Peripheral gangrene in children with atypical hemolytic uremic syndrome.

Abstract

Atypical hemolytic uremic syndrome (aHUS) is a thrombotic microangiopathy with severe clinical manifestation, frequent recurrence, and poor long-term prognosis. It is usually caused by abnormalities in complement regulation. We report 2 cases of children affected by a catastrophic extrarenal complication. A 4-year-old Indian girl developed gangrene of the finger tips 2 days after initial presentation of aHUS. Factor H autoantibodies were identified. Renal function continued to decline despite daily plasma exchanges, and she was started on peritoneal dialysis 5 days after admission. The distal tips of the left hand remained gangrenous with a line of demarcation. Three weeks later, she did not return for follow-up and died at home because of dialysis-related complications. An Arabic girl developed end-stage renal disease due to aHUS in the fourth month after birth. A de novo activating C3 mutation was found. At age 9 months, she suddenly developed ischemic changes in fingers of both hands and several toes. The lesions progressed, and several finger tips became gangrenous despite intense plasma exchange therapy. The
decision was made to administer complement blocking therapy with the C5 antibody eculizumab. All nonnecrotic digits rapidly regained perfusion. The 3 already gangrenous fingers healed with loss of the end phalanges. During maintenance, eculizumab aHUS activity subsided completely and some late recovery of renal function was observed. aHUS may present by thrombotic macroangiopathy of small peripheral arteries. Eculizumab appears effective in preserving tissue viability if administered before gangrene occurs and should be considered as first-line rescue therapy in such cases.

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