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Case Report

Radiotherapy for Langerhans cell histiocytosis with paraplegia: A rare oncologic emergency case report in infancy and literature review

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Abstract

Background: Langerhans cell histiocytosis (LCH) is a clonal disease with focal or disseminated lesions that may compress the surrounding tissues, including the spinal cord. Because few reports have described the spinal symptoms as the first manifestation of pediatric LCH, the long-term neurological outcomes remain unclear.

Case report and literature review: We report a 21-month-old boy who presented with sudden-onset paraplegia. Imaging analyses revealed that osteolytic lesions and epidural tumors compressing the spinal cord at the T7-9 vertebrae. Twelve days after he developed leg weakness, emergency radiotherapy was started after a tumor biopsy. During the course of radiotherapy, paralysis steadily ameliorated. After we excluded infections and determined the pathological diagnosis of LCH, multi-drug chemotherapy was started. Apparent improvement in his complete paraplegia was observed after a total 15 Gy of radiotherapy and subsequent chemotherapy, leaving no neurological sequelae at 4 years of age. Through a literature search of studies published from 1980 to 2017, we found that children with LCH showed a generally favorable recovery from neurological dysfunction after the acute phase of spinal symptoms.

Conclusion: This report underscores the utility of emergency radiotherapy for the neurological recovery of spinal LCH in infants. Our long-term observation further denotes the value of this treatment in terms of the intact survival with preserved motor functions and physical growth.

Keywords: Langerhans cell histiocytosis; Spine; Paraplegia; Radiation therapy

1. Introduction

Spinal cord compression symptoms can present as the initial manifestation of posterior mediastinal tumors in childhood. The treatment choice of such an oncologic

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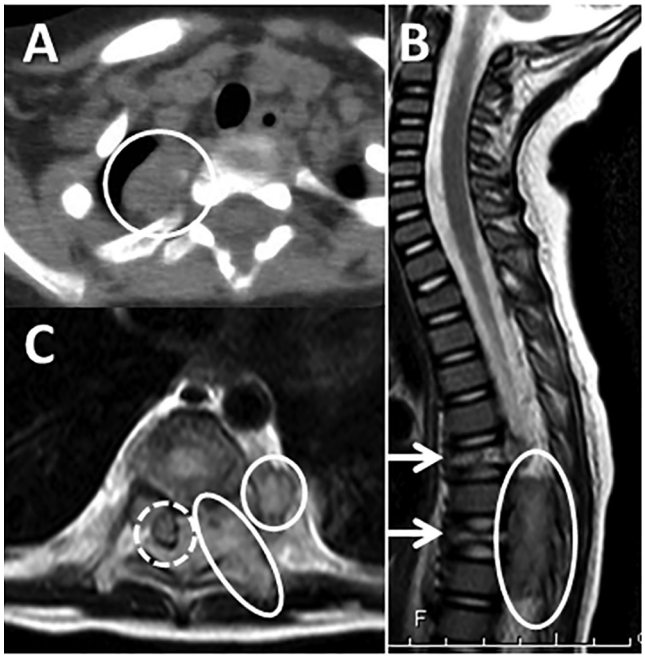

Fig. 1. Computed tomography and magnetic resonance imaging of the whole spine. (A) Mass lesion in the right upper lung. (B) T2-weighted images show compression deformity of the T6 and T8. (C) A mass lesion in the epidural region at the Th7-9 levels compresses the spinal cord (arrows). Ovals and dashed circles indicate the appearance of LCH tumor (A-C) and the rightward-deviated spinal cord (C), respectively.

emergency is challenging because of the variable treatment response of the tumor, its-involved sites, and the duration from the neurological onset.

Langerhans cell histiocytosis (LCH) is a clonal disease that involves one or more organ(s) [1]. Granuloma formation accompanied by eosinophilic and/or lymphocytic infiltration compresses the adjacent tissues. The majority of patients are under 10 years of age, and the spine is one of the most commonly affected sites [2]. However, spinal symptoms are rarely reported as the first manifestation of pediatric LCH [2,3]. We herein report a 21-month-old patient with LCH who presented with spinal cord compression symptoms.

2. Case report

A 21-month-old boy who had no history of developmental or growth delay was transferred to our hospital for the further study of his leg weakness. Two weeks earlier (day 1), he had begun to adopt a rigid, inflexible posture of the trunk and frequently grizzled without clear reasons. His mother suspected that he might have back pain. On day 4, he first visited our affiliated hospital because of cough, and a diagnosis of pneumonia was made based on the chest X-ray findings and increased levels of C-reactive protein (CRP) (10.0 mg/dL). Intravenous antibiotic therapy failed to improve the pain of the afebrile patient.

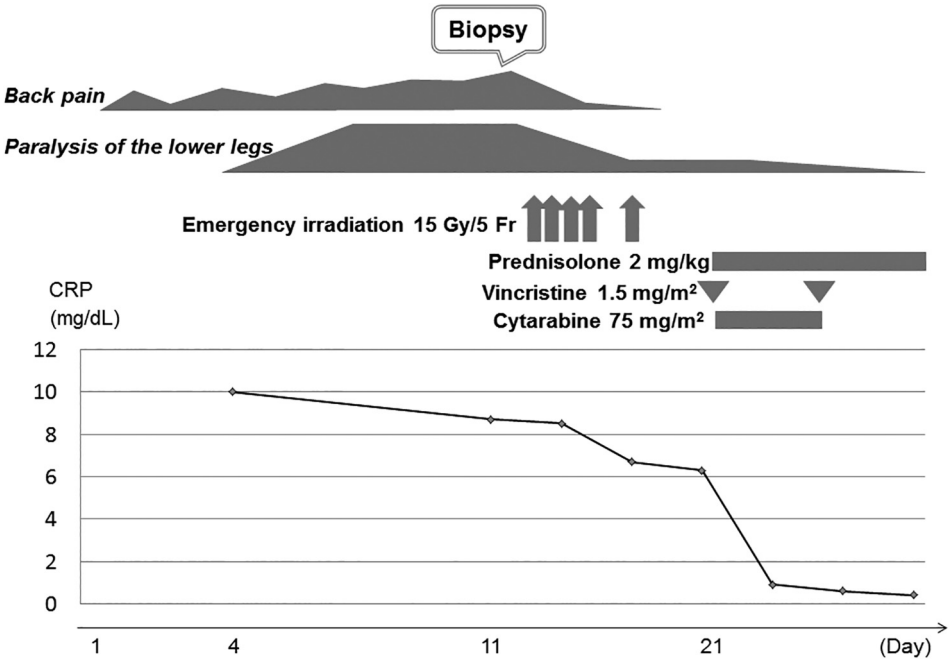

Fig. 2. Clinical course of the present case. This case was admitted to our hospital presenting with rapidly progressive back pain and paralysis (gray shades). At the 12th day of illness, the emergent radiotherapy was started (vertical arrows). The acute-phase treatment successfully relieved both back pain and paralysis within the next 2 weeks. The radiotherapy was followed by 2 mg/kg/day prednisolone for 42 days and the combined chemotherapy with 1.5 mg/m^2 vincristine and 75 mg/m^2 cytarabine. The ameliorating signs of this patient were correlated with the time course of CRP levels after admission (line plot at the bottom).

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