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Case Report

A child with acute transverse myelitis requiring permanent pacemaker implantation

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Abstract

We diagnosed a 3-year-old girl with acute transverse myelitis (ATM). She presented with weakness of the limbs and developed urination difficulty and respiratory disturbance. Magnetic resonance imaging revealed a symmetric area of high signal intensity on T2-weighted images involving the lower end of the medulla oblongata to the level of the fourth thoracic vertebra. Anti-aquaporin-4 antibody was negative. She was treated with intravenous methylprednisolone pulse therapy, immunoglobulin therapy, and plasmapheresis; however, her clinical symptoms did not change. At 10 and 20 days after symptom onset, cardiac arrest occurred on postural change, requiring cardiopulmonary resuscitation. A permanent pacemaker was implanted 23 days after onset. In the presence of sympathetic nerve hypofunction, relative hyperactivity of the parasympathetic nerves may have led to severe bradycardia and cardiac arrest in the presence of an inducer, such as a postural change. This is the first reported case of pacemaker implantation for management of ATM.

Keywords: ATM; Cardiac arrest; Treatment; Children

1. Introduction

Acute transverse myelitis (ATM) is an acute disease in which inflammation of the spinal cord induces motor paralysis, sensory disturbance, or dysautonomia [1,2]. Although some patients with cervical or superior thoracic cord lesions show autonomic symptoms involving the cardiovascular system, such as hypotension or bradycardia, therapeutic intervention is rarely required [1,2].

2. Case report

A 3-year-old girl presented with weakness of the left upper limb. She had no history of vaccination within 1 month before onset, antecedent infection, or trauma. She had no family history of cardiovascular diseases, including arrhythmias and sudden cardiac death. The following day, the patient developed weakness of the right upper and bilateral lower limbs, fever, and cervical

Abbreviations: acute transverse myelitis, ATM

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pain; therefore, she was emergently admitted. Paralysis of the limbs progressed and urination difficulty and respiratory disturbance appeared 2 days after onset of the paralysis. The patient was intubated and mechanically ventilated. Magnetic resonance imaging of the whole spine 3 days after onset revealed an area of slightly low signal intensity without enhancement effects on T1-weighted images and a symmetric area of high signal intensity on T2-weighted images involving the lower end of the medulla oblongata to the level of the fourth thoracic vertebra (Fig. 1A). Brain magnetic resonance imaging revealed no significant lesions involving the optic nerves. On blood testing, the levels of coagulation and fibrinolysis system markers were within the reference ranges, and antinuclear antibody, anti-dsDNA antibody, anti-SSA and -SSB antibodies, and anti-aquaporin-4 antibody were negative. A cerebrospinal fluid test showed an increased protein level of 59 mg/dL without pleocytosis. Oligoclonal bands and herpes virus in the serum and cerebrospinal fluid were negative by polymerase chain reaction, and no atypical cells or viral antigens were found in the cerebrospinal fluid, stool, or nasal discharge.

Based on the clinical course and imaging findings, we diagnosed the patient with ATM. She was treated with intravenous methylprednisolone pulse therapy (30 mg/kg/day, 3 days), immunoglobulin therapy (400 mg/kg/day, 5 days), and plasmapheresis; however, her clinical symptoms did not change. Follow-up magnetic resonance imaging 10 days after onset revealed a reduction in the size of the lesion, but enhancement effects were partially detected (Fig. 1B, C). On the same day, she developed sinus arrest during a passive postural change (Supplementary Fig. 1A). Recovery of spontaneous circulation was obtained after 40 s. Eleven hours after the initial arrhythmic event, sinus arrest recurred, requiring 4 min of cardiopulmonary resuscitation (Supplementary Fig. 1B). In the recovery phase, her electrocardiogram was normal without any conduction abnormalities, and her echocardiogram showed normal structure and function. Her serum electrolyte concentrations and thyroid function were normal. She had not been administered any drugs associated with sinus node dysfunction. We thereafter initiated low-dose continuous intravenous injection of isoproterenol and continued the intravenous methylprednisolone pulse therapy and plasmapheresis. However, 20 days after onset of the paralysis, a third arrhythmic event of severe sinus bradycardia followed by sinus arrest occurred, requiring 4 min of cardiopulmonary resuscitation. We implanted a temporary external pacemaker followed by a permanent pacemaker for recurrent life-threatening sinus pause. After implantation, the pacemaker was programmed in VVI mode at a pacing rate of 40 ppm. Pacemaker telemetry showed that her autopulse was 60–80 bpm. During 18 months of follow-up remote monitoring, no critical bradyarrhythmia appeared. Here serum catecholamine levels were undetectable. Rehabilitation improved her motor function; however, she became bedridden and upper limb-predominant paralysis remained 2 years after onset, but her intellectual outcome was favorable. Tracheotomy management and intermittent urethral catheterization were being continued at the time of this writing. We obtained informed consent about treatment and publishing from the patient's parents.

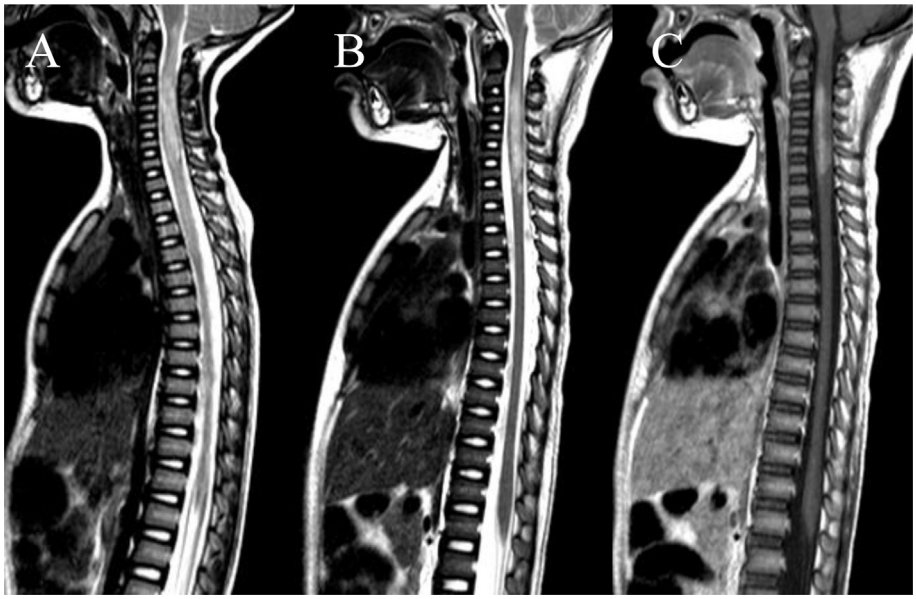

Fig. 1. Magnetic resonance imaging. **(A)** T2-weighted images revealed a symmetric area of high signal intensity involving the lower end of the medulla oblongata to the level of the fourth thoracic vertebra 3 days after onset. At 10 days after symptom onset, T1-weighted images showed **(B)** a reduction in the size of the lesion but **(C)** partial enhancement effects.