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***Olgu Sunumu***

# Early-onset acute partial transverse myelitis: Case report

# Erken başlangıçlı akut parsiyel transvers miyelit: Olgu sunumu

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**ABSTRACT**

**Acute partial transverse myelitis is an acute inflammatory process of the spinal cord and it is a rare clinical syndrome. Acute partial cord lesions cause unilateral or markedly asymmetric sensory and motor dysfunction. In this paper, we report a case of 11 month-old girl who developed acute onset monoparesia following a viral respiratory infection which has not been described in the pediatric age group so far. Magnetic resonance imaging of the spinal cord disclosed expansive and high signal-intensity in the left part of the cervical region. A diagnosis of acute partial transverse myelitis was made and patient was treated with IV immunoglobulin and oral prednisolone. The child had eventually a good neurologic outcome by second month of the treatment.**

**Key words:** Acute partial servical transverse myelitis, childhood, treatment

**ÖZET**

**Akut parsiyel transvers miyelit spinal kordun akut inflamatuar bir durumu olup nadir bir sendromdur. Akut parsiyel kord lezyonları tek taraflı veya belirgin asimetrik duyusal ve motor disfonksiyona neden olurlar. Bu yazıda, viral solunum yolu enfeksiyonunu takiben akut başlangıçlı monoparezi gelişen 11 aylık bir kız olguyu daha önce pediatrik yaş gurubunda bildirilmemiş olduğundan sunduk. Spinal kanalın manyetik rezonans incelemesinde servikal bölgenin sol parçasında expansif ve yüksek sinyal yoğunluğu saptandı. Akut parsiyel transvers miyelit tanısı konuldu ve hasta iv. immunoglobulin ve oral prednizolon ile tedavi edildi. Çocuk tedavinin ikinci ayında iyi bir nörolojik gelişme gösterdi.**

**Anahtar kelimeler:** Akut parsiyel servikal transvers myelit, çocukluk çağı, tedavi

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## INTRODUCTION

Acute flaccid paralysis (AFP) is a clinical syndrome characterized by rapid onset of weakness, including (less frequently) weakness of the muscles of respiration and swallowing, progressing to maximum severity within several days or weeks. The term "flaccid" indicates absence of spasticity or other signs of disordered central nervous system motor tracts such as hyperreflexia, clonus, or hyperactive extensor plantar responses [(1)]. As a cause of AFP, acute transverse myelitis (ATM) is less frequent than Guillain-Barre syndrome or paralytic nonpolio enterovirus infection. The reported annual incidence of ATM is less than one case per 2 million population [(2)]. ATM is a focal inflammatory disorder of the spinal cord, resulting in motor, sensory, and autonomic dysfunction [(3)]. In the initial phase of spinal shock, the common manifestations include weakness of the lower extremities, AFP, urinary distention (neuroge-

nic bladder), constipation, hyporeflexia, sensory impairment, severe pain, and paresthesia. Complete transverse myelitis and partial transverse myelitis are different entities. Spinal cord damage in ATM generally affects all cord functions. Acute partial cord lesions cause unilateral or markedly asymmetric sensory and motor dysfunction. An urgent magnetic resonance imaging (MRI) is required to exclude acute spinal cord compression. MRI lesions are usually small, located in the lateral or posterior part of the spinal cord in the acute partial transverse myelitis patients (APTM) [4]. In this paper, we report a case of very early-onset APTM.

## CASE

A 11-month-old girl presented with acute onset weakness of the left upper limb, fever and generalized maculopapular rashes. Six day prior to her admission, fever and maculopapular rashes had become apparent. Because fever persisted she was admitted to the emergency service of our hospital and amoxicilline-clavulonic acid and ibuprofen were prescriped. On the same day she hadnat able to crawl because of left limb flask paralysis. His previous medical history and family history were unremarkable. At admission she presented with generalized maculopapular rash and physical examination demonstrated an asymmetrical lower motor neuron type of weakness affecting upper left limb with total inability to perform any movement. The muscle tone was decreased, the deep tendon reflexes of m. triceps, m.biceps and m.brachioradialis could not be elicited. No cognitive or cranial nerve disturbances were noted. Muscle power was considered as 1/5 on proximal and distal parts of the left limb. There was no meningeal irritation finding. Laboratory findings were as follows: hematocrit 30.3% white blood cell count [15000 cells/mm$^3$ (27% neutrophils, 67% lymphocytes)], and normal platelet count. Coagulogram, ionogram, blood urea nitrogen and creatinine concentrations and C-reactive protein were all within normal range. Cranial magnetic resonance imaging (MRI) was normal and spinal cord MRI disclosed an enlargement from C3 to C7 on sagital T2-weighed images (Figure 1). On transverse T2-weighed images, linearly increased signal intensity was noticed over the left spinal segments (Figure 2). These features were suggestive of APTM. Antibodies against HIV, herpes simplex virus, hepatitis, TORCH, Ebstein-Barr virus and mycoplasma

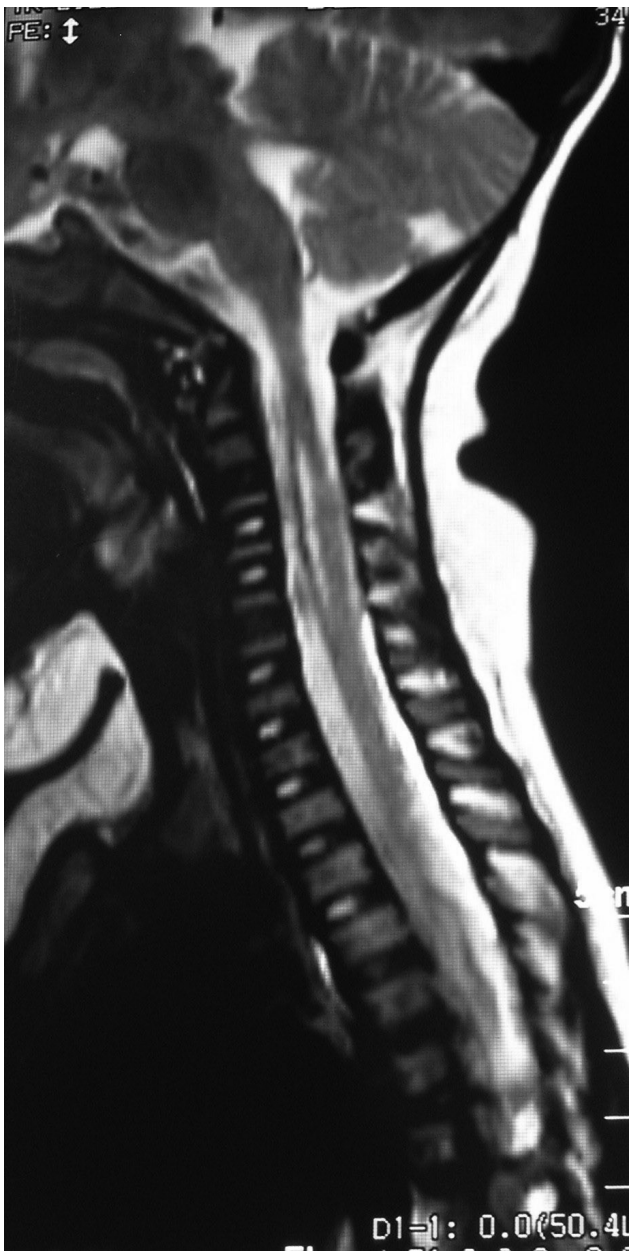


**Figure 1. Cervical MRI disclosed enlargement from C3 to C7 spinal segments on sagital T2-weighed images.**

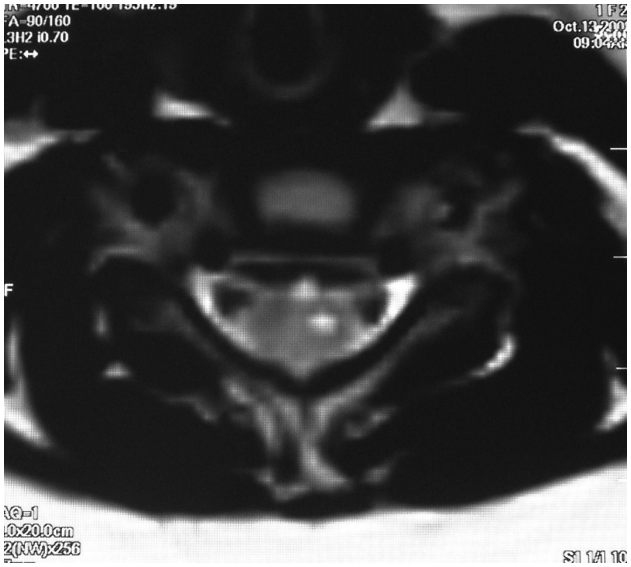

**Figure 2. Cervical T2-weighed images, linearly increased signal intensity was noticed over the left spinal segments on tranverse image.**

were not detected in the serum samples of the patient. During the follow up, maculopapular rashes regressed, after treatment with intravenous immunoglobulin (2 gr/kg) and muscle power was improved to 3/5 on the left limb. Then oral prednisolone (2 mg/kg/day) was started and she was assigned to included in the physical therapy programme. One month later control cervical MRI showed regressed expansion and edematous pattern. Prednisolone was tapered to every other day after 2 months, because of her cushingoid appearance. In the third month left limb muscle power was evaluated as complete, and the prednisolone therapy discontinued at the sixth month.

## DISCUSSION

AFP is a disease with sudden onset of weakness of a limb or paralysis over a period of 15 days in a patient less than 15 years of age. It may manifest itself as monoplegia, paraplegia, hemiplegia, facial palsy or any transient weakness. Concomitant disorders presenting with AFP are polio virus infection and vaccination sequelae, Guillain-Barre syndrome, brain stem stroke, encephalitis, acute transvers myelitis, ischemic myelopathy, compressive myelopathy, spinal radiation, intramedullary tumor, spinal arteriovenous malformation, infectious myelitis (e.g., Lyme disease, HHV-1, 6, mycoplasma pneumonia), diphtheritic neuropathy and porphyria [4]. ATM is a rare clinical syndrome in childhood. In this group it mostly occurs in children over 5 years of age [5]. Small children, 3 years of age or younger, develop spinal cord dysfunction over hours to a few days. They have a history of an infectious disease, usually of viral origin, or an immunization within the few weeks preceding the 1 first development of their neurological difficulties. The clinical loss of function is often severe and may seem complete. Although a slow recovery is common in these cases, it is likely to be incomplete. Paralysis begins as flaccidity, but over a few weeks spasticity develops [6].

Magnetic resonance imaging (MRI) is the primary modality of investigation in the diagnosis of ATM and also in differentiating this entity from other spinal cord lesions such as tumours [7]. Reports of MRI findings in patients with ATM have described local enlargement of the spinal cord and increased signal intensity. Commonly, three to four segments of the spinal cord were involved [8]. We found high signal intensity extending over several cervical spinal segments but only left part of the cord was affected, and a diagnosis of APTM was made.

Diagnostic criteria for TM include bilateral (not necessarily symmetric) sensorimotor and autonomic spinal cord dysfunction, clearly defined sensory level, progression to nadir of clinical deficits between 4 hours and 21 days after symptom onset, demonstration of spinal cord inflammation (cerebrospinal fluid pleocytosis or elevated IgG index) or MRI revealing a gadolinium-enhancing cord lesion, exclusion of compressive, postradiation, neoplastic and vascular causes [9]. After a mass lesion associated with spinal cord compression or complete subarachnoid column block from spinal cord swelling is ruled out, a lumbar puncture is indicated. The number of mononuclear cells is usually elevated minimally and the level of

CSF protein is elevated mildly. We couldnot perform lumbar puncture because her family didnot consent.

The term APTM has received somewhat less attention, but may also be defined [10]. MRI lesions of APTM tend to be smaller, often on the dorsal or lateral aspect. Complete ATM tends to be associated with large edematous-appearing lesions, which may expand the cord over multiple levels and produce T2-weighted abnormalities involving the central or (when viewed from axial view) entire cord. These syndromes may be distinguished by the extent to which spinal cord function is impaired. The term ATM refers to complete or near complete dysfunction at a spinal level, and APTM is defined as mild, patchy or grossly asymmetrical spinal cord dysfunction [11].

The clinical picture of ATM includes partial or complete paraplegia or quadriplegia, decrease or loss of deep tendon reflexes, sensory impairment and varying degrees of bladder and bowel disturbances. In a few patients, the disease has an ascending course with risk of asphyxia when upper cervical segments (C3-C5) are involved [12]. Our patient also had a sudden onset, and her left limb weakness with C3 to C7 cervical spine cord involvement was detected. Nevertheless she did not develop primary respiratory insufficiency, presumably owing to left side (partial) involvement she had a good neurological recovery.

The differential diagnosis of an AFP comprises a wide range of conditions. Bacterial abscesses, spinal cord tumors, vascular malformations, and hematomas can be usually ruled out by imaging modalities [13]. ATM is a clinical syndrome which can be associated with a number of different conditions. It is therefore necessary to identify either a direct infection, a systemic disease or an autoimmune (post-infectious or post-vaccinal) process bresponsible. A number of infectious agents have been incriminated in the ATM etiology. These include herpes simplex, herpes zoster, echovirus, hepatitis A-B, influenza, Lyme disease virus, HIV, mycoplasma pneumoniae, measles, mumps, varicella and Ebstein-Barr virus [14,15]. Our patient had no evidence of a systemic disease (like connective tissue disorder or malignancy), previous history of vaccination, radiation, or clinical evidence of infections caused by varicella-zoster virus, hepatitis, Ebstein-Barr virus, HIV or tuberculosis. Presumably our patient was an idiopathic APTM case. To our knowledge our patient will be the youngest APTM patient cited in the literature.

The management strategies have included supportive treatment and therapy for the causative agent. An intense search for etiological factors should be carried out, as therapy for a precisely identified factor may be beneficial in many instances. Treatment with high-dose intravenous methylprednisolone may be effective in children with ATM [10]. There are some case reports of patients who had improved with intravenous immunoglobulin therapy [11,16]. We preferred to use IVIG (2 g/kg) and then oral prednisolone (2 mg/kg) therapy in order to exclude the upper respiratory tract infection.

In conclusion, hence there has been no reported pediatric APTM case in the literature so far, we should emphasize investigations for the presence of APTM in the distinctive diagnosis of acute flask monoparesia in the childhood.

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