initiation of treatment are important to reduce the disability and the length of stay in hospital. Beside that, more clinical trials are needed to discover the most effective treatments for ATM among children in future.

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