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Clinical Letter

A Serendipitous Case for Shorter Steroid Course in Infantile Spasms

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Background

The treatment of infantile spasms (IS) has remained a topic of discussion given the limited efficacy and multiple adverse effects of currently available treatments. Hormonal therapy, including intramuscular adrenocorticotropic hormone (ACTH) and oral prednisolone, is first-line in the treatment for most patients with IS.[1] Given the cost-effectiveness and feasibility of administration, our institution favors high-dose oral prednisolone over ACTH in the treatment of IS, following the protocol of the United Kingdom Infantile Spasms Study, which utilized two weeks of steroid therapy followed by a 15-day taper.[2–4] However, using steroids or ACTH to treat IS carries a risk of immunosuppression and can predispose infants to primary infections or reactivation of previous infections with potentially devastating consequences.[5] Here, we present an infant who developed a varicella-zoster virus (VZV) infection in the setting of high-dose steroids for IS.

Patient description

This previously healthy five-month-old infant with up-to-date immunizations presented to the emergency department after three days of abnormal movements. His parents described clusters of intermittent extension of his arms with simultaneous flexion of his hips that occurred around the sleep-wake transition, consistent with IS. On examination, he had several pustular lesions on his back and occiput. His neurological exam was normal. Routine electroencephalography showed classic hypsarrhythmia. He was treated with 15 mg of prednisolone three times daily. Over the first 48 hours of therapy (six doses), his spasms continued and the rash worsened (Fig. 1). A skin lesion was cultured and was positive for VZV DNA. He was started on acyclovir at meningitic dosing. On further questioning, his family reported that the infant had come into contact with an uncle with a recent shingles infection. The lesions continued to worsen and prednisolone was stopped after 12 total doses due to concern for worsening varicella infection in the setting of immunocompromise. Alternative therapies were discussed, including vigabatrin and the ketogenic diet. However, by day five at the hospital, his clinical spasms ceased. On day six at hospital, cerebral spinal fluid examination was normal, including negative VZV polymerase chain reaction. He completed a

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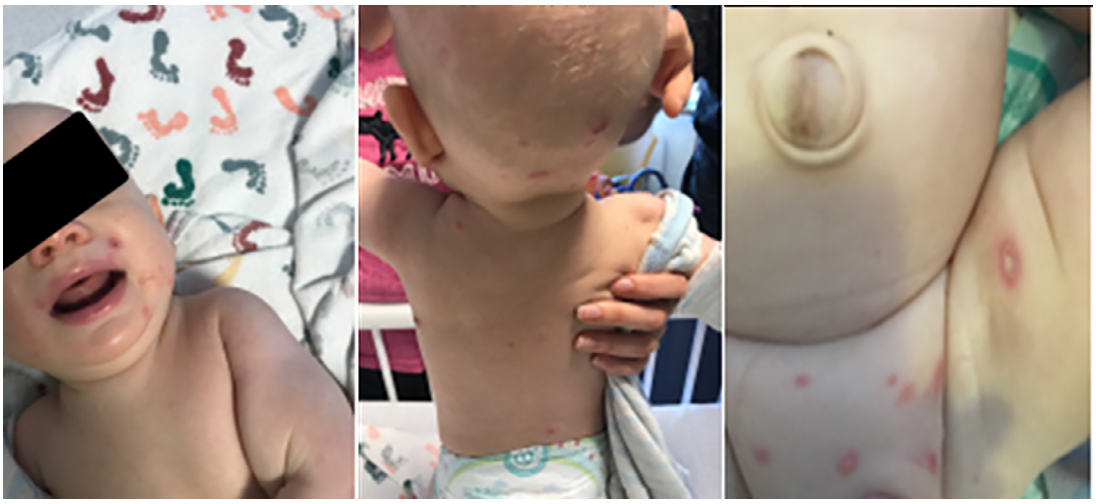

FIGURE 1. Vesicular lesions of the face, occiput, and thighs, consistent with varicella-zoster virus.

10-day course of acyclovir and his rash resolved. He had no further spasms, brain magnetic resonance imaging with and without contrast was normal, and his electroencephalography on day seven was normal. Now at age nine months, he is developing normally, without further spasms and without antiseizure medications.

Discussion

Given the history of exposure to a family member with shingles, the rash with positive viral culture, and the improvement on acyclovir, our patient's course was consistent with a diagnosis of VZV infection in the setting of high-dose steroids for IS. While the use of steroids put him at risk for complications of VZV, the risk of developmental regression from IS was initially thought to be greater than that of worsening VZV infection, particularly given the negative cerebral spinal fluid VZV polymerase chain reaction and absence of clinical features suggestive of central nervous system (CNS) infection.

IS typically affects children too young to receive certain immunizations, exacerbating the risk for infection when treated with steroids. Our patient had outgrown maternal antibody coverage for VZV but had not yet received the standard varicella immunization at one-year of age. This lapse in coverage and his exposure led to development of a mild VZV rash that worsened in the setting of steroids.

Although previous literature has described an association between infection and the development of IS, particularly that of congenital cytomegalovirus affecting the CNS,[6] no cases of VZV infection have been reported in association with IS, with or without CNS involvement.

Although our patient received a briefer course of steroids than typical for IS, his spasms and hypsarrhythmia resolved. This suggests that a shorter duration of hormonal therapy in patients with IS may be advisable. Future research should investigate the optimal length of therapy for IS to balance treatment of spasms against adverse side effects.

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