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Life-Threatening or Organ-Impairing Henoch-Schönlein Purpura: Plasmapheresis May Save Lives and Limit Organ Damage

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Abstract
Adult-onset Henoch-Schönlein purpura (HSP) tends to become chronic-relapsing, yet rarely leads to organ impairment, e.g. due to chronic glomerulonephritis. Bed rest, compression and nonsteroidal anti-inflammatory drugs are usually sufficient to control the active phases. We report 2 cases of adult HSP with an unusually severe evolution. One patient required intensive-care treatment for hypovolemic shock caused by hemorrhagic pancolitis; the other had progressive and extremely extensive vasculitic leg ulcers. Both were refractory to common immunosuppression with systemic corticosteroids (oral and pulse) and additive steroid-sparing immunosuppressive drugs. Only after the introduction of plasmapheresis did both patients show a dramatic improvement in the disease, with rapid and almost complete healing. Plasmapheresis is a rarely used therapeutic tool in the treatment of severe HSP, but the growing literature on its highly beneficial effect underlines its potential usefulness.

Introduction

Henoch-Schönlein purpura (HSP) is a systemic small vessel leucocytoclastic vasculitis characterized by vascular deposition of IgA immune complexes. It mostly affects children, being the most common vasculitis of childhood with 50% of all cases occurring before the age of 5 years [1]. The exact pathogenesis of serum IgA immune-complex-level dysregulation remains unknown; however, group A streptococcal tonsillitis/pharyngitis and unspecific upper airway infections are the most common triggering factors [2]. HSP has also been reported to appear in association with malignancy, mostly with solid tumors but also in the setting of plasma cell dyscrasias like multiple myeloma [3–7], and also as a first manifestation of myeloma [5–7]. The disease typically presents with a classic palpable purpura, mostly involving the lower extremities, due to erythrocyte extravasation into the skin. Polyarthralgia, usually of the knees and ankles, is present in more than 80% of patients [1]. More than half of the patients have colicky abdominal pain, which can be associated with mild bloody diarrhea. The most feared feature of HSP is glomerulonephritis (IgA nephropathy), occurring in 40–50% of patients and manifesting in most cases as proteinuria, microhematuria and erythrocyte casts. Nephropathy represents the most common cause of morbidity and death related to HSP, with 12–19% of children progressing to transient renal failure and up to 3% to chronic end-stage renal disease [8, 9]. Other rare complications include central nervous system vasculitis, orchitis, and cardiac and ophthalmological involvement [10].

HSP usually has a spontaneous self-limiting favorable evolution, with up to 95% of affected patients showing a complete remission within a few weeks. The overall prognosis is excellent, even with one third to one half of patients having 1 or more recurrences of symptoms (usually within 6 weeks, but also as late as 3–7 years) afterwards [11].

Clinical manifestations in adult-onset HSP tend to be more pronounced and recurrences are more frequent. In rare cases, HSP can become organ-compromising or even life-threatening. The 2 reported cases are good examples of refractory or even life-threatening HSP, for whom systemic corticosteroids, both oral and intravenous pulse, and steroid-sparing agents did not stabilize the disease. Eventually, plasmapheresis led to a rapid and long-lasting remission in both patients.

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