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Case report

Complex regional pain syndrome in a 15-year-old girl successfully treated with continuous epidural anesthesia

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Abstract

A 15-year-old girl developed severe pain in her right upper limb within a few days after she experienced an astatic epileptic seizure accompanied by falling on her right side. She was treated with fluid infusion through a cannula into her right hand. Swelling, mild flaring, and muscle weakness of the right arm subsequently appeared. Pharmacotherapy and stellate ganglion block were ineffective, and continuous epidural anesthesia was commenced 14 days after the falling event. The pain and accompanying symptoms completely disappeared within 5 days. Early treatment with continuous epidural anesthesia may be a promising option for the management of complex regional pain syndrome during childhood.

Keywords: Complex regional pain syndrome (CRPS); Reflex sympathetic dystrophy (RSD); Nerve block; Continuous epidural anesthesia

1. Introduction

Complex regional pain syndrome (CRPS) type I is a condition that is often triggered by trauma or blood puncture of the extremities, and it is characterized by devastating pain disproportionate in severity and persistence to the precipitating event, and various autonomic manifestations, including vasomotor instability and residual motor disabilities [1]. Onset of less than 10% CRPS cases happens during childhood and adolescence [2], and occurs predominantly in young adolescent girls. Although early initiation of treatment correlates with a better prognosis in CRPS [3], the relatively rare association of childhood CRPS with preceding traumatic events compared with adult CRPS and the under-recognition of this syndrome by pediatricians often result in considerable delay in proper diagnosis.

CRPS was formerly called reflex sympathetic dystrophy, but this term was discarded because the assumption of sympathetic nerve hyperactivity was not validated. The pathophysiology of the syndrome remains unclear, with no definitive hallmark for the prediction of outcome, and the effect of each treatment regimen varies considerably among individual patients. These factors

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have led to certain controversies regarding treatment strategy. The classic literature suggests that childhood CRPS is generally self-limited and has a better prognosis than adult CRPS, and that conservative management by physiotherapy with a mobilization program is sufficient [4]. Furthermore, some authors claim that psychological problems play a significant role in the pathogenesis of childhood CRPS, and that medication and nerve stimulation/block therapies are inappropriate because these allow family members to avoid facing the psychosocial issues [5]. However, the efficacy of physiotherapy for CRPS was not proved by a randomized control trial [6]. Residual pain in young adulthood has been recognized in at least half of sufferers [7]. There are children with refractory courses and/or persistent trophic sequelae [8] and invasive treatment, including sympathetic blocks, may need to be considered in some cases. Here, we report the case of a girl who showed early and complete recovery from CRPS, achieved by continuous epidural anesthesia.

2. Case report

The patient was a 15-year-old, right-handed girl who developed normally until the age of 9 years, when she developed weekly epileptic seizures with visual and/or olfactory aura accompanying impairment of consciousness. Brief seizures persisted after treatment with clobazam, occurring once or twice per year, and she was referred to our hospital for further evaluation. The neurological findings were unremarkable, and her intelligence quotient was assessed as 117 on the Wechsler Intelligence Scale for Children-IV, with scores in subscales of Verbal Comprehension Index 105, Perceptual Reasoning Index 113, Working Memory Index 120, and Processing Speed Index 113. Interictal electroencephalography (EEG) showed frequent spikes over the right frontotemporal area. Magnetic resonance imaging revealed a gadolinium-enhanced lesion at the right temporal tip (Fig. 1A and B). Thereafter, she experienced weekly episodes of nausea with trembling of the body and hand, visual hallucination of herself and family members, sensation of ear fullness, or brief visual disturbance. A 96-h video monitoring EEG under reduction of clobazam dosage identified four episodes of motion arrest with impaired consciousness coincident with the emergence rhythmic theta waves over the right frontotemporal region. Moreover, trembling of the body appeared several times, which was not accompanied by a change in EEG. As we had evidence of true epileptic seizures, lesionectomy was planned.

Two months later, the patient experienced a seizure with headache, nausea, and unconsciousness at school. She fell down and hit her right shoulder on the floor. She was referred to a local hospital, and was subjected to blood examinations and treatment with fluid infusion

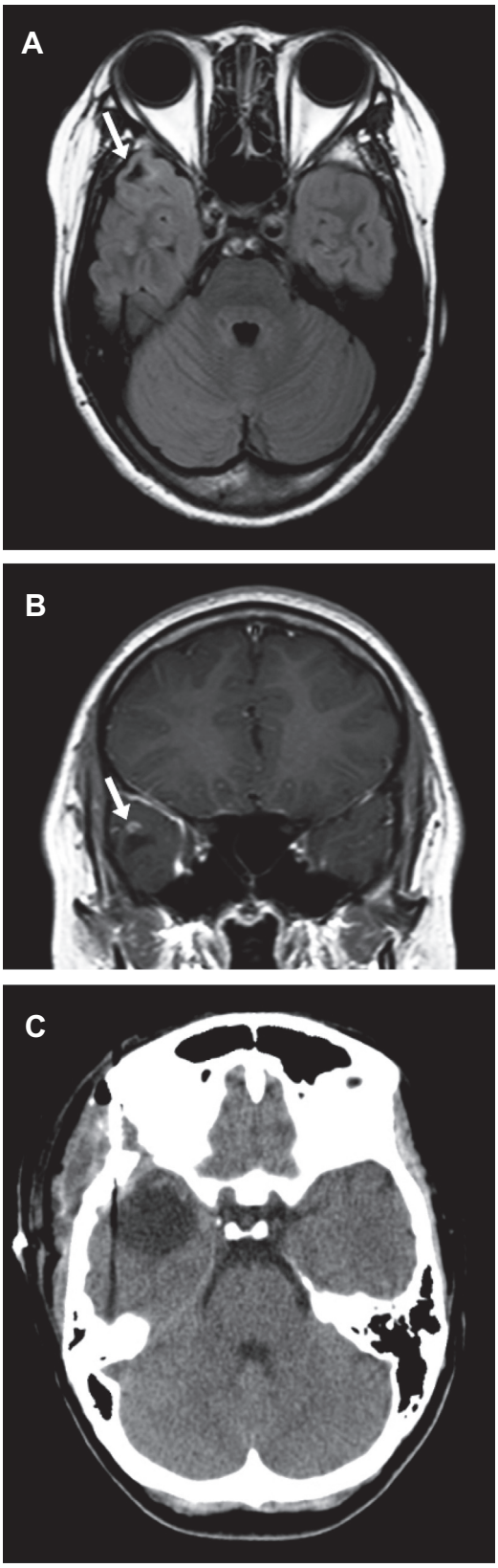

Fig. 1. Neuroimaging of the patient. (A) Fluid-attenuated inversion recovery image. (B) Gadolinium-enhanced image. (C) Computed tomography of the brain after lesionectomy. In A, a lesion with low signal intensity (arrow), partly rimmed by high signal striae, is visible. In B, a peripheral area of the lesion is enhanced (arrow).