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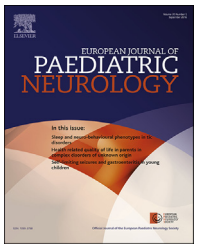

Case Study

Acute inflammatory demyelinating polyradiculoneuropathy in a newborn infant

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ABSTRACT

Background: Acute inflammatory demyelinating polyneuropathy (AIDP), also known as Guillain-Barré syndrome, is an immune-mediated polyneuropathy usually triggered by infections or vaccinations. In childhood AIDP is commonly described after the first year of life. Here, we present a case of a newborn infant with AIDP manifestation directly after delivery.

Case study: A newborn girl with a healthy mother, without known exposure to immunomodulating factors, was admitted to the neuropediatric department due to ascending hypotonia, weakness, pain and areflexia in the lower extremities. The clinical presentation, laboratory and neurophysiological studies supported the diagnosis of AIDP. The infant showed first signs of clinical improvement following administration of intravenous immunoglobulin and her recovery was complete at one year.

Conclusion: AIDP should be considered as a differential diagnosis in ascending hypotonia also in the neonatal period.

1. Introduction

Acute inflammatory demyelinating polyneuropathy (AIDP), commonly known as Guillain-Barré Syndrome, is an immune-mediated polyneuropathy usually triggered by infections or more rarely vaccinations. In childhood it generally occurs after the first years of life but can manifest earlier as demonstrated by cases with infection-related AIDP in the first year.[1] There are also a few reported cases of congenital AIDP in infants whose mothers had a chronic or acute autoimmune disease[2–4] and one infant to a healthy mother who presented with AIDP during the first week of life.[5] As this represents an important differential diagnosis in neonatal infants with pronounced and progressive hypotonia, we here report a case of congenital AIDP in a female infant without known exposure to immunomodulating factors or infections.

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2. Case report

A Caucasian girl was delivered with planned cesarean section due to reduced fetal movements, oligohydramnios, previous traumatic delivery for the mother and borderline pelvis diameter at gestational age 37 weeks + 1 day. The mother, otherwise healthy and with two previous normal pregnancies, reported reduced fetal movements at the 29th gestational week which motivated weekly ultrasounds until week 36. The serial ultrasounds showed that the fetal growth was within normal curves (−5%, Fig. 1). They also showed a decreased amniotic fluid index which varied between 25 and 78 mm; index was almost normalized at week 35 (78 mm) but then decreased again in week 36 + 4 (25 mm). The examinations confirmed the decrease of fetal movements. No signs of fetal distress were evident.

Serology for toxoplasma, rubella, cytomegalovirus (CMV), herpes simplex virus (HSV) type 1 and 2 and parvovirus was normal in week 33. The combined ultrasound and biochemical (CUB) test was also normal.

The newborn girl had birth weight 2750 gg, length 48 cm and head circumference of 33. The Apgar score was 10 at 1 min, 9 at 5 min and 7 and 10 min. The newborn showed a mild respiratory distress at around 7 min and needed positive pressure with mask but then had a normal respiration at 15 min. It was however evident that the infant did not display normal movements in her legs and both plantar and patellar reflexes were absent bilaterally. Moreover, both legs but in particular the left one, were quite swollen with long capillary refill time of about 6 s. During the first hours of life a more generalized hypotonia, including the arms and the trunk, was evident but with otherwise normal peripheral oxygen saturation, heart and respiratory rate. The infant was not interested in eating and received a naso-gastric tube. Blood tests showed no signs of infection during postnatal days 1 and 2 and both ultrasound of the brain and radiology of the legs were normal.

The hypotonia progressed slowly over the following days and she was perceived as weaker and less responsive

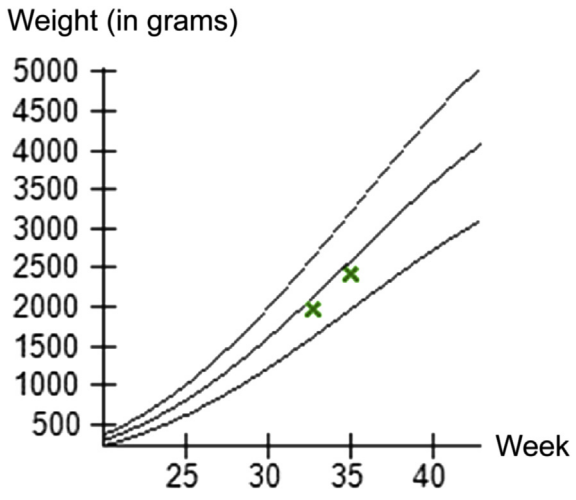

Fig. 1 – Growth curve at 34 + 3 and 36 + 1 weeks of gestation.

although breastfeeding was soon initiated satisfactory. Magnetic resonance imaging (MRI) of the spine and brain on postnatal day 3 and MRI of the spine with angiography on day 7 merely showed a minimal intraspinal sacral lipoma without clinical significance. A lumbar puncture was performed on the 5th day and showed cerebrospinal fluid (CSF) leucocytes: 274 ($\times 10^6$/L), monocytes: 134 ($\times 10^6$/L), albumin 950 mg/L (normal:<225 mg/L), and erythrocytes 183,200 ($\times 10^6$/L) due to traumatic procedure. A second lumbar puncture on the 13th day was again traumatic. Electroneurography (ENeG) performed on the 6th day displayed long distal latencies in tibial nerves with conduction velocities less than half of normative values for age and no F-responses upon stimulation of the right tibial nerve. Sensory evoked potentials of the tibial nerves showed no evidence of a central component in the paralysis. The findings were considered indicative of demyelination as seen in AIDP or neurometabolic disease.

Repeated ultrasound of the legs showed no signs of a venous thrombosis but radiology of the lower limbs revealed a left proximal tibial fracture. The orthopedic follow up manifested bilateral acetabular dysplasia that was managed with hip abduction braces. She also had a systolic murmur upon cardiac examination and a subsequent cardiac ultrasound revealed a small patent foramen ovale without hemodynamic significance. An eye examination was also performed with normal findings.

Due to the progressive fatigue, the clinical signs and the neurophysiological examination a diagnosis of AIDP was considered as highly possible and a five-day treatment with intravenous immunoglobulins (IVIG, 0.4 mg/kg daily) was initiated at day 8. In parallel, diagnostic work-up for other possible causes of the infant's paralysis continued. On postnatal day 12 a repeated ENeG showed unaltered values with very low nerve conduction velocities in the legs and even in the right medianus nerve indicating a pronounced sensorimotor demyelinating polyneuropathy (Table 1). Muscle responses upon proximal stimulation were highly fragmented with temporal dispersion. Electromyography (EMG) of right tibialis anterior and right vastus lateralis muscles on the same day recorded brisk spontaneous activity with fibrillations and positive sharp-waves in both muscles indicating significant axonal injury. No motor units could be recorded from tibialis muscle but bursts of low amplitude motor units were seen from vastus lateralis without any movement in the leg. The neurophysiological follow up supported the initial findings of acute demyelinating polyradiculopathy with axonal damage.

As part of the continued diagnostic work-up she was found to have normal values for blood count, all infectious parameters, TPO (*thyroid peroxidase*) and TRAK (thyrotropin receptor *antibodies*) antibodies, antinuclear antibodies (a positive ANA staining pattern was considered nonspecific), neuronal intracellular and surface antibodies, coagulation tests, homocysteine, cobalamin, methylmalonic acid, MAG (myelin associated glycoprotein)-antibodies, gangliosid (GM1, GM2, GM3, GT1b, GQ1b, GD1a, GD1b)-antibodies, sulfatide-antibodies, neurometabolic workup with plasma aminoacids, p-acetylcarnitine, urinary organic acids, u-creatinine, s-CMV/HSV/rubella/toxoplasma, csf-enterovirus/HSV/VZV (varicella zoster virus), f-enterovirus and s-campylobacter jejuni/CMV/enterovirus/rubella. Analyses for EGR2 (early growth