Subacute Sclerosing Panencephalitis Presenting With Hemiparesis in Childhood: Case Report
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What is This?
Subacute Sclerosing Panencephalitis Presenting With Hemiparesis in Childhood: Case Report

ABSTRACT

Subacute sclerosing panencephalitis is a chronic and fatal disease of the central nervous system. Most patients present with progressive psychosocial disturbances. A 14-month-old girl was admitted to our hospital because of left-sided hemiparesis. During hospitalization, focal and generalized seizures occurred. The electroencephalogram (EEG) revealed that periodic lateralized discharges consisted of polyspikes and high-voltage slow waves in the left hemisphere. Clinical and EEG findings and positive serology of measles antibodies in both serum (>1:24) and cerebrospinal fluid (>1:8) prompted us to study the cerebrospinal fluid and serum measles antibodies. Elevated measles antibody titers were detected in both serum and cerebrospinal fluid by complement fixation (1/512 and 1/16, respectively). The titers of measles IgG in the cerebrospinal fluid and cerebrospinal fluid/serum index were 71.5 mg/dl and 178.7, respectively. The patient was diagnosed with subacute sclerosing panencephalitis. The patient was put on carbamazepine, isoprinosine, and β-interferon therapy. There was no change in either neurologic deficit or mental assessment scores in her follow-up visit 10 months later.

Discussion

The diagnosis of subacute sclerosing panencephalitis is based on clinical manifestations, abnormal EEG findings, and high titers of serum (>1:24) and cerebrospinal fluid (>1:8) measles antibodies by complement fixation. Additionally, Baram et al reported a 22-month-old girl who had subacute sclerosing panencephalitis without cerebrospinal fluid measles antibodies. They detected measles virus genes that had typical multiple mutations for subacute sclerosing panencephalitis in postmortem brain-derived ribonucleic acid. Because the clinical and laboratory findings of our patient met the diagnostic criteria for subacute sclerosing panencephalitis, we did not consider a brain biopsy.

Generally, four of five clinical stages are used to describe the clinical course of subacute sclerosing panencephalitis. We did not observe the myoclonic jerks and/or akinetic drop attacks that are the typical features of clinical stages I and II. Thus, the classic progression of stages in subacute sclerosing panencephalitis did not apply to our patient.

The presenting symptoms in patients with subacute sclerosing panencephalitis are multiple and variable. Most patients present with subtle personality and mental changes. Decreased visual acuity, focal seizures, and headache or increased intracranial pressure are less frequent initial symptoms. Decreased visual acuity, focal seizures, and headache or increased intracranial pressure are less frequent initial symptoms. Decreased visual acuity, focal seizures, and headache or increased intracranial pressure are less frequent initial symptoms.
Poser et al pointed out significant variability in the EEG patterns in subacute sclerosing panencephalitis. The most characteristic finding in the EEGs of patients was periodic high-amplitude slow-wave complexes. They also reported that the characteristic periodic complexes would occur sooner or later. Accordingly, serial EEGs were required in patients with suspected subacute sclerosing panencephalitis. This was exactly what we have seen in our patient: in the first EEG, there were no typical features, but the next EEG was concordant with subacute sclerosing panencephalitis. Although the periodic complexes were usually bilateral, synchronous, and symmetric, asymmetric periodic complex amplitude was also defined in a recent study. The authors thought that the asymmetry of the periodic complex implied that one of the hemispheres was less damaged.

The radiologic staging of subacute sclerosing panencephalitis is mainly based on MRI results. Brismar et al reported that the severity of MRI changes did not always correlate well with the clinical findings. They showed that several patients with clinically advanced disease had normal or almost normal findings on MRI examinations. They and other authors noted that the progression of MRI abnormalities seemed to follow a constant pattern, with focal, high T2-intensity white-matter changes in the early period, atrophic changes in the later periods and loss of most of the white matter in the final stage of the disease.

The median interval between natural measles and the onset of subacute sclerosing panencephalitis was 8 years, with a wide range of 3 months to 14 years. The factors predisposing to a shorter latent period have not been explained clearly. Anlar et al hypothesized that a shorter latency might result from an inadequate immune defense against measles encountered at young ages. However, they did not find any correlation between the age at onset of measles and the duration of the latent period. Further, the latent period might also be affected by external factors, such as toxin or nutrition. Other viral infections might influence viral latency and reactivation. We accepted that our patient had measles subclinically. The effect of clinical or subclinical measles on the latent period was not known. But it was reported that children with measles who are younger than 1 year of age have a high risk of developing subacute sclerosing panencephalitis.

We report an infant with an atypical presentation for childhood subacute sclerosing panencephalitis. Although focal neurologic signs are frequent in adults with subacute sclerosing panencephalitis, they rarely manifest in children. This suggests that in infants with focal neurologic signs, subacute sclerosing panencephalitis should be considered.
We report a case of acute, painful polyneuropathy in a boy with newly diagnosed type 1 diabetes mellitus associated with a precipitous drop in hemoglobin A₁c. After initiation of insulin, the patient's hemoglobin A₁c dropped from 14.1 to 7.6%, and he developed severe pain in his feet, which prevented him from walking. Nerve conduction studies were consistent with mild to moderate sensorimotor peripheral neuropathy. Initially, he required opiate analgesics for pain control. Three months after presentation, the patient showed dramatic improvement and regained his ability to walk. Although not well described in the pediatric literature, this case represents insulin neuritis, one of the few diabetic neuropathies that has a favorable outcome.

**Acute Painful Neuropathy (Insulin Neuritis) in a Boy Following Rapid Glycemic Control for Type 1 Diabetes Mellitus**

**ABSTRACT**

We report a case of acute, painful polyneuropathy in a boy with newly diagnosed type 1 diabetes mellitus associated with a precipitous drop in hemoglobin A₁c. After initiation of insulin, the patient’s hemoglobin A₁c dropped from 14.1 to 7.6%, and he developed severe pain in his feet, which prevented him from walking. Nerve conduction studies were consistent with mild to moderate sensorimotor peripheral neuropathy. Initially, he required opiate analgesics for pain control. Three months after presentation, the patient showed dramatic improvement and regained his ability to walk. Although not well described in the pediatric literature, this case represents insulin neuritis, one of the few diabetic neuropathies that has a favorable outcome. (J Child Neurol 2003;18:365–367).

We report rapid-onset sensory and motor polyneuropathy in a child with type 1 diabetes mellitus associated with a precipitous decrease in hemoglobin A₁c. To our knowledge, there is only one other report of this association in a child, although it has been reported more frequently in the adult literature under the name “insulin neuritis.”

**Patient Report**

A 14-year-old boy of Hispanic origin presented to the pediatric neurology service with a 6-week history of paresthesias and pain in both lower extremities. The paresthesias were in the form of prickle and tinges. The pain, localized in his feet, was both sharp and burning in character. It occurred in response to pressure, that is, the touch of the floor against his feet. It was so severe that it prevented him from walking, and he stopped attending school. The patient also reported tingling in both hands. He had been treated with gabapentin and acetylsalicylic acid, with no improvement in symptoms. There were no complaints of incontinence, erectile dysfunction, change in power, fever, nausea, vomiting, diarrhea, or weight loss.

The patient had been diagnosed with diabetes mellitus type 13 months previously, when he presented with ketoadidosis. Hemoglobin A₁c at that time was 14.1% (normal range 4.6–6.5%). A diabetes autoimmune panel had been obtained prior to the initiation of insulin therapy. Human insulin antibodies were 7.4 µU/mL (normal < 5), anti-GAD65 antibodies were 3.7 U/mL (normal < 0.5), and ICA 512 (islet cell) antibodies were not present. The patient was started on a typical dose and schedule of Lente and lispro insulins. His diabetes quickly became controlled, with a hemoglobin A₁c of 7.6% 9 weeks after diagnosis (target range for age is 7–8%).

Examination was remarkable for absent muscle stretch reflexes at the left knee and both ankles. The right knee and upper extremity muscle stretch reflexes were barely elicitable. He showed 5 of 5 strength. Sensory examination showed allodynia when pressure with a hand or pin was applied to the skin below the knees bilaterally. Proprioception, vibration, and warm/cold differentiation were intact. He walked with an antalgic gait.

Nerve conduction studies performed at presentation (6 weeks into the course of his symptoms) showed slowing of motor velocity and reduced amplitude of sural nerve sensory responses (Table 1). The results were consistent with a mild to moderate sensorimotor peripheral neuropathy. Other laboratory results at the time of evaluation included a normal rapid plasma reagin (RPR) as well as normal thyroid-stimulating hormone, vitamin B₁₂, and folate levels.

The patient’s neuropathy was attributed to the rapid improvement in hyperglycemia, as demonstrated by the significant drop in hemoglobin A₁c. His insulin was adjusted downward to loosen his diabetes control, in hope of improving his neuropathic symptoms. The patient was started on phenytoin for the sharp, jabbing pain, gabapentin was continued, and acetylsalicylic acid was weaned. Seven weeks after onset of symptoms, the patient stopped all medication. An electromyogram (EMG) performed 10 weeks after presentation was essentially the same as the first EMG. Three months after onset of symptoms, the pain had significantly decreased. Examination at that time showed absent muscle stretch reflexes in all extremities; the sensory examination was normal. He was able to walk normally. Hemoglobin A₁c was 8.0%. Owing to complaints of being depressed, he was referred for outpatient psychiatric treatment.

**Discussion**

In the current case report, a temporal relationship between the onset of painful sensory neuropathy and the improvement in hemoglobin A₁c following initiation of insulin therapy was identified in a 14-year-old child with diabetes mellitus type 1. Such a rapid decline in the hemoglobin A₁c to the target range is not frequently seen after the initiation of therapy in a patient with new-onset diabetes, and it was hypothesized that this may have contributed to the patient’s presentation. Although it has been known for many years that chronic diabetes is associated with painful peripheral neuropathy in adults, less is known about the acute painful neuropathy associated with the short-term establishment of glucose control, particularly in the pediatric population. In 1933, Caravati might have been the first to describe insulin neuritis, the painful neuropathy immediately following initial treatment of diabetes with insulin. Symptoms of this syndrome include burning pain in the feet more than in the hands, paresthesias, and allodynia. The pain is severe, limiting function, and often disrupts walking and daily activities. Insulin neuritis can also be associated with profound weight loss and severe depression (diabetic neuropathic cachexia). It is self-limited and responds to simple, symptomatic treatment.